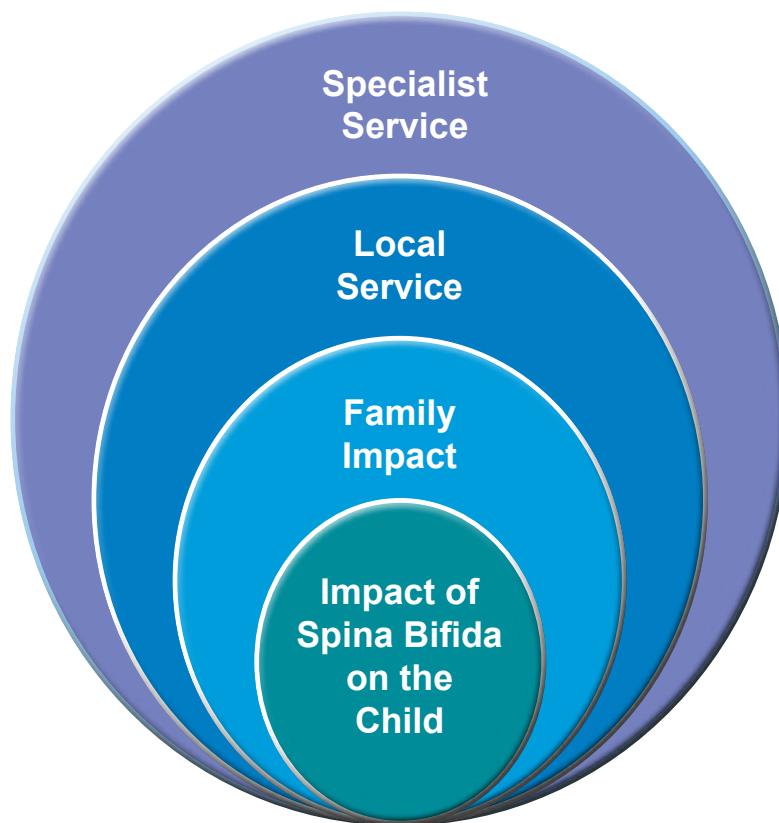


# **The Health and Therapy Needs of Children with Spina Bifida in Ireland**



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This report has been prepared in good faith on the basis of information available at the date of publication without any independent verification. Readers are responsible for assessing the relevance and accuracy of the content of this publication.

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# Executive Summary

## The Health and Therapy needs of Children with Spina Bifida in Ireland

### Chapter 1: Background

Spina bifida is a type of Neural Tube defect (NTD) which has been described as one of the most complex congenital conditions compatible with life (Liptak and El Samra, 2010). Ireland has one of the highest rates of NTDs in the world with a prevalence of 1.17 per 1000 live births (McDonnell et al., 2014). It is estimated that there are at least 500 children (0-18 years) with spina bifida currently in Ireland (EUROCAT, 1991).

Spina bifida can affect nearly all body systems to some degree depending on the level of the lesion; with potential physical, motor, sensory, continence, cognitive and psychosocial difficulties (Northrup and Volcik, 2000). The life-long impact of spina bifida can have an estimated health services cost of €500,000 (Yunni et al, 2011).

Provision and coordination of care for children with spina bifida along with multidisciplinary team (MDT) working is essential to optimise health and therapy outcomes (Kinsman et al., 2000, Brustrom et al., 2012, Aldana et al., 2010). Temple Street Children's University Hospital (TSCUH) is the national paediatric neurosurgical centre providing care for all children born with spina bifida since 2008. TSCUH also provides a MDT Spina Bifida clinic to support these children and families. This is consistent with international best practice (Merkens, 2006).

As a result of reported service inequities and the need to develop and improve services, a review of the health and therapy needs of children with spina bifida and their access to local and specialist services was undertaken. This project was completed by an Occupational Therapist, Physiotherapist and Consultant Paediatrician and was funded by the Children's Fund for Health within TSCUH fundraising department.

### Chapter 2: Aims and Methodology:

The overall aim was to identify the health needs of children (aged 0 – 18 years) in Ireland with spina bifida. Specific objectives included:

1. To identify the availability of current services for children with spina bifida and make recommendations regarding children's health and therapy needs
2. To highlight the impact of spina bifida on children and their families
3. To provide an up to date literature review of international best practice

#### 4. To gather feedback regarding the MDT spina bifida service in Temple Street

This mixed-method study gathered information from service providers (professionals working with children with spina bifida throughout Ireland) and parents / guardians over two phases (phase 1: quantitative questionnaires, phase 2: qualitative interviews). Ethical approval was obtained from the ethics committee of TSCUH. A focus group was completed with children (8-18 years) which was aided by the completion of the KINDL spina bifida quality of life questionnaire.

Quantitative data gathered from questionnaires was analysed with SPSS using correlations, frequencies and means. Qualitative data was transcribed verbatim and analysed using thematic analysis as described by Braun and Clarke (2006).

### Chapter 3: Samples

Questionnaires were completed by 155 parent/guardians from a national spread which represented a 30% response rate of the total estimated population. 86% of respondents were mothers who represented children with a mean age of 5.7 years. Qualitative interviews were completed with 26 parents.

247 service providers completed questionnaires. They were represented by Physiotherapists (41%), Occupational Therapists (32%), Speech and Language Therapists (8%), Paediatricians (3%), Psychologists (4%), Clinical Nurse Specialists (2%), Social Workers (2%), Public Health Nurses (1.6%), Dietitians (1.2%) and other (6%). Service providers had an average of 8.5 years' experience and 7.02 children with spina bifida on their open caseloads. 25 service providers participated in qualitative interviews.

4 children participated in a focus group consisting of an 8 and a 10 year old boy, and a 13 and a 15 year old girl.

### Key Findings

#### Chapter 4: Impact of spina bifida on children and families

A high proportion (20%) of the sample reported having a family history of Neural Tube Defects.

Children with spina bifida present with a variety of complex needs affecting multiple body systems and impacting on their ability to actively participate in daily life.

- 64% have a CSF shunt in place to manage hydrocephalus.
- 69% of children over 3 years require a wheelchair.
- 93% of children over 5 years require support to achieve continence.
- 55% have visual difficulties.
- Obesity is 5 times more prevalent than in the typically developing population.

- 14% have an intellectual disability.
- 50% of school-age children have low self-esteem.
- 96% require ongoing physiotherapy, 87% require ongoing occupational therapy.
- 89% of school-age children access mainstream schooling; however access can be dependent on whether supports for physical and toileting needs are available.
- 43% of children accessing special education fall within the average cognitive range.

Half of the school-aged children suffered from low self-esteem, which was influenced by age and difficulties with continence and weight. Social, emotional and psychological issues were prevalent against a background of limited availability of ongoing psychological support.

### **Impact of spina bifida on families**

The impact of spina bifida on the wider family unit was identified. Parents reported the significant burden of having to fight for services, co-ordinate and communicate between service providers. Parental difficulty acknowledging and envisaging their child's needs into the future was also identified.

## **Chapter 5: Health services to support children with spina bifida**

The complex needs of children with spina bifida results in their need to access both local and specialist services.

### **Local services**

Although there was timely initial access to teams, geographical disparities existed in relation to frequency of access to individual professionals. Children had good access to Physiotherapists, Occupational Therapists and Paediatricians, but limited involvement with Speech and Language Therapists, Social Workers, Family Support Workers, Psychologists, Dietitians and Orthotists. High demands on local services to provide equipment was identified as 83% of children already had equipment in place to support their needs. Orthoses, wheelchairs and standing frames were the equipment items most frequently utilised, requiring both trained professionals (Physiotherapist, Occupational Therapist and Orthotist) and funding for assessment and provision. The average wait-time for receipt of equipment was 6 months which caused frustration.

### **Specialist Services**

Only 46% of children had access to the MDT Spina Bifida clinic in TSCUH. Parents and service providers identified seven key professionals as most important to be present at the MDT Spina Bifida clinic: Neurosurgeon, Urologist, Orthopaedic Surgeon, Nurse Specialist, Paediatrician, Physiotherapist and Occupational Therapist. At the time of data collection, a Urologist, Orthopaedic Surgeon and Occupational Therapist were not available at the clinic. Not all children who had access to the MDT Spina Bifida clinic were reviewed annually, as recommended by international best practice, due to inadequate resources and staffing levels.

## Chapter 6: Key Recommendations

There is a comprehensive list of recommendations within this report, with the key recommendations outlined below.

Equity of access to professionals:

1. Parents and families require prompt access to information and specialist advice following the diagnosis of spina bifida.
2. A fully-staffed MDT spina bifida service should be available for inpatient care with an annual clinic review for all children with spina bifida in Ireland. The clinic team should consist of the following seven key professionals: Neurosurgeon, Urologist, Orthopaedic Surgeon, spina bifida Nurse Specialist, Paediatrician, Physiotherapist and Occupational Therapist. There should also be access to a Social Worker, Neuropsychologist, Speech and Language Therapist, Dietitian, Play Therapist, Neuro-ophthalmologist, Orthoptist and necessary imaging services.
3. Local MDTs should be available for children with spina bifida throughout childhood with an emphasis on early intervention. Local access to a Physiotherapist, Occupational Therapist, Speech and Language Therapist, Psychologist, Social Worker, Nurse, Orthotist, Dietitian, Paediatrician and Family Support Worker is required.
4. There should be increased frequency of provision of therapy services which is based on assessment of the individual child's needs.

**Improved psychosocial support:**

1. Family, peer and sibling support should be available antenatally, post-discharge from the tertiary hospital and on an ongoing basis to provide practical and emotional assistance.
2. All team members should be aware of and responsive to the child's psychosocial needs.

**Specific health and therapy needs should be met:**

1. A multidisciplinary approach to prevention and management of weight gain is necessary. Height, weight and BMI (centiles and Z scores) should be measured every 6 months by local professionals.
2. A comprehensive functional visual assessment should be available to all children with spina bifida who present with visual difficulties.
3. Trained nursing professionals should be available outside the national centre to assist with support and training of families and children with the objective of achieving social continence.
4. The process of prescribing, funding and providing equipment needs to be reviewed to enable timely provision that benefits the child and avoid unnecessary waste.

### **Inclusion of children in society and education:**

1. Mainstream education should be encouraged for children with spina bifida, provided this is appropriate to their learning needs. Access to mainstream school should not be influenced by physical or continence needs. Children may require Special Needs Assistants, Classroom Assistants, Resource Teachers or School Nurses to provide the necessary support.
2. Improvements in physical accessibility of the schools and community environments are necessary to increase independence and active participation in daily life.
3. Increased inclusion and engagement of children in activities such as adapted sports,
4. extra-curricular activities and social events is essential for their health and well-being.
5. There is a need for increased public awareness about pre-conceptual care and folic acid supplementation in Ireland.

### **Development of transition process to adulthood:**

1. Specific and directed transition planning should commence early in adolescence (between 12 and 14 years) as a priority and involve collaboration between the MDT spina bifida service, local services and adult services.
2. Co-ordinated spina bifida care within adult services requires development.

# SECTION ONE

# CHAPTER ONE

# BACKGROUND



## BACKGROUND

Spina bifida, is a type of neural tube defect (NTD), which has been described as the most complex congenital abnormality compatible with life which presents with a variety of complications (Liptak and El Samra, 2010, Liptak, 2003). NTDs consist of incomplete closure of the neural tube at any point along the formation of the spinal cord from the brain to the sacrum and occur in the first month of pregnancy (Northrup and Volcik, 2000). The most common forms of NTDs are anencephaly and spina bifida. Spina bifida meaning “split spine”, is also called spinal dysraphism and makes up a term used to myelomeningocele, meningocele, lipomyelomeningocele, and occulta (Fletcher and Brei, 2010).

Myelomeningocele is the most common form of spina bifida, it is also called open spina bifida or spina bifida aperta and results in an exposed area of the spinal cord and meninges protruding through a defect in the vertebral arch, most commonly in the lumbar region (Northrup and Volcik, 2000).

In the past, prior to antibiotic and surgical advances, life expectancy was often less than a year. Many children were not treated surgically, and those who had surgical repair often suffered complications such as infections and subsequent intellectual disabilities (Hunt, 1981, Liptak and El Samra, 2010). Improvements in the management of infants, children, adolescents and adults with spina bifida and hydrocephalus has meant that many are now not only surviving into adulthood, but live active and happy lives (Crosthwaite et al., 2001, Shin et al., 2012).



## Impact on health

The impact of spina bifida on the child and family can be substantial and complex. Multiple body systems are affected to some degree depending on the level of the lesion. This includes paralysis of the lower limbs below the level of lesion, anaesthesia of the skin as well as contractures or abnormalities of the lower limbs (Northrup and Volcik 2000). The pattern of muscle weakness and mobility level will vary with each individual, depending upon the site and extent of the spinal defect (Crosthwaite et al., 2001). Physical and orthopaedic complications depend on the level of the lesion, with common orthopaedic complications seen such as clubfoot, hip dysplasia, spinal deformities, and secondary flexion contractures due to immobility (Northrup and Volcik, 2000). Children can often present with difficulties with incontinence also.

Hydrocephalus and Chiari Type II malformations are common (Crosthwaite et al., 2001). Children with spina bifida require various neurosurgical procedures throughout their life including closure of their lesion and they frequently require the insertion of a cerebrospinal fluid (CSF) shunt for the management of hydrocephalus which can be present in up to 80% of cases (Northrup and Volcik, 2000). Chiari Type II Malformations of the hindbrain are common and treatment may be necessary if symptomatic. Complications such as syringomyelia, Chiari II Malformation or tethered spinal cord may also require surgery if they are symptomatic (Adzick et al., 2011).

Almost all individuals with spina bifida will have some degree of bladder and bowel dysfunction because the low sacral nerves innervate the distal bowel, anal sphincter, bladder and internal and external bladder sphincters (Shaer et al., 2007). Early urological evaluation and appropriate monitoring and interventions are essential to protect renal function and promote continence.

The neuropsychological impact includes difficulties with perceptual motor skills, numerical reasoning, attention, memory and organization (Northrup and Volcik, 2000). As a result, although most children with spina bifida have average intelligence, specific learning difficulties can make it challenging to obtain gainful employment and autonomy in daily living (Northrup and Volcik, 2000).

Complications secondary to spina bifida such as obesity, scoliosis, pressure sores, reduced independence and restricted participation in school and society can also have considerable impact on the child and family as described by Simeonsson et al. (2002). Other areas of impact which require consideration are sexual function, precocious puberty and latex allergy (Bowman et al., 2001, Merkens, 2006, Verhoef et al., 2005b). These factors all need to be considered when delivering health and therapy care to these children.

## Prevalence

Ireland has one of the highest rates of NTDs in the world with an incidence of 1.17 per 1000 (McDonnell et al., 2014), about half of whom present with spina bifida. Northrup and Volcik (2000) report a variation in incidence by ethnicity with the Celtic nations specified as having high incidence compared to North America. Although no central database exists for incidence of children with spina bifida in Ireland, calculations based on the European Surveillance of Congenital Anomalies (EUROCAT, 1991) data, Temple Street Children's University Hospital (TSCUH) and Spina Bifida Hydrocephalus Ireland (SBHI) reported numbers, and incidence and birth rates would suggest that there are approximately 500 children (0-18 years) with spina bifida currently in Ireland. Reasons cited for high rates of spina bifida in Ireland include a genetic predisposition, a history of low peri-conceptual folic acid intake (Food Safety Authority of Ireland, 2008) and the fact that termination of pregnancy for foetal anomaly (TOPFA) is not legal (McDonnell et al., 2014).

The availability of prenatal diagnosis and the rates of elective termination of pregnancy can affect the live birth rate (Shaer et al., 2007, Bowman and McLone, 2010). In Wales, Czapran et al. (2012) reported that 83% of pregnancies with an antenatal diagnosis of spina bifida were terminated in 1998. However, they noted that the termination rate was 62% ten years later in 2008. This trend may suggest that prospective parents are now more willing to proceed with a pregnancy due to better support and services available for them and their children. Improved prognosis and more specific antenatal counselling can also give a more accurate prediction of outcomes. Lorber (1972) noted that in Sheffield, UK, nearly double the number of babies born with NTD would have been born between 1974 and 1984 if it were not for the availability of TOPFA. The lack of TOPFA in Ireland contributes to the high incidence in comparison with other countries (McDonnell et al., 2014). A proportion of NTD affected pregnancies in Ireland are terminated abroad, while this information is not readily available to the EUROCAT Registry, it is estimated to be at a rate of 20% (EUROCAT 2011).

## Role of Folic Acid

Considerable evidence exists to recommend that women of childbearing age who could become pregnant should take 400 micrograms ( $\mu\text{g}$ ) (0.4 mg) of folic acid a day for at least 3 months before conception to prevent NTDs (MRC, 1991, Shaer et al., 2007). A Randomised Control Trial in Hungary identified statistically significant results suggesting periconceptual folic acid intake reduced the first occurrence of NTDs (Czeizel and Dudas, 1992) with other studies showing a reduction in recurrence of NTDs by between 71 and 91% (Czeizel, 2000). For women with a previous family history of a pregnancy affected by a NTD a randomized double-blind prevention trial showed that a higher dose of 4000  $\mu\text{g}$  of folic acid daily (10 times the general recommendation) as well as supplementing their diet with folate rich food reduced their recurrence risk by 70% across 7 countries (MRC, 1991). The study established that 29% of NTDs were not prevented by folic acid (MRC, 1991) suggesting other factors at play

including genetics.

Folate is available from green vegetables, fruits and juices. In Ireland there is a system of voluntary food fortification which means that under the Flash Labelling Scheme manufacturers can choose to supplement products with extra folic acid. This means that folic acid can sometimes be present in breakfast cereals, milks, yogurts however these are recommended to only include one sixth or one half Recommended Dietary Allowance (RDA) (Food Safety Authority of Ireland, 2008). Natural folate is less easily absorbed into the body than folic acid taken as supplements or in fortified foods therefore most people do not get enough folic acid through food alone. The North/South Ireland Food Consumption Survey found that over a third (35%) of women of child bearing age in Ireland report that they consume no folic acid at all (Irish Universities Nutrition Alliance., 2001). In 1993 in Ireland, the Department of Health and Children advised that all women of childbearing should age take an additional 400µg of vitamin folate daily as a supplement, prior to conception and during the first 12 weeks of pregnancy by eating more natural food folate, by eating foods fortified with folic acid, or by taking folic acid supplements or a combination of these. This advice has had little impact on reducing the incidence of NTDs in Ireland which is partially impacted by the fact that less than 50% of pregnancies are planned resulting in women learning they are pregnant without having had the opportunity to follow recommended advice for planning a pregnancy (FSAI, 2006). Many countries have introduced mandatory folic acid food fortification, the impacts and consequences of this will be discussed further in the 'folic acid and prevention section' in chapter 4.2.

Additional risk factors for NTDs have been cited such as maternal medication, pre-pregnancy obesity and diabetes, previous pregnancy with spina bifida or a positive family history and genetic predisposition (Kennedy et al., 1998, Lynch, 2005, Chen, 2005).

## **Health Care Services**

The health care needs of children with spina bifida are complex. They need specialists, generalists, and integrated systems to deliver this complex care (Liptak and El Samra, 2010). Children with spina bifida in Ireland may be accessing services from various settings including tertiary centres, local hospitals, and community teams or through individual disciplines to support their health and therapy needs. These services should be readily available, on-going, comprehensive and coordinated throughout the lifespan to ensure children with spina bifida can access and actively participate in their environment (Merkens, 2006). The establishment and development of these services are described below.

## **Local Services**

In Ireland, Primary Care services play a central role in the provision of health care (McDaid 2009). The Primary Health Care Strategy was launched in 2001 by the Department of Health and Children (DoHC., 2001) stating that 'Primary care should be readily available to all people regardless of who they are, where they live, or what health and social problems they may have'. Following from this strategy was the recommendation for the development of multidisciplinary community health care teams which should include an essential skill mix including assessment, diagnosis, prevention, and rehabilitation as well as access to occupational therapy, physiotherapy, nursing, midwifery, home help, health education, counselling, administration, management and social services. Early intervention was seen as a priority (DoHC., 2001).

Early intervention teams (EITs) provide services for children in their preschool years, School age teams (SATs) provide services for children with physical disability through the school ages until 18 years. Teams may be located within the HSE services, or through joint initiatives pooling resources from a variety of organisations (for example Enable Ireland, CRC, Brothers / Daughters of charity, St John of God's Carmona Services, voluntary organisations). The local health services can be staffed and funded by a variety of organisations to deliver services to children with spina bifida in Ireland. These services might include:

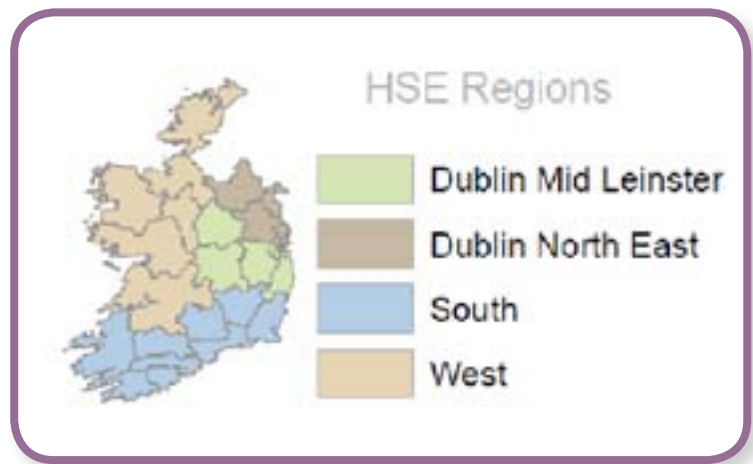
The Health Service Executive (HSE) provides a range of services for people with intellectual, physical and sensory disabilities or autism. These services include basic health services as well as assessment, rehabilitation, income maintenance, community care and residential care. Some services are provided directly by the HSE. Many of the community, residential and rehabilitative training services are provided by voluntary organisations with grant aid from the HSE. They may access community care services such as: Public Health Nurses, Home Helps, Personal Assistance, Psychological Services, Speech and Language Therapy, Occupational Therapy, Social Work Services, Physiotherapy, Day care and Respite Care ([www.hse.ie](http://www.hse.ie))

Enable Ireland provide a service to children with disabilities covering all aspects of a child's physical, educational, and social development from early infancy through adolescence. They cover 40 locations in 14 counties ([www.enableireland.ie](http://www.enableireland.ie))

The Central Remedial Clinic (CRC) carries out a wide range of services, carrying out assessment, diagnosis and treatment of children with a wide range of physical conditions to different parts of Ireland, as well as locally in Dublin ([www.crc.ie](http://www.crc.ie))

The Prospectus Report (DoHC., 2003) realigned the reconfiguration of the health boards into four administrative units under the control of the Health Service Executive (Harvey, 2007). These four units or geographical areas are named as HSE Dublin Mid Leinster, HSE Dublin

North East, HSE West, HSE South. In analysis within this report these units are used to group and compare regional access to services. This organisational framework is currently under review with a possible move toward clinical directorate. Variations in access to services exist across Ireland (National Disability Authority, 2011). Previous reports suggest regional and county variations in referral and access to



services in Ireland (Carroll et al., 2013). To address the ad hoc development of children's disability services, the HSE highlighted the need to reconfigure services for children with disabilities (HSE., 2010) and initiated a national programme "Progressing Disability Services for Children and Young People" in joint partnership with nongovernmental agencies, the DoHC and the Department of Education and Skills. This programme, which was launched in 2011, aims to implement the new structures for disability services within the context of the primary care model. This process was underway during the timeframe of this report. Following the reconfiguration of services in line with progressing disability services, access is dependent on the geographic area, the age of the child, and types of disabilities or developmental concerns. Therefore services which were previously unique to physical disability will now include children with a range of developmental needs and therapists will be reallocated between services.

## ***Specialist Services***

In 2005 the "Report of the committee to review neurosurgical services in Ireland" examined existing neurosurgical service provision nationally and concluded that there was compelling evidence that neurosurgical services in Ireland were insufficient to meet the needs of the population and required significant expansion for development and staffing for paediatric neurosurgery in Dublin (Committee to Review Neurosurgical Services, 2005). Following this and the Horwath report (Horwath Consulting Ireland, 2008), Temple Street Children's University Hospital (TSCUH) became the national neurosurgical centre and tertiary care centre for children with spina bifida and hydrocephalus born in the Republic of Ireland after the 1st of September 2008. The Spina Bifida MDT, however, was only partially resourced with allocation of staffing and funding being largely subsumed by the broader Neurosurgery needs rather than the specific needs of the children with spina bifida. TSCUH shares care with local services as recommended by the NHS (2011). Spina Bifida multidisciplinary clinics have been repeatedly cited in the literature by as the best method of specialist service delivery for these children (Brei, 2007, Brustrom et al., 2012, Delmarva Foundation, 2006, Aldana et al., 2010). A multidisciplinary Spina Bifida clinic was established in TSCUH in 2011 to cater for the specialist care needs of these children.

Due to the high incidence of NTDs in Ireland and the fact that all children born with NTDs since 2008 now attend TSCUH, this centre has one of the largest caseloads of children with NTDs in Europe resulting in a unique opportunity to gather data regarding this complex condition.

## **Participation and Engagement in Their World**

Similar to many children with disabilities, active participation and engagement in life is the desired outcome for children with spina bifida. The World Health Organisation's International Classification of Function Disability and Health (ICF) aims for participation of an individual in life situations with an emphasis on capacity and performance (World Health Organisation, 2001). Using the ICF model to describe spina bifida highlights areas of concern related to body function and structures (e.g., paraplegia, upper extremity function, and learning disability), activities (e.g., mobility, self-care, and performance of school-related tasks), and participation (e.g., social activities, work, and independent living).

In the past, most of the focus on outcomes from interventions was on body function and structure rather than on activities and participation, which is now being seen as a priority (Kinsman et al., 2000). Participation in everyday life and out of school activities are recognized as critical to children feeling confident, establishing meaningful relationships, and attaining life satisfaction (Rosenbaum and Stewart, 2004, Law et al., 2006). However, King et al. (2013) reported that children with disabilities participate in significantly fewer out of school activities than children without disabilities, therefore highlighting the need for this engagement in activity to be fully inclusive for the child with disabilities. As well as out-of-school activities, engagement in appropriate school placement and in school activities which are positive for children can contribute to better psychological wellbeing and functioning (Essner and Holmbeck, 2010).

Having a child with spina bifida can have a significant impact on families due to the complexity of the condition, the need for various medical procedures and the chronic impact of mobility and continence issues (Vermaes et al., 2008). Parental stress, sibling anxiety, financial and work implications and psychological distress and restricted social lives can present as issues with families and psychological assistance and support is required (Adzick et al., 2011, Vermaes et al., 2008)

## **Reviews, Guidelines and Standards of Care**

Reviews, Guidelines and Standards of Care exist for some aspects of spina bifida care such as neurosurgical specialist needs, Specialist MDT Clinics, health care services and non-surgical issues which are detailed below:



The Children's Neurosurgical Specification Standards set out Areas of care, Principles and Best Practice for specialist neurosurgical service delivery for Spinal Neural Tube Defects. They stipulate that there must be access to 24/7 neurosurgical advice, urgent scanning and imaging, ongoing specialist MDT review, shared care and good communication and coordination with local teams (NHS., 2011).

In 2006, the Delmarva Foundation (2006), assisted by the Spina Bifida Association of America (SBA), the Centers for Disease Control and Prevention (CDC), the Agency for Healthcare Research and Quality (AHRQ) and members of the spina bifida community surveyed Spina Bifida clinics throughout the United States (US). This review investigated staffing, patient load, funding, strengths, perceived needs and involvement in clinical research and quality improvement.

Results from the Delmarva national review suggested that Spina Bifida clinics in the US range in size, availability and the number of patients seen during a clinic (between 4 and 60 patients per clinic). Most children are reviewed at least once a year, and clinics vary in length from 2.5 hours to 12 hours. Although there is variation in the specialists available at spina bifida MDT clinics, the survey found that more than half of the clinics in the US included a Urologist (91%), Orthopaedic Surgeon (87%), Physiotherapist (85%), Social worker (79%), Neurosurgeon (72%), Clinical nurse specialist (69%), Occupational therapist (66%), Paediatrician (66%) and Dietitian/nutritionist (65%) with 87% of clinics also having a dedicated care coordinator. Various assessments and interventions were provided through MDT clinics including prenatal care planning (62%), coordination with community services (99%) and coordination with schools and teachers (91%). About half of the Spina Bifida clinics were involved with research and 60% in ongoing quality improvement activities. The report highlighted the need for MDT Spina Bifida clinics and the need to provide coordinated care for children and adults with spina bifida. (Delmarva Foundation, 2006).

The Spina Bifida Association of America (SBA) established "Guidelines for Spina Bifida Health Care Services throughout lifespan" (Merkens 2006) following a conference on evidence-based practice in Washington D.C (Liptak 2004). The guidelines aimed to provide practitioners with recommended directions for treatments and interventions for patients with spina bifida based on the best available research findings at that time. This document is laid out in chronological order and highlights priority outcomes, principles and interventions for each stage as detailed in the table below Merkens (2006).





Table 1

“Guidelines for Spina Bifida Health Care Services throughout lifespan”		
Merken's (2006) - Priority Outcomes		
PRIORITY OUTCOMES		APPLIES FROM THIS AGE THROUGHOUT LIFESPAN
Prenatal	<ul style="list-style-type: none"> <li>• Receive accurate information about spina bifida &amp; range of outcomes, respect autonomy</li> <li>• Delivery method chosen and families happy</li> <li>• Baby referred to a MDT in a NICU</li> </ul>	
Newborn	<ul style="list-style-type: none"> <li>• Closure of open spine</li> <li>• Control hydrocephalus</li> <li>• Baseline studies of neurosurgery orthopaedic &amp; urology needs</li> <li>• Referral to spina bifida MDT</li> </ul>	<ul style="list-style-type: none"> <li>• Parents and siblings feel supported during this challenging time</li> <li>• Latex precautions followed</li> <li>• Receive on-going, comprehensive, coordinated care throughout the lifespan</li> </ul>

<p>Infant &lt;1 year</p>	<ul style="list-style-type: none"> <li>• Attention to bowel function &amp; continence</li> <li>• Enrolled in Early Intervention Team (EIT)</li> <li>• Identify cryptorchidism &amp; testicular torsion</li> </ul>	<ul style="list-style-type: none"> <li>• Parents/siblings receive adequate information and support</li> <li>• Normal intracranial pressure and CNS functioning maintained</li> <li>• Optimal cognitive function preserved</li> <li>• Bone and muscle maintained in functional alignment</li> <li>• Kidney function and drainage in normal range</li> <li>• UTIs controlled</li> <li>• Weight is appropriate for length/height, primary care team is providing care</li> <li>• Genetic counselling and information about folic acid for parents</li> <li>• Parents using latex precautions</li> <li>• Has all routine immunizations</li> </ul>
<p>Toddler 1-3 years</p>	<ul style="list-style-type: none"> <li>• Bowel continence program continued</li> <li>• Enrolled in EIT</li> </ul>	<ul style="list-style-type: none"> <li>• Optimal mobility is maintained</li> <li>• Family aware of folic acid</li> </ul>
<p>Preschool 3-5 years</p>	<ul style="list-style-type: none"> <li>• Wheelchair obtained for mobility &amp; transport if appropriate</li> <li>• Enrolled in preschool program</li> </ul>	<ul style="list-style-type: none"> <li>• Bowel &amp; urine programs establish social continence</li> <li>• Latex precautions used by all</li> <li>• Overall development is progressing</li> <li>• No deterioration of skills evident (cord tethering)</li> </ul>
<p>School age</p>	<ul style="list-style-type: none"> <li>• Increase independence in self-care (bladder/bowel skin)</li> <li>• Child aware of latex precautions</li> <li>• Onset of puberty at optimal age sought &amp; precocious puberty in girls identified &amp; treated</li> <li>• Prevention of sexual abuse</li> </ul>	<ul style="list-style-type: none"> <li>• Bowel &amp; urine continence well established</li> <li>• Prevention of secondary disability due to tethered cord</li> <li>• Has friends &amp; engages in recreational activities</li> <li>• Regular exercise programme</li> <li>• In appropriate school with full inclusion in class &amp; extra-curricular</li> </ul>

Adolescent		<ul style="list-style-type: none"> <li>• Independent in all self-care activities</li> <li>• Receives adequate education &amp; vocational service &amp; information/supportive services</li> <li>• Participates in adaptive drivers education course</li> <li>• Understands folic acid supplementation</li> <li>• Aware of sexual issues</li> <li>• Prevention of sexual misuse/abuse expands</li> <li>• Knowledgeable about spina bifida management &amp; prevention of complications</li> </ul>
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Adapted from (Merkens, 2006)

\*MDT = Multidisciplinary Team, NICU = Neonatal Intensive Care Unit

Lipak and El Samra (2010) performed a comprehensive literature review of spina bifida health care delivery. The article reviewed the history of spina bifida care, healthcare delivery, research progression and the main medical issues. Discussion included multidisciplinary clinics which remain the model for healthcare delivery for children with spina bifida and the overall goal to maintain or improve health and wellbeing through cost-effective solutions. However methods to achieve these goals remained uncertain and further research and review was recommended (Liptak and El Samra, 2010).

## Aim of the report

Children with spina bifida require appropriate support by both local and specialist teams and services to manage their complex health and therapy needs. Some standards of care have been established in both the United Kingdom and the United States to guide aspects of care delivery for these children. These will be referenced throughout the document as examples of evidence-based guidelines for people with spina bifida as well as other literature relating to spina bifida.

The aim of this report is to determine the health, therapy and service needs of children with spina bifida (0-18 years) from the perspectives of parents and service providers, to compare these to international practice and to help guide development of recommendations for service development.

# **CHAPTER TWO**

# **METHODOLOGY**

## **Methodology**

This chapter aims to describe the research methodology of this study to explain the sample selection, to describe the procedure used in designing the instrument and collecting the data and finally to provide an explanation of the statistical procedures used to analyse the data.

The overall aim of this study was to identify current health needs of children aged 0-18 years with spina bifida in Ireland. Specific objectives of this study were:

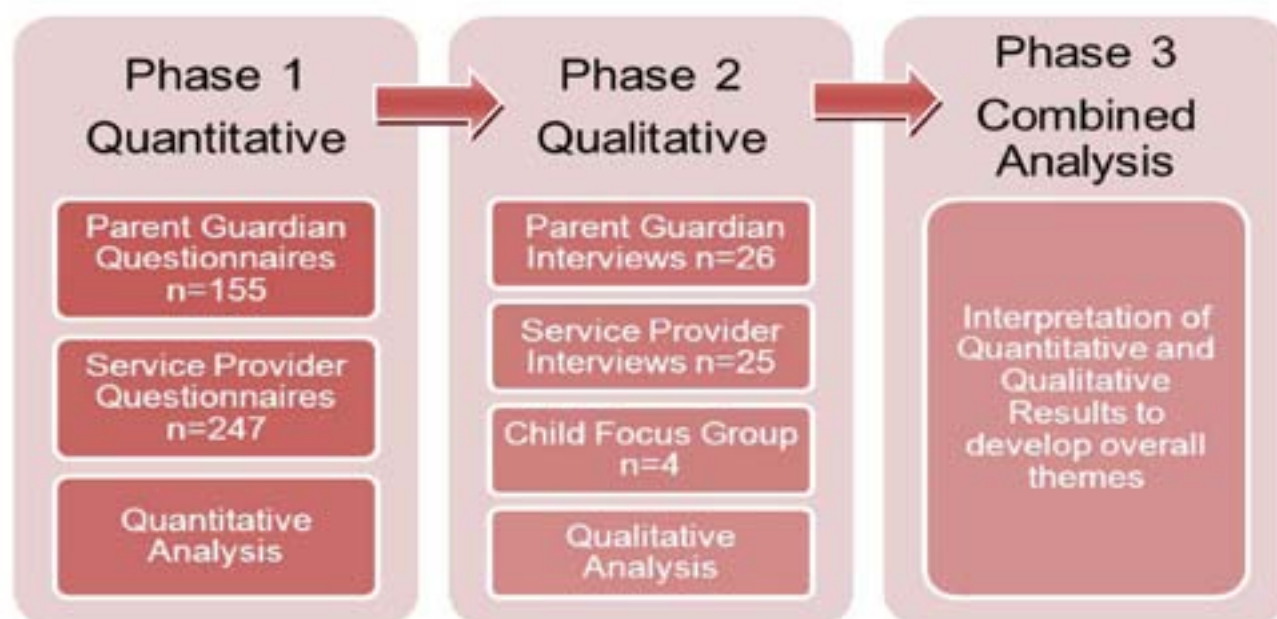
1. To identify the availability of current services for children with spina bifida from the perspectives of families and service providers nationally.
2. To gather the recommendations of service providers, expectations of parents / guardians, opinions of children with spina bifida regarding health and therapy needs.
3. To highlight the impact of spina bifida on the quality of life of children and their families in order to develop recommendations to support client centred practice.
4. To provide an up to date literature review of international best practice in relation to the health and therapy needs of children with spina bifida.
5. To gather feedback regarding the MDT Spina Bifida clinic in TSCUH and how this service compares with other international centres.

The main research question was: 'What are the current health and therapy services available for children with spina bifida in Ireland and how does this compare with international best practice and the needs identified by service providers and parents / guardians and the children themselves?'

## **Research Methodology**

A mixed method sequential explanatory design was used for this study which consists of three phases, collecting data from three different population groups (service providers, parents / guardians, children 8-18 years) (Figure 1). Mixed methods research aims to "consider multiple viewpoints, perspectives, positions and standpoints" in order to synthesize qualitative and quantitative findings (Johnson, Onwuegbuzie, & Turner, 2007, p. 113). Using mixed-methods can assist in gathering a holistic picture of the influences and processes children encounter within a cultural setting (Harkness et al., 2006). The research followed the pragmatism paradigm which Creswell and Piano-Clark (2007) described as being appropriate in mixed-methods research due to its focus on the research question and real-world problem centred approach.

Figure 1.  
Methodology



In the first phase purposely-designed quantitative questionnaires were completed by service providers and parents / guardians (See Questionnaires in Appendix 2). The second phase consisted of one to one interviews with service providers and parents / guardians to further explore results derived from analysing quantitative data. Those unable to attend interview were given the opportunity to complete telephone interviews. Interviews are a rich method of data collection, which are discovery-oriented, responsive to the phenomena and reflexive (Finlay, 1999). Concurrently, a small focus group was conducted, and quality of life questionnaires completed with children aged 8-18 years to gather their own perspectives of service needs. As gathering information from children themselves can provide a clearer understanding of their perspectives eliciting the child's voice is supported in research (Ashton, 2008; Curtin, 2001, Morrow & Richards, 1996).

### **1. Sampling:**

A purposive sampling method was employed to gather information from the three population groups (service providers, parents / guardians, children 8-18 years) using gatekeepers. Purposive samples aim to be representative and participants are deliberately chosen (Sim & Wright, 2000).

#### **Service Providers:**

The service providers included all clinical or professional staff working with children with spina bifida in the following services: Enable Ireland, Central Remedial Clinic (CRC), Health Service Executive (HSE) teams (early intervention teams [EIT], school age team [SAT], and primary care teams [PCT]), and charitable organisations e.g. SBHI. A gatekeeper was identified by phone

for each for these teams who agreed to forward an email containing information leaflets with a link to an online questionnaire to the remainder of the team members. An estimated population of 1000 service providers was calculated based on fully-staffed teams.

Responses to online questionnaires were received from 247 service providers which represents a response rate of 25%.

Volunteers for follow-up interviews were sought at the time of questionnaire completion and with reminders closer to the time. Interested volunteers were sent further information leaflets and consent forms to be completed and suitable interview times were identified. Twenty five service providers completed follow up interviews following a semi-structured format which were developed following initial questionnaire analysis.

### **Parent / Guardians:**

Parents / guardians of children with spina bifida aged 0-18 years were contacted through gatekeepers. Online questionnaires were forwarded by the Spina Bifida Hydrocephalus Ireland (SBHI) administration through their databases, placed on their Facebook page and on their website. Disability Paediatricians were contacted nationally and paper questionnaires were sent for onward posting by their administration staff. Paper questionnaires were also sent from the administrator for the TSCUH Spina Bifida MDT Clinic Database. Information posters with a link to the online questionnaire were distributed to consenting healthcare facilities.

An estimated prevalence of children with spina bifida in Ireland is between 2.3 – 4.45 per 10,000 live births (Eurocat data 2013, TSCUH database). This would indicate an incidence of approximately 1550 adults and children nationally based on current population numbers. We estimated that 30% of these are under 18 years, which results in a figure of 500 children with spina bifida nationally. This calculation was in line with the SBHI database, which estimates 500 families under the age of 18.

Responses to either online or postal questionnaires were received from 155 parents / guardians, which represents a response rate of 32% from the overall population.

Volunteers were sought for follow-up interview at the time of questionnaire completion. Information leaflets on the interview process and consent forms were included with the postal questionnaires. Volunteers who returned completed consent forms were contacted and a suitable interview time was identified. Twenty six parent / guardians completed follow up interviews following a semi-structured guide (See Appendix 4) which were developed following initial questionnaire analysis.

## **Children 8-18 years:**

The final population group of children with spina bifida (8-18 years) were also accessed through the various gatekeepers (SBHI, Disability Paediatricians, TSCUH spina bifida MDT Clinic). Parental consent and child assent was required for participation. Child Focus Group Information leaflets, child assent forms and parental consent forms were included with the postal questionnaires. The focus group was held at the annual Spina Bifida and Hydrocephalus Ireland Conference where parents and children were in attendance. One focus group was held with 4 participants aged 8-16 years. An adolescent focus group was not completed due to lack of response from this population. Curtis, Roberts, Copperman, Downie, & Liabo (2004, p. 168) noted that children with disabilities can be “hard to reach” for research and are therefore less well represented in research due to difficulties with access. The participants of this study consisted of a hard to reach population as according to McDonnell et al (2014) there is no complete Irish database available regarding children with spina bifida.

### **Inclusion criteria:**

A service provider for children with spina bifida within Enable Ireland, Central Remedial Clinics (CRC), Health Service Executive (HSE) teams (early intervention teams [EIT], school age team [SAT], and primary care teams [PCT]), charitable organizations. A parent of a child / children (0-18 years) with spina bifida A child aged 8-18 years with spina bifida with parental consent

### **Exclusion criteria:**

Neural Tube Defects not included in the spina bifida description (i.e. Anencephaly). Isolated congenital hydrocephalus Children with spina bifida under the age of 8 years were excluded from the focus groups due to difficulties with insight into impacts on quality of life and limited ability to engage in a focus group.

## ***2. Instrument Design and Data Collection***

### **Phase one Data Collection: Questionnaires**

As no existing measure which captured the extent of data required by this study was identified by an initial literature search, two structured self-developed questionnaires were specifically designed for this study.



## **Parent Guardian Questionnaire**

A questionnaire was devised into 3 sections:

- **Demographics:** this captured information on parent / guardian relationship, child's age, gender, ethnicity, family structure and employment status, geographical location, and educational setting.
- **Medical Information:** this section gathered information relating to family history, level of lesion, hydrocephalus and related surgeries, level of mobility, equipment including provision and wait time, height and weight, pressure sores, visual difficulties, bladder and bowel difficulties, orthopaedic difficulties and surgeries, cognitive ability
- **Therapy information:** this section gathered information about the therapeutic interventions, access to professionals and frequency of review, access to Spina Bifida clinic and specialists, satisfaction. (See appendix 1 for copy of Parent/Guardian Questionnaire)

## **Service Provider Questionnaire:**

A questionnaire was devised, and consisted of the following three sections:

- **Role and Caseload:** this section gathered details of profession, service type and area, years of experience, team structure, number of children on active caseload, wait times for access of service.
- **Assessment and Interventions:** this section investigated the assessment and intervention practices currently in place nationally, in order to establish where gaps in service may be evident. Questions asked about the frequency and location of review, involvement and timing of equipment assessment, range of available interventions provided, specific interventions provided for bladder and bowel management, pressure relief, education, height and weight monitoring practices, prevalence of social, emotional or psychological concerns, speech and language concerns, and visual difficulties.
- **Transition, shared care and Service Development:** this section investigated the transition for children with spina bifida into adulthood and some of the service involved in this process. It also examined the shared care model between the national Spina Bifida clinic and the local community services. (See appendix 2 for copy of Service Provider Questionnaire)

Questionnaires were sent to colleagues for proofing and piloting to ensure ease of use and appropriateness. Closed ended dichotomous questions and Likert scales were used to gather quantitative data. Qualitative comments were also gathered. All questionnaires

were anonymous, confidentiality was ensured, and paper questionnaires were stored in a locked cabinet for the duration of the study. Return of the completed questionnaire was deemed consent to engage in this part of the study. An online or postal questionnaire was chosen for phase one of data collection due to its cost efficiency, speed of administration and convenience.

## **Phase Two Data Collection:**

### **Interviews**

Initial quantitative analysis was completed on both the service provider and parent / guardian questionnaires on Survey Monkey. Frequencies and means were calculated and qualitative comments reviewed. This informed the development of a semi-structured interview guides (See Appendix 4) for the service providers and parent guardians using open ended, non-threatening questions. The semi-structured interview guide approach allows the researcher to remain in the “driver’s seat” yet still maintain flexibility based on participant prompts (Turner, 2010, p. 755).

#### **Service Provider Interview Guide:**

This aimed to gather a range of professionals’ opinions regarding their role, the accessibility of their services, whether they felt their service met the needs of children with spina bifida, their involvement in and the types of interventions provided, and finally to gather recommendations which might help to improve services in the future. 25 service providers volunteered for interview. (See appendix 4 for copy of service provider interview guide).

#### **Parent Guardian Interview Guide:**

This aimed to gather parents’ experiences from diagnosis, initial hospital experience, current services accessed, how spina bifida had impacted on their child and their family. We also explored how they felt these experiences could have been improved upon and recommendations were gathered for the future development of the service. 26 parent/guardians volunteered for interview. (See appendix 4 for copy of parent/guardian interview guide).

Following completion of consent forms, Interviews were completed one to one at a convenient location. Telephone interviews were arranged when face to face interview was not possible to arrange.

Interviews were transcribed verbatim by the researchers and by an outsourced agency. All transcriptions were proof-read by researchers for errors.

## **Child Focus Groups:**

Following completion of parental consent and child assent forms a Focus Group was held at the SBHI AGM which was attended by both parents and children. For research with children to be seen as ethical, parental consent is often a key criterion (Morrow & Richards, 1996). This focus group followed a semi structured format aiming to gather the children's perspectives and experiences of their life with spina bifida. A quality of life questionnaire was completed at the beginning of the session to introduce topics for discussion. This questionnaire was not analysed quantitatively due to the limited number of participants. The quality of life questionnaire selected was the KINDL (Revens-Sieberer et al., 2001). This standardized questionnaire has demonstrated validity and reliability in measuring health related Quality of Life for Children, including those with spina bifida. (See appendix 5 for copy of focus group guide and appendix 6 for copy of KINDL measure).

## **3.Data Analysis**

Quantitative data gathered from questionnaires was inputted through Survey Monkey and exported to Microsoft Excel. All information was coded, anonymized and further exported to SPSS. Data was analysed using descriptive statistics including frequency distributions, measures of central tendency and bivariate relationships using SPSS.

Qualitative data gathered from interviews and focus groups was recorded via tape recorder and transcribed verbatim for analysis. These transcripts were inputted into Weft QDA for coding. To ensure rigour an audit trail was established by documenting methods, field notes, interview-guide and reflections. Dickie (2003) found that field notes often constitute an early step of analysis. These were written shortly after the interviews and included observations of both participants and the interviewer. Finlay (2002, p. 532) defined reflexivity as "thoughtful, conscious self-awareness" which is necessary for the trustworthiness of qualitative research as a way to analyse one's own personal influence.

Once coding was completed, overall themes were identified as described by (Braun and Clarke 2006). Thematic analysis has been described as a flexible and accessible approach which is often poorly demarcated or acknowledged (Braun and Clarke, 2006). Overall themes should capture recurrent patterns in the data (Merriam, 2009), therefore the 4 major themes were identified highlighting parents and service providers perspectives in relation to:

1. Impact of spina bifida on the child,
2. The impact of spina bifida on the family
3. The need for local services
4. The need for specialist services to support children with spina bifida.

## **Conclusion**

This research aimed to gather an up-to-date representation of international best practice in relation to the management of spina bifida, information in relation to current services available in Ireland, as well as recommendations on how to further develop these services. Data analysis and discussions were then collated to develop recommendations and dissemination of these findings at national and international conferences as well as to policy makers and service managers will continue to be completed.

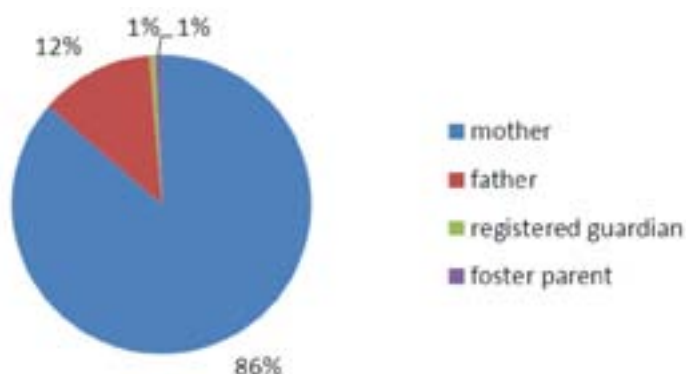
# CHAPTER THREE

## SAMPLES

### 3.1 Parent / Guardian Sample

Figure 2

#### Parent / Guardian Respondents



Respondents consisted of 155 parent / guardians who completed postal or online questionnaires completing information in relation to their child's demographics, medical and therapy service provision and recommendations for the development of services.

Eighty six per cent of respondents were mothers, 12% were fathers, with 1 registered guardian and 1 foster parent (Figure 2).

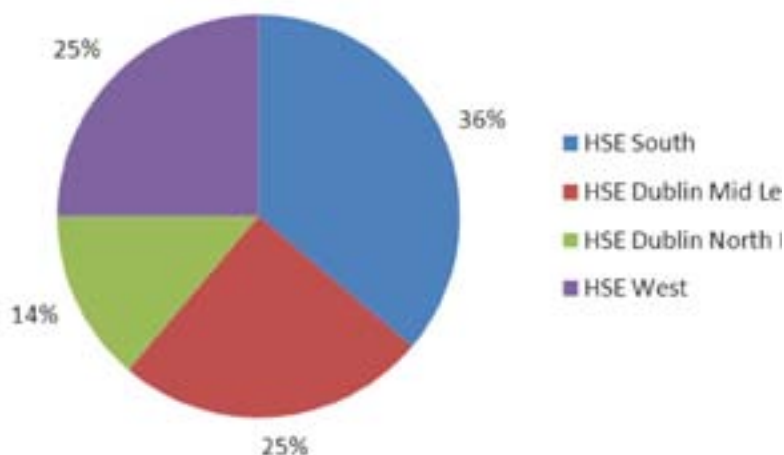
The mean age of the children was 5.7 years (SD 4.7) with a range from 2 months to 18 years. Although sampling methods aimed to capture all children in the 0-18 year age range, a greater number of young children were represented. 54% of the children represented were female and 46% male. The male to female ratio was 1:1.17.

147 (96%) respondents were from a white ethnic background, 3 respondents were black ethnicity, 1 reported to be Asian, 1 eastern European and 1 from the Travelling community.

The parents represented a national spread of children across the four HSE regions (Figure 3) with a slightly greater representation from the HSE South (36%) and slightly lower from Dublin North East (14%). 26 parents took part in qualitative interviews with researchers.

Figure 3

#### Parent / Guardian Address



### 3.2 Service Provider Sample

247 service providers responded to the online questionnaire which provided information about their role and caseload, assessments and interventions completed and recommendations and information about transition, shared care and service development.

A variety of service providers were sought through multidisciplinary teams nationally and the spread of professionals are presented below (Figure 4).

Figure 4

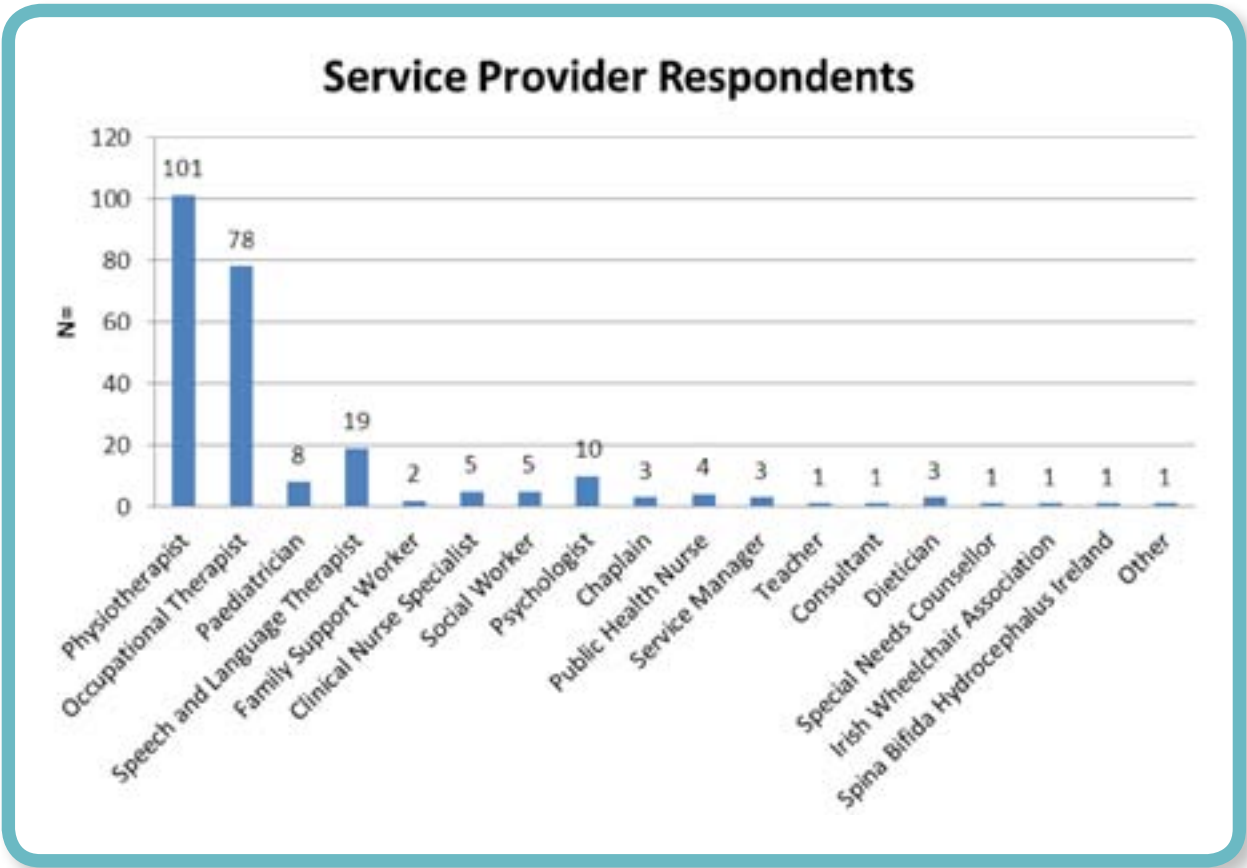
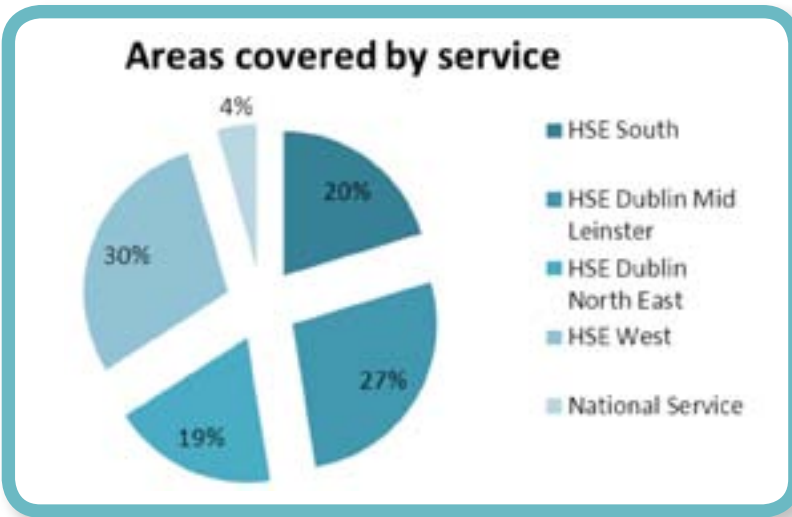


Figure 5



41% of service providers were physiotherapists with 32% being occupational therapists. Speech and language therapists represented 8%, psychologists 4%, and paediatricians 3%. Ninety per cent of respondents were female and 10% male. The total sample had a mean of 8.51 years of experience (SD 5.992, range 0-35 years) with children with spina bifida.

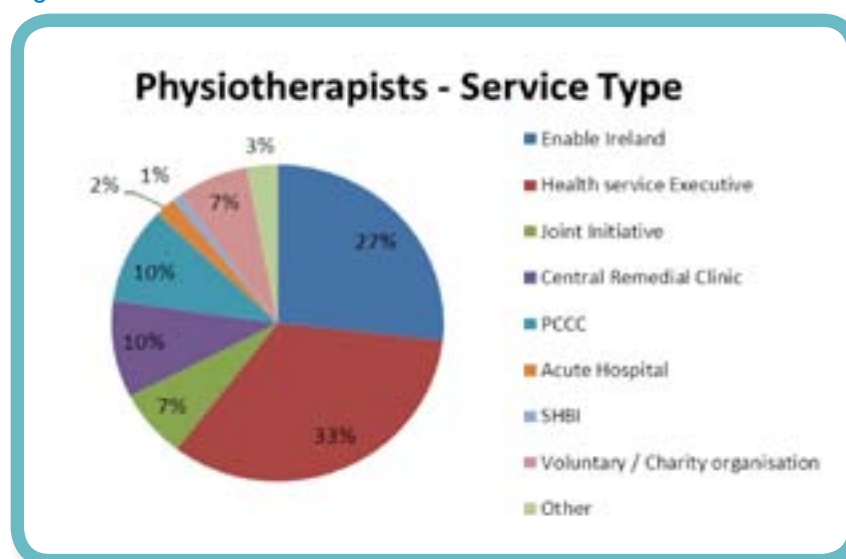
A national service was provided by 4% of the service providers with 30% of service providers reported that their service covered the HSE Dublin Mid Leinster region, 27% covering HSE Dublin North East, 20% HSE South and 19% HSE West (Figure 5)

Therapists locally do not have many children with spina bifida on their active caseloads. With individual professionals reporting between 2 and 7 children on their active open caseloads on average. Out of the whole sample, 85% respondents reported that they provide some kind of clinical input to children with spina bifida (6% were involved in the prescription of equipment only and 9% were not involved as the services were provided elsewhere).

As a large proportion of respondents were Physiotherapists, Occupational Therapists, Speech and Language therapists, Paediatricians and Psychologists, and data was analysed and reported individually from the perspective of professionals most frequently involved with these children these samples are detailed below. The large proportion of Physiotherapy and Occupational Therapy respondents may have been influenced by the fact that the two principal authors who distributed questionnaires and completed interviews were from these professions.

## Physiotherapists

Figure 6



One hundred and one physiotherapists responded; 89.1% were female (n=90) and 10.9% were male (n=11). The services in which they worked are presented in the chart below. A third of the Physiotherapists are employed by the HSE, over a quarter by Enable Ireland Services. Ten per cent of physiotherapists were employed by CRC, a further

10% by the Primary, Community and Continuing Care (PCCC). Other employers were acute hospitals, Spina Bifida Hydrocephalus Ireland and Voluntary or Charitable organisations

The largest proportion of physiotherapists worked in early intervention community teams (40%), followed by school age teams (19%) and teams providing services to children from 0-18 years (18%). Others worked in CRC, acute hospitals, and voluntary and charitable organisations.



Figure 7

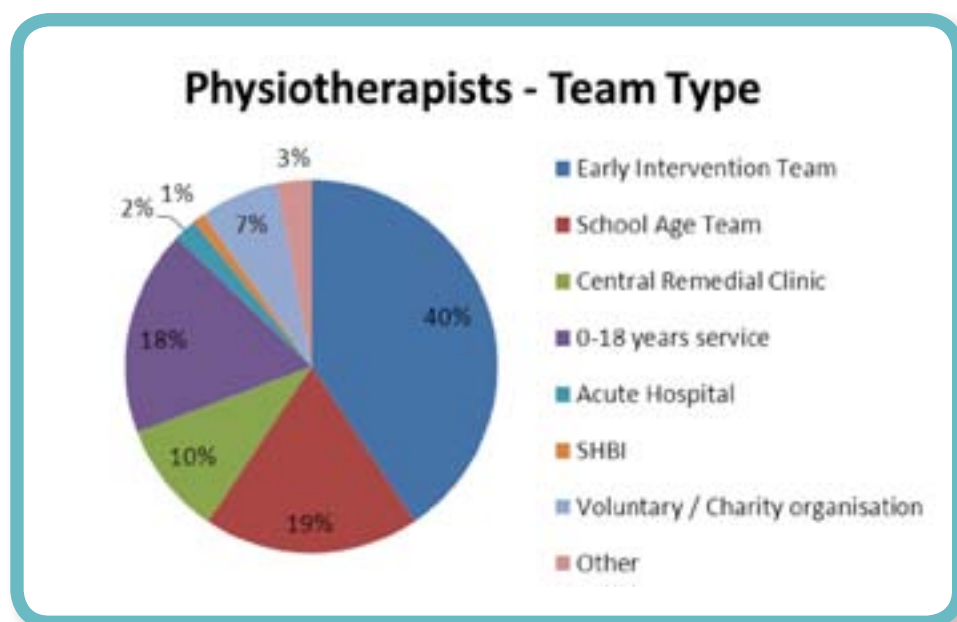


Figure 8

A small proportion of physiotherapists (4%) provided a national service. A smaller proportion reported providing a service to HSE South compared to other service areas as demonstrated in the pie chart above.

Physiotherapists had a mean of 9.2 years' experience working with children with spina bifida, ranging from 0-35 years and a

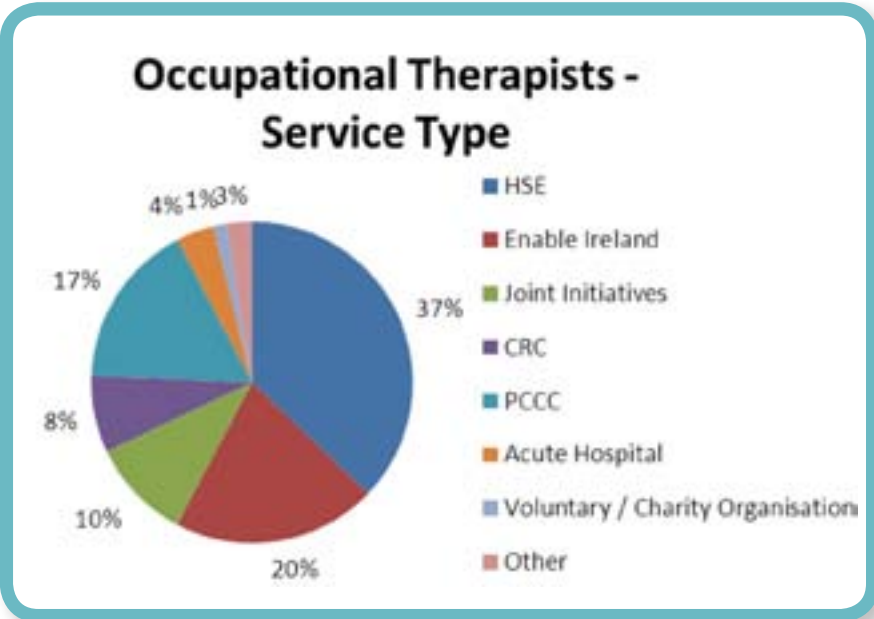
Standard Deviation of 7.461. 87% reported to be involved in providing clinical inputs, with a mean of 4.33 children (SD=10.994) with spina bifida on their open caseloads. Physiotherapists reported having no children with spina bifida on their waiting lists to access intervention.

Seven Physiotherapists also took part in qualitative interviews with the researchers.



Occupational Therapists:

Figure 9



78 Occupational Therapists responded by questionnaire. 84.4% (n=65) were female and 15.6% (n=12) were male. The services where they worked are presented in figure 9

The largest proportion of Occupational Therapists were employed by the HSE (37%), a fifth employed by Enable Ireland Services. Seventeen per cent of Occupational Therapists were employed by

the Primary, Community and Continuing Care (PCCC), and 8% by the CRC. Other employers were acute hospitals, and voluntary or charitable organisations.

Figure 10

The largest proportion of Occupational Therapists worked in services providing services for children from 0-18 years (3%) followed by early intervention community teams (29%) and school age teams (24%). Others worked in CRC, acute hospitals, and voluntary and charitable organisations.

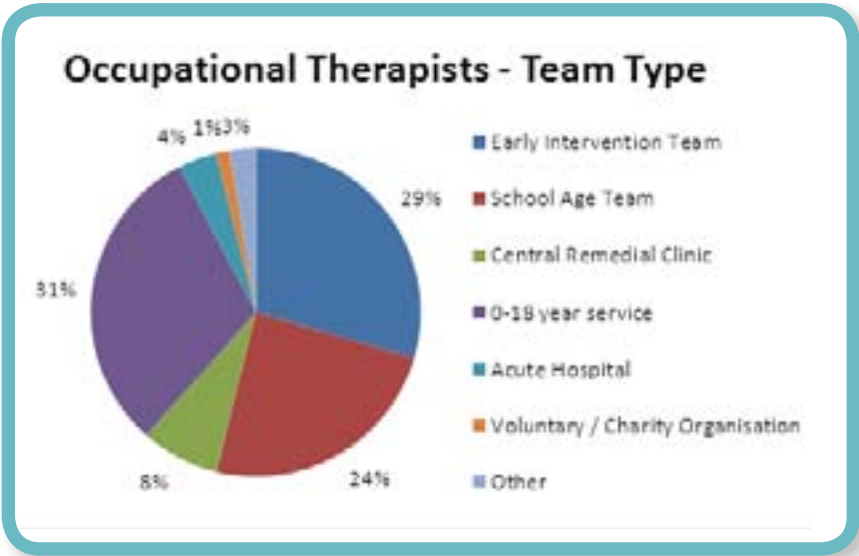
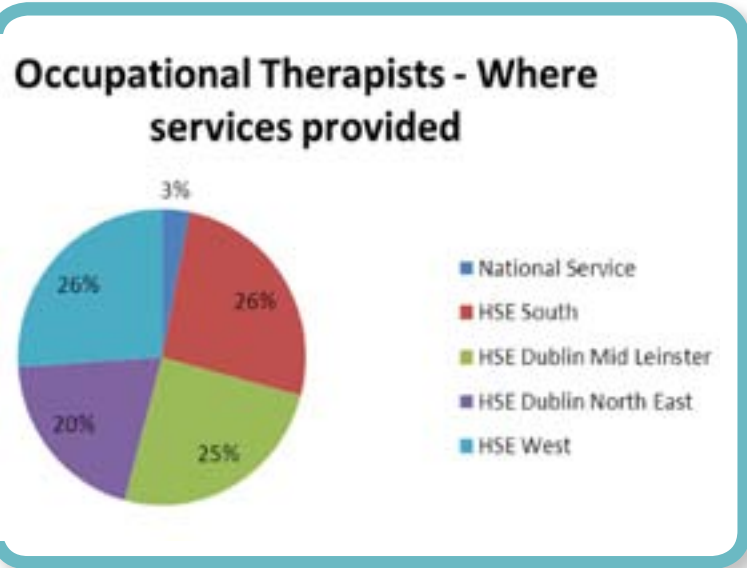


Figure 11



A small number of Occupational Therapists (n=3) reported to provide occupational therapy services to clients on a national basis.

Other HSE regions were equally represented as indicated on the chart below

Occupational Therapists had a mean of 8.1 years' experience working with children with spina bifida (SD 4.0). Of those respondents, 77% reported being involved in clinical inputs, 15% were involved in the prescription of equipment only, and a further 8% were not directly involved in interventions for children with spina bifida and therefore did not complete further questions. Occupational Therapists reported to have a mean of 2.8 children (SD = 3.0) with spina bifida on their active caseloads. They also reported to have a mean of 0.2 children with spina bifida on waiting lists for intervention, and 0.6 children with spina bifida on their closed caseloads awaiting equipment.

Seven Occupational Therapist also took part in qualitative interviews with researchers.

### ***Speech and Language Therapists:***

Nineteen Speech and Language Therapists responded to the questionnaire, all of whom were female.

Figure 12

A quarter of Speech and Language therapists were employed by Enable Ireland, a further quarter by joint initiative services. A third were employed by either PCC or HSE. In relation to teams, early intervention and school age teams are equally represented.

Speech and Language Therapists had a mean of 4.62 years' experience (range 1 – 16 years) with a mean of 9.85 children with spina bifida on their active caseloads.

They also reported to have a mean of 3.5 children with spina bifida on their waiting lists. One Speech and Language therapist took part in Qualitative Interviews with the researchers.

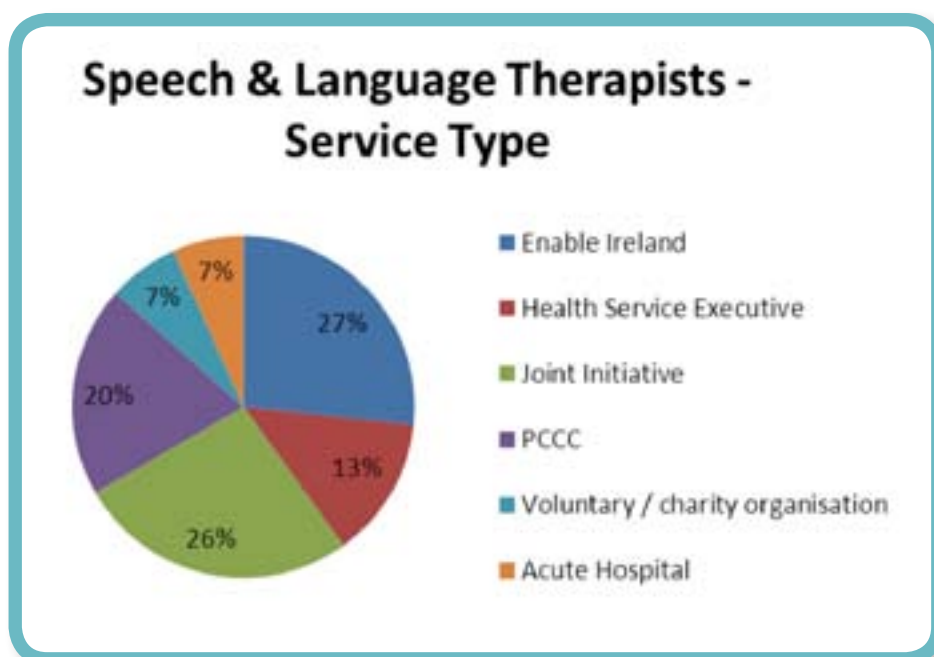
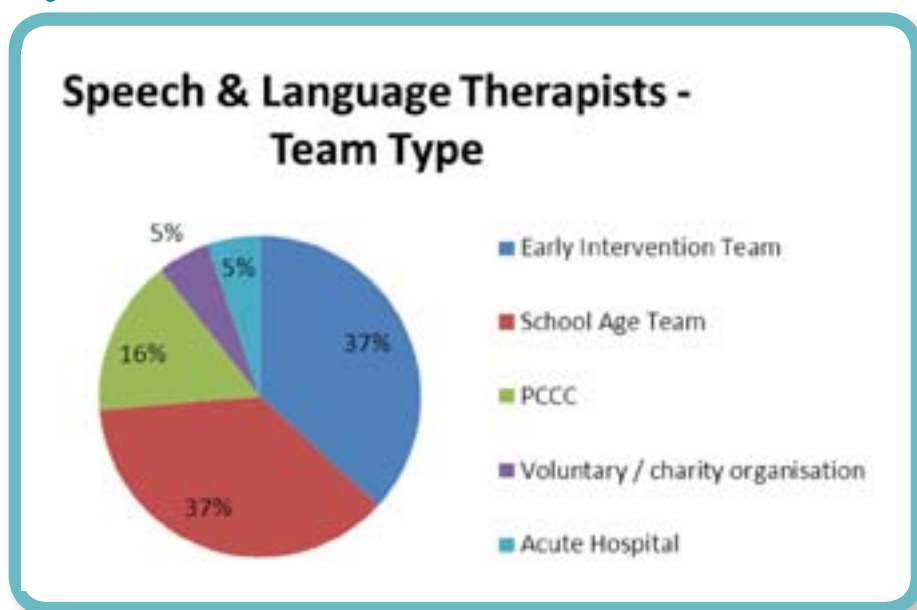


Figure 13



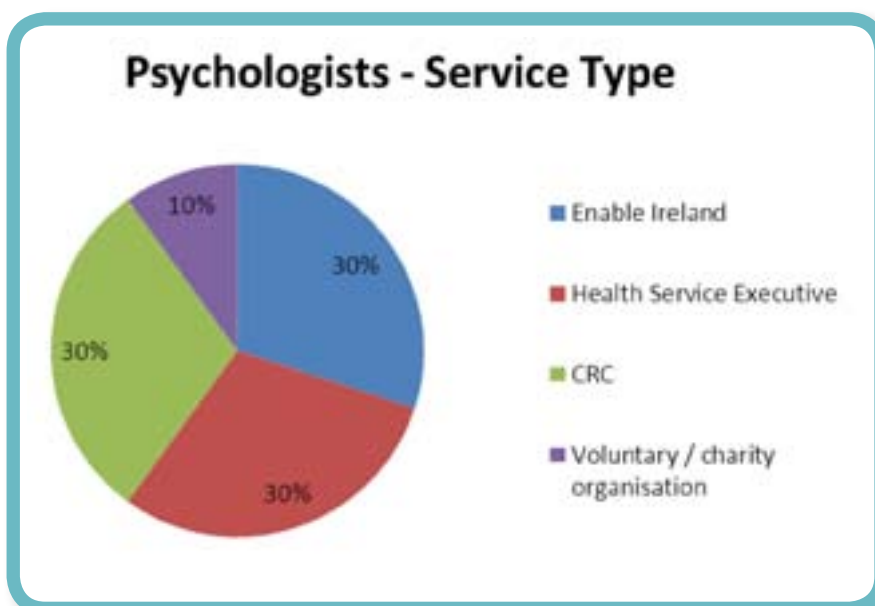
## Psychologists:

Ten Psychologists responded by questionnaire, all of whom were female.

Figure 14

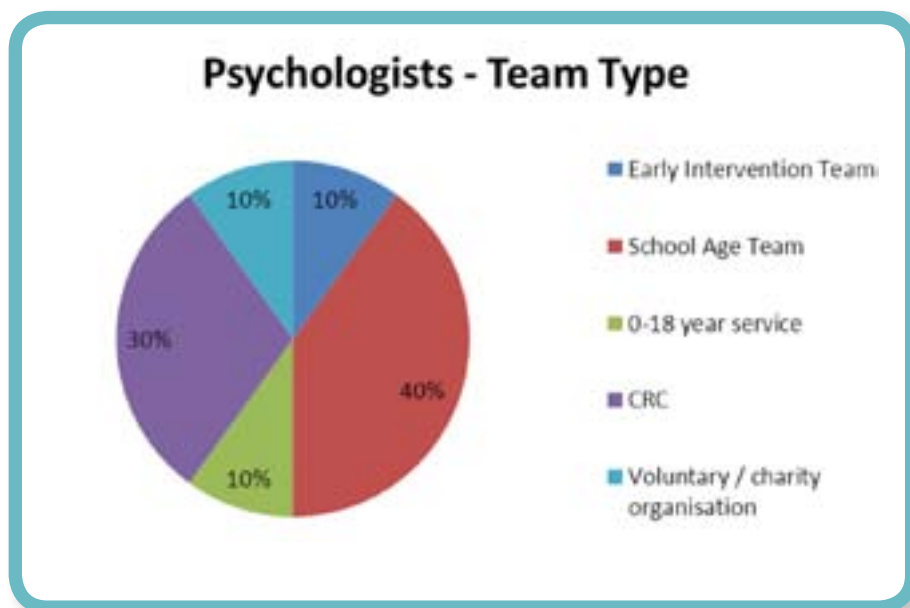
Psychologists were evenly spread between HSE, Enable Ireland and CRC with 10% employed by voluntary or charitable organisations. The largest proportion of Psychologists (40%) were on school age teams, followed by CRC.

The psychologists had a greater representation in the Dublin Mid Leinster area and no psychologist provided a national service.



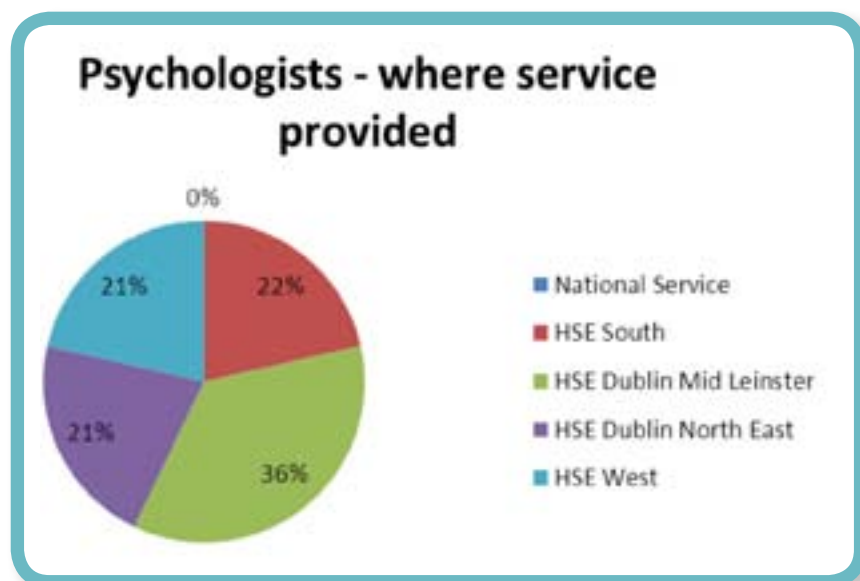
Eighty per cent of psychologist respondents reported to be involved in clinical inputs for children with spina bifida. They had a mean of 8.4 years of experience (range 6-10 years) with a mean of 7.8 children with spina bifida on their active caseloads. They reported that a mean of 1.67 children with spina bifida were on waiting lists for intervention.

Figure 15



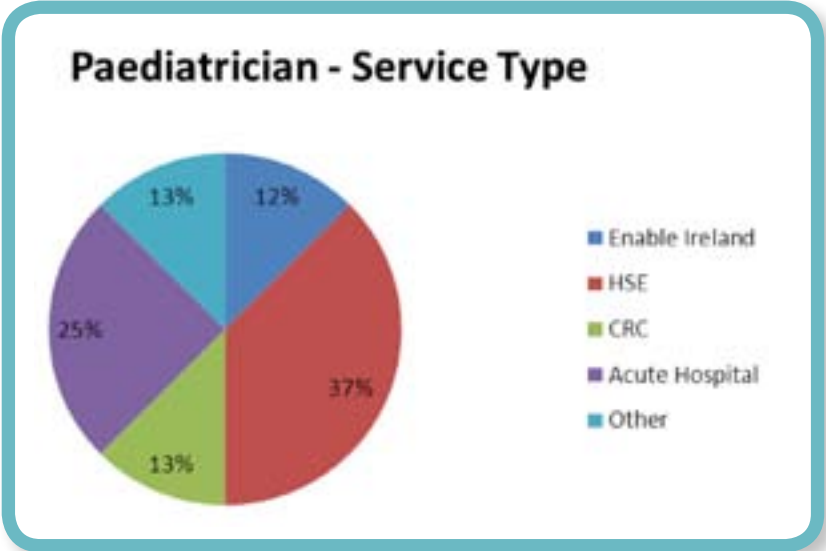
One psychologist took part in qualitative interviews with researchers

Figure 16



**Paediatricians**

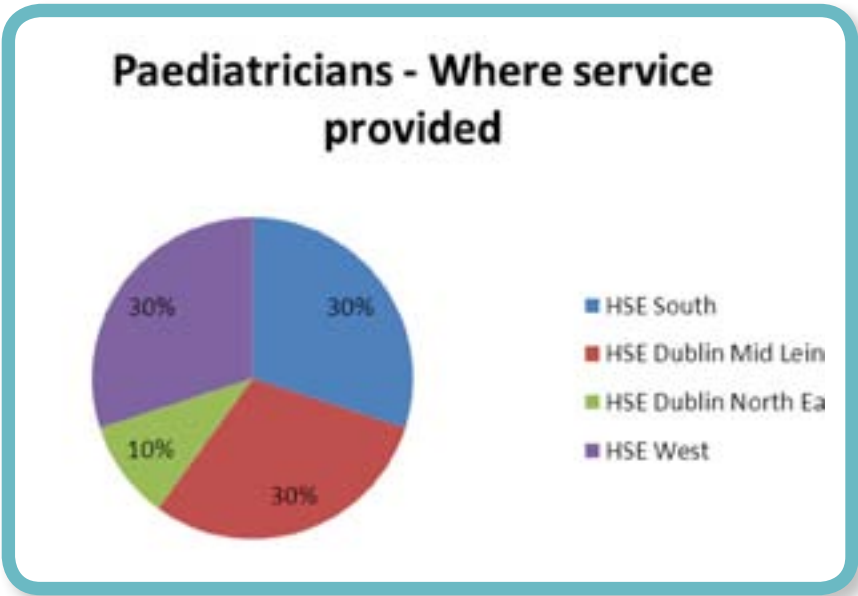
Figure 17



Eight Paediatricians responded by questionnaire. Two (25%) were male, and 6 (75%) were female.

The majority of Paediatricians were employed by the HSE or Acute hospitals. Of note, all paediatricians from the HSE and Enable Ireland were working on Early Intervention Teams.

Figure 18



The paediatricians represented all HSE areas, with less from the HSE Dublin North East area. 87.5% were involved in providing services to children with spina bifida with a mean of 11 years of experience (range 9-14 years). Paediatricians had a mean of 4.3 children with spina bifida on their active caseload and none reported having children with spina bifida on their waiting lists. Three consultant paediatricians with a special interest in

community child health took part in qualitative interviews with researchers.

# SECTION TWO

**CHAPTER FOUR**

**RESULTS:**

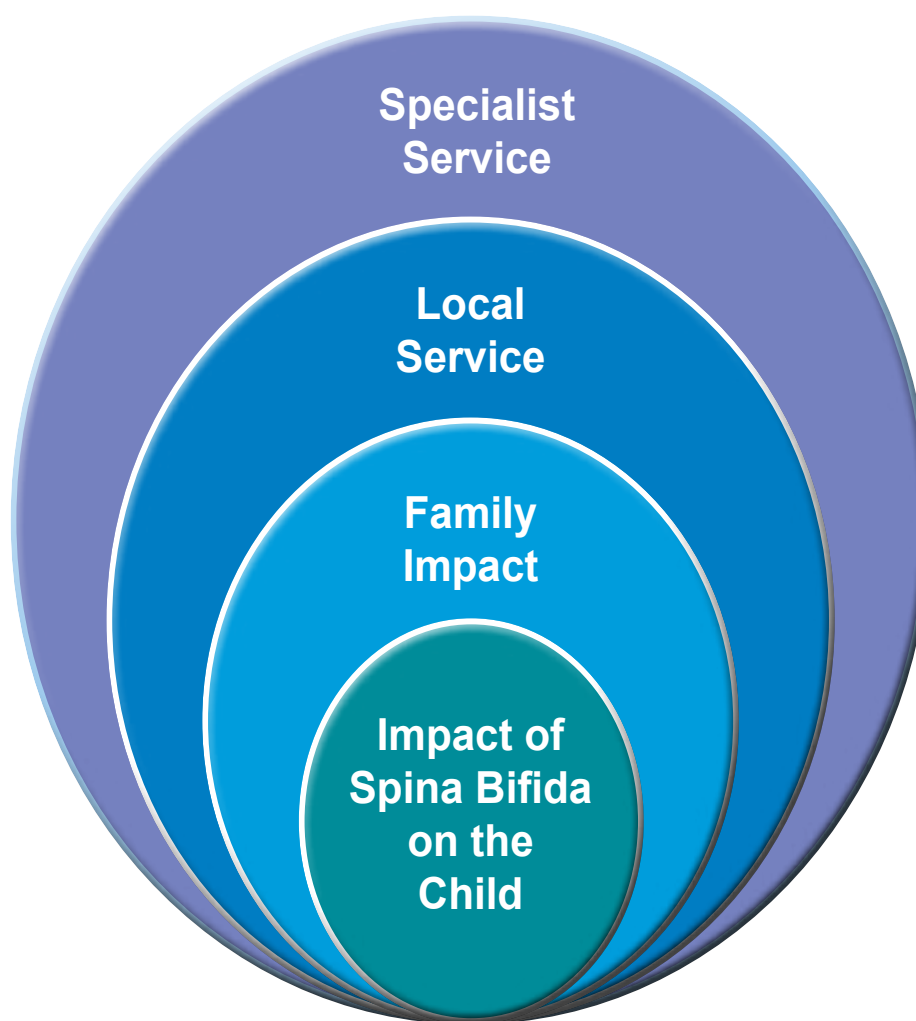
**IMPACT OF SPINA  
BIFIDA ON THE CHILD  
AND FAMILY**



# RESULTS

The four overall themes from the results are represented in the figure below. These themes demonstrate the significant impact of spina bifida on the child, on their family and the need for both local and specialist services to support these children and their families.

Figure 19 Overall result themes

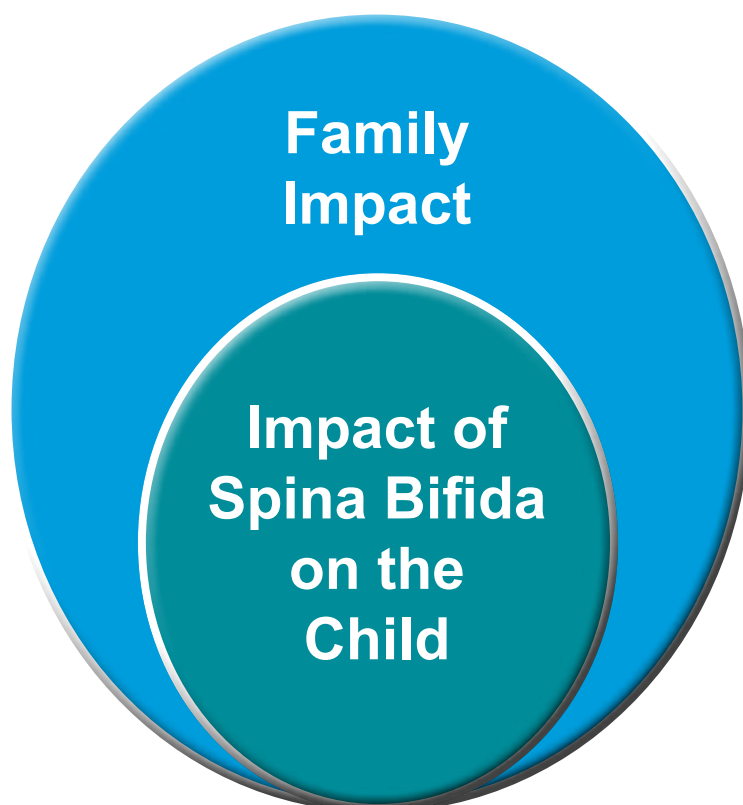


Results will be presented theme by theme in the following chapters. Firstly the impact of spina bifida on the child will be discussed, with results highlighting the physical impact and cognitive and psychosocial impact. Next results relating to the impact of spina bifida on the family are presented. Finally results relating to the health services necessary to support the child with spina bifida at both a local service and specialist service level are represented. Each subsection in these chapters includes a background, results, discussion and recommendation

section. Background includes the context and literature review relating to that specific area, results include results from the research questionnaire and interviews with parents and service providers, discussion compares findings to international practice and literature and recommendations are listed based on research findings and reviews. All recommendations are repeated in a table format in the recommendations chapter.

## **IMPACT ON THE CHILD AND FAMILY**

Figure 19b



### **4.1 PHYSICAL IMPACT**

As previously mentioned, spina bifida is a complex disability affecting the child in many aspects of their life (Liptak and El Samra, 2010). The condition-specific physical difficulties can include contractures and orthopaedic deformities of the spine and lower limbs, mobility impairment, incontinence, cognitive challenges, and pressure sores due to impaired sensation; all of which may require surgery and interventions throughout childhood to manage (Alriksson-Schmidt et al., 2013). Early closure of the lesion requires neurosurgical intervention and insertion of a CSF shunt for management of hydrocephalus is common (Vinck et al., 2006). There is an impact on mobility in most people with spina bifida which is related to the functional level of lesion (Alriksson-Schmidt et al., 2013). Weight, nutrition and general health can impact on the physical well-being of children of spina bifida with a higher rate of obesity noted in this

population group (Neter et al., 2011, Buffart et al., 2008b). Simeonsson et al. (2002) describes secondary complications as a result of spina bifida which are direct or indirect preventable consequences of the underlying primary condition. These can include reduced participation in society, delayed school attendance, scoliosis, pressure sores, obesity and dependence, renal failure and mental health issues. These can be more apparent at various life stages and are also influenced by the child's personal characteristics as well as their ability to interact with the environment demonstrating the dynamic aspect of the disability of spina bifida throughout the lifespan. The World Health Organisation's International Classification of Function Disability and Health (ICF) as previously discussed aims to ensure participation in life (World Health Organisation, 2001). Although the underlying disability is present from birth, the implications and secondary complications can be reduced by appropriate surveillance and intervention.

Other areas of considerations are sexual function, precocious puberty and latex allergy. Sexual function may be affected in spina bifida (Verhoef et al., 2005b, Cardenas et al., 2008). Erectile dysfunction can affect from 22 – 88% of males (Gamé et al., 2006). Urinary incontinence, hydrocephalus, restricted mobility and dependence on others are barriers to for adolescents and adults with spina bifida to engage in sexual relationships (Cardenas et al., 2008, Sawyer and Roberts, 1999). Verhoef et al. (2005b) reported that there is limited specific education given to adolescents in relation to sexual development and recommends sensitive discussion by health-care professionals of the impact of spina bifida on sexual and reproductive health which could alleviate unnecessary anxiety and help with the transition of children with disabilities into adulthood (Sawyer and Roberts, 1999).

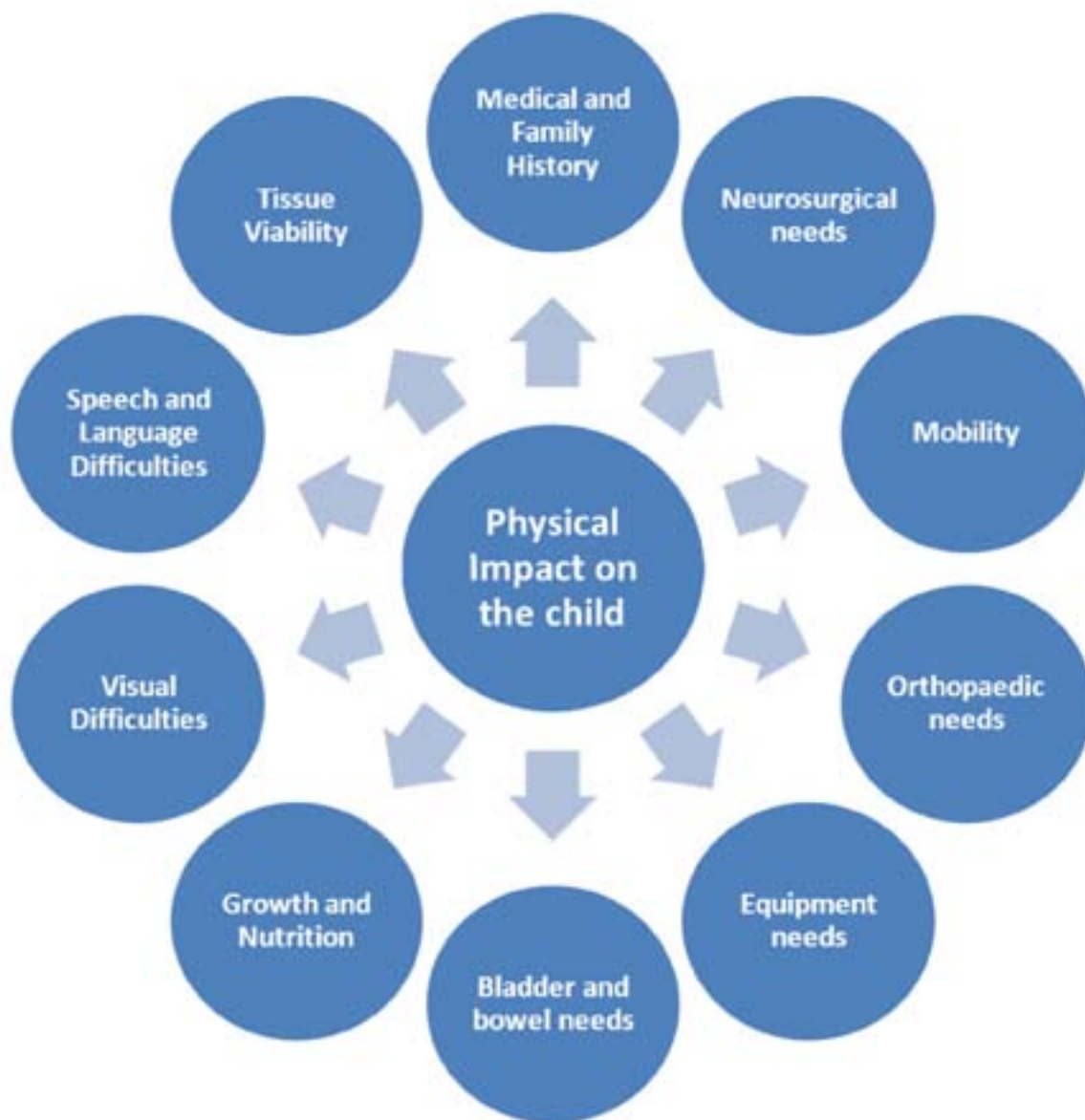
Precocious puberty is the early onset of puberty and often precedes emotional maturity which may lead to significant internal tension. Almost 20% of females with spina bifida will enter puberty before the age of 8 years compared with only 1% of the typically developing population (Levey, 2007). Verhoef et al. (2005a) advocates for counselling in relationships and sexuality which should be part of the regular care for adolescents and adults with spina bifida. Monitoring by a paediatrician and endocrinologist is necessary.

Up to a third of people with spina bifida have varying degrees latex allergy (Bowman et al., 2001, Crosthwaite et al., 2001). Some authors believe this association is due to the increased numbers of surgical procedures resulting in latex exposure however the exact mechanism is unknown (Woodhouse, 2008). It is therefore important that prophylactic measures are taken to avoid the exposure to prevent potentially serious allergic reactions (Rendeli et al., 2006).

The specific physical implications of spina bifida on the child which will be discussed in this chapter relate to the following headings (see Figure 20): medical and family history, neurosurgical needs, mobility, orthopaedic needs, equipment needs, bladder and bowel needs, growth and nutrition, visual difficulties, speech and language difficulties, and tissue viability. The cognitive and psychosocial impact will be discussed in the next section.

Figure 20

## Physical impact on the child



## ***NEUROSURGICAL NEEDS:***

### **Background**

Although it is widely accepted that folic acid prevents about 70% of Neural Tube Defects (Czeizel, 2000), the excess risk among family members with NDT suggests there are genetic risk factors involved (Byrne, 2010, Byrne and Carolan, 2006). McDonnell et al. (2014) reported a genetic predisposition for increased risk of spina bifida in Ireland. There is also a higher risk (3-5%) of having an offspring with a Neural Tube Defect if there is a history within a first degree relative (Au et al., 2010, Byrne, 2010).

**Results**

Within our sample, a positive family history of Neural Tube Defects was reported in 20%, suggesting family history of Neural Tube Defects is substantial within our population. Byrne and Carolan (2006) reported an adverse pregnancy outcome in 17.4% maternal first cousins of spina bifida, although outcomes included preterm deliveries, stillbirths, and miscarriages.

**Level of lesion**

When asked regarding the level of lesion, the largest proportion of children were reported to have a lumbar (27%), lumbosacral (19%) or sacral (12%) lesion. However, a very large proportion of parents (34%) were unsure of the level of their child’s lesion. This is represented in the figures below. When the figures are adjusted by removing those that indicated they did not know their level of lesion, the spread of respondents is presented in Figure 22.

Figure 21

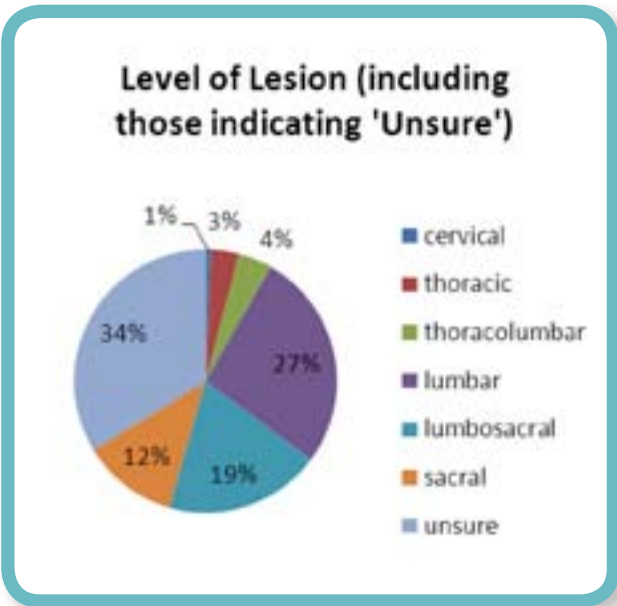


Figure 22

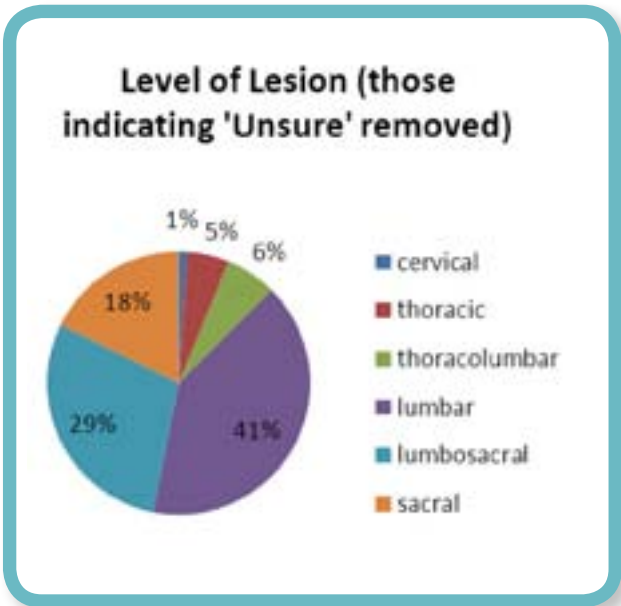


Table 2 compares the spread of level of lesions in our sample (adjusted and non-adjusted charts above) to other studies internationally. The unadjusted chart includes the large proportion of where the level of lesion was unknown and would suggest a lower number of low level lesions compared to other studies. Possible explanations for a lower proportion of low lumbar and sacral lesions reported within our sample may be explained by a lack of parental knowledge about the level of lesion and the lack of termination of pregnancy in Ireland resulting in more high level lesions being carried to term. The lack of parental knowledge is of concern due to the physical implication of the level of lesion and makes comparison with other studies difficult.

Table 2

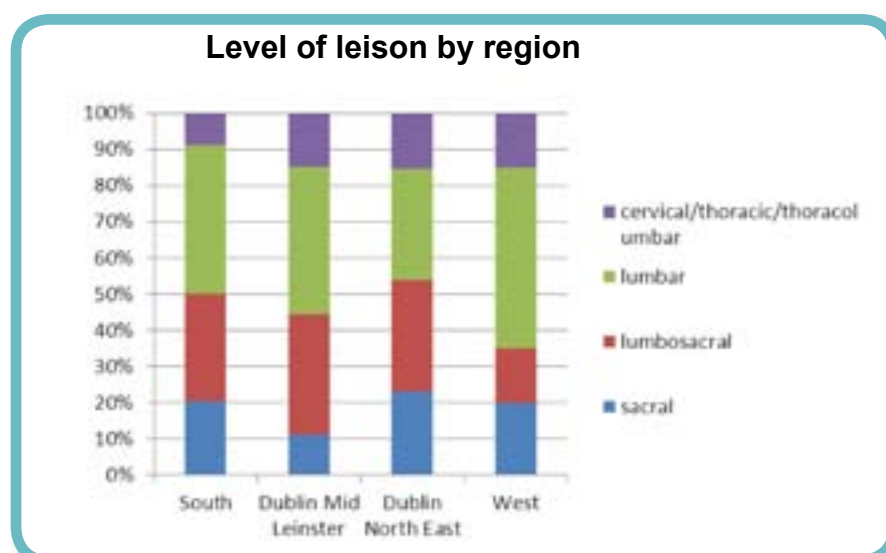
## Level of lesion compared to literature

	Location	N=	Cervical	Thoracic / Thoracolumbar	Lumbar / Lumbosacral	Sacral
Current Study	Ireland	155	1%	7%	46%	12%
Current Study (adjusted chart)	Ireland	155	1%	11%	70%	18%
(Johnson et al., 2007)	Seattle, US	348	NIL	29%	36%	29%
Alatise et al., 2006	Nigeria	106	1%	6%	57%	16%
RINTOUL 2002	Philadelphia, US	297	NIL	23%	62%	15%
(Iborra et al., 1999)	Barcelona, Spain	332	NIL	21.7%	60.8%	17.3%

In the adjusted chart however, the numbers in each level of lesion group are comparable to previous studies. Internationally, antenatal counselling based on level of lesion and brain abnormalities can influence whether parents choose to continue with a pregnancy (Rintoul et al., 2002). These factors may impact on the level of lesion of fetuses that are carried to term. These would warrant comparison with clinical data to determine the spread of lesions within Ireland.

No significant difference was seen in the levels of lesion across the four national HSE regions, suggesting that a geographical difference is not present in this current population. In all areas other than Dublin North East, Lumbar lesions were the most prevalent. This spread is represented in the chart (Figure 23).

Figure 23



# Hydrocephalus

## Background

Hydrocephalus co-occurs with spina bifida in 80-95% of cases (Shaer et al., 2007). Del Bigio (2010) defines hydrocephalus as a “pathological enlargement of the intracranial cerebrospinal fluid (CSF) filled spaces; usually this refers to the cerebral ventricles” which is a result of an imbalance between production and absorption of CSF. Hydrocephalus is secondary to Chiari II malformation of the hind brain; and these children typically require CSF shunts to reduce intra-cranial pressure (Vinck et al., 2006).

The Chiari II malformation affects almost all children with spina bifida resulting in the downward herniation of the cerebellum and brainstem into the foramen magnum and the cervical canal (Crosthwaite et al., 2001). Chiari II malformation remains an important cause of death and disability in spina bifida. However, whether surgical intervention will occur is very dependent on the presenting symptoms as the results of early and aggressive surgery are mixed (Liptak, 2003).

Both spina bifida and hydrocephalus can be identified antenatally depending on their severity (Shaer et al., 2007). If hydrocephalus is not already present at birth, it often develops soon after the surgical repair or closure of the open spina bifida lesion. In the 1950's Holter and Spitz developed the first effective ventricular shunt system to treat hydrocephalus, redirecting the fluid away from the ventricles through a one way valve system. Prior to this development uncontrolled hydrocephalus was the leading cause of death in children with open spina bifida (Shaer et al., 2007, Liptak and El Samra, 2010).

The development of the ventriculo-peritoneal (VP) shunt was reported in a 25 year review to have revolutionised care for those with spina bifida and has led to increased survival rates (Bowman et al., 2001). While no standard protocol for the management of hydrocephalus currently exists (Liptak, 2003), most studies report the use of cerebrospinal fluid (CSF) shunts including VP shunts to manage hydrocephalus. CSF shunts are generally used with infants who have rapidly progressing hydrocephalus or those who present with acute neurological changes, such as stridor, swallowing dysfunction or central apnoea, with or without significant change in the ventricular size (McLone and Dias, 2003). Initial shunt insertion used to be inserted at birth concurrently with lesion closure in the presence of hydrocephalus, However, research now advocates active surveillance of the newborn; with mild to moderate ventriculomegaly both clinically and radiographically to determine which infants will benefit from permanent CSF diversion (McLone and Dias, 2003, Chakraborty et al., 2008, Warf et al., 2009, Fletcher and Brei, 2010, Bowman and McLone, 2010).

Unfortunately inserting a CSF shunt for management of hydrocephalus brings with it various complications and has been associated with shunt infection in between 3 and 35% of cases

(Margaron et al., 2010, Kulkarni et al., 2001). Long-term complications of shunt insertion include infection, blockage, malfunction and multiple revision (Hunt et al., 1999, Barf et al., 2003, Bowman and McLone, 2010). A high rate of revision (especially when occurring after the second birthday) results in lower levels of independence and employment in later life, higher mortality, and reduced memory and quality of life (Hunt et al., 1999, Oakeshott et al., 2010, Dennis et al., 2002, Hetherington et al., 2006). Lomax-Bream et al. (2007) identified that the presence of hydrocephalus and shunt significantly impacted development of cognition and motor skills, but did not have an effect on the development of language. Specific results relating to cognition and social, emotional and psychological functioning will be discussed in chapter 4. Rates of shunt insertion have been reported to be between 52% and 86% in children with spina bifida (Johnson et al., 2007, Chakraborty et al., 2008, Bowman et al., 2001, Iborra et al., 1999, Adzick et al., 2011)

Rintoul et al. (2002) reported that the median number of shunt revisions was 2 per patient with a range of 0 to 21 with the majority of these occurring in the first year of life. He also noted that the requirement for shunt was statistically related to the functional and anatomical level of lesion, with sacral level lesions being statistically less likely to require shunting. Bowman et al. (2001) in a 25 year follow up study found a revision rate of 95% suggesting these difficulties may persist into adulthood.

## Results

Within our sample, 69% of parents reported that their child had a history of hydrocephalus, and of the total sample 64% of the children had a CSF shunt (the phrasing in the questionnaire was VP shunt) in place to manage their hydrocephalus. Of the 96 children who had a shunt in situ, 44% indicated that a shunt was placed within the first week of life, with an additional 50% requiring a shunt within the first 6 months of life. Only 5% of parents indicated that their child had a shunt first inserted later than the first 6 months of life.

Table 3

When Ventriculo-peritoneal Shunt inserted

Ventriculo-peritoneal Shunt	N=	%
First week of life	42	28
First 6 months of life	48	32
First two years of life	4	2.7
After 2 years	1	0.7
Removed and not replaced	1	0.7
No Shunt	54	36

No information was sought in relation to complication or infection rates. The largest proportion of children (31%) had only one shunt surgery, the average number of shunt-related surgeries was 2.8 and only 20% of the respondents indicated that their child required more than 4 shunt related surgeries.



## Discussion

A strong family history of neural tube defects was noted in this sample as compared to previous studies (Byrne, 2010, Byrne and Carolan, 2006). A review of pre-conceptual care and education of parents about increased risk, as well as increased public awareness relating to folic acid fortification and supplementation is recommended in line with recommendations by (McDonnell et al., 2014).

Lumbar level lesions were evident with the largest proportion of children, followed by lumbosacral lesions. However a third of the parents sampled were not aware of the level of their child's lesion suggesting a need for further education from the specialist centres. With such a high proportion of parents being unsure of the level of lesion it is difficult to compare our sample with other international studies. Education to parents about their child's level of lesion should occur from an early stage through the spina bifida MDT from as early as the antenatal stage.

The numbers with shunts in situ in this study (64%) are lower than historical reports of 86% (Bowman et al., 2001), but in line with more up to date literature who recommend a more conservative approach to Shunt insertion with rates as low as 52% (Fletcher and Brei, 2010, Chakraborty et al., 2008). Of those that do require CSF shunting, the largest proportion were placed within the first 6 months of life and only required one surgery however 20% required more than 4 shunt related surgeries.

## Recommendations

1. Directed education regarding future pregnancy risk to extended families of children born with neural tube defects should be implemented by the national spina bifida service in Temple Street
2. Parents should receive education about their child's level of lesion to help with understanding the potential impact.

# **MOBILITY**

## **Background**

There is a huge variation in the levels of mobility achieved in children with spina bifida which can be influenced by many factors such as level of lesion, spasticity around hip or knee, lower limb contractures, hip dislocation, scoliosis, impaired cognitive status, impaired hand function, visuo-spatial function, the involvement of the parents, and the therapy program (Danielsson et al., 2008, Thomson and Segal, 2010, Liptak and El Samra, 2010). Bartonek and Saraste (2001) further commented that while lower limb contractures, hip dislocation, or surgical repair of scoliosis do not significantly impact ambulation potential, they do affect the energy expenditure required for ambulation. In addition, balance disturbances, which were significantly higher in children whom had 2 or more shunt revisions, significantly affect ambulation (Bartonek and Saraste, 2001).

The functional level of lesion is very much related to the child's mobility potential. Mayfield (1991) reported that 100% of children with a sacral lesion can achieve ambulation, compared with 95% of lumbar lesions. Similarly, Bowman et al. (2001) in a 25 year follow up study reported that 93% of patients with a sacral level continue to ambulate 100% of the time, 91% with a L5 lesion ambulate and 57% of patients with an L4 lesion ambulate. None of the patients with a lesion above L3 ambulated as their main mode of locomotion.

Ambulatory potential is significantly related to the presence of quadriceps power grade 4 - 5 in the Oxford grading system (functional lesion level L3/4 or below) (Danielsson et al., 2008, McDonald et al., 1991, Huff and Ramsey, 1978, Mazur and Menelaus, 1991). Mazur and Menelaus (1991) reported that 98% of their sample of children with spina bifida with a quadriceps strength of grade 4-5 were at least household ambulators with 82% being community ambulators. Of those with quadriceps strength of grade 3 or less, 88% were exclusive wheelchair users. Swank and Dias (1994) identified the motor level as well as sitting balance to be most associated with the potential to ambulate.

In order to achieve ambulation appropriate orthotic devices and equipment are essential (Brown, 2001). This requires collaboration of the orthopaedic surgeon, Physiotherapist, Orthotist and Occupational Therapist. The onset of walking in children with spina bifida has been reported to be delayed by an average of 2 years and continues to develop until 6 years of age (Teulier et al., 2009, Bartonek, 2010).

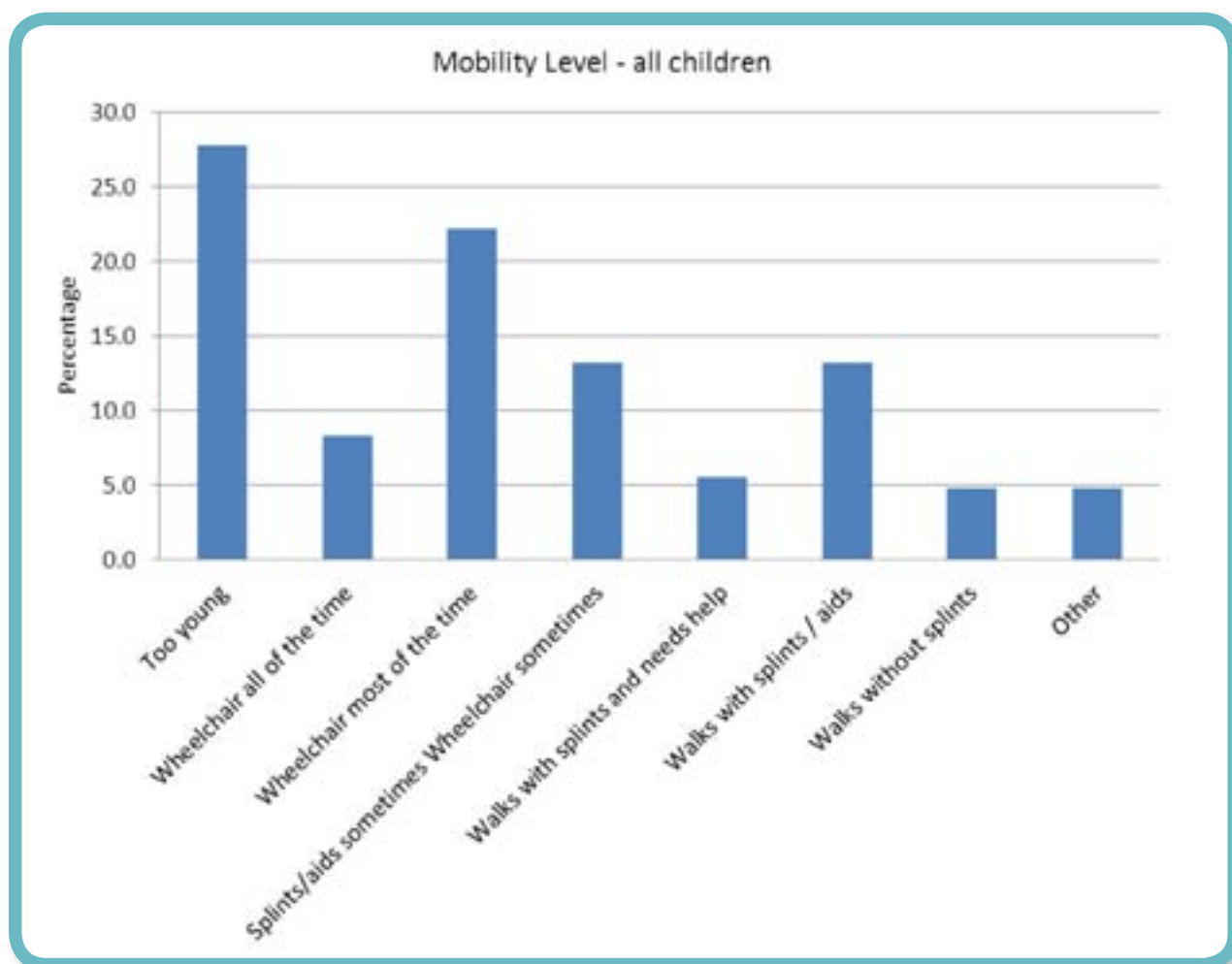
Very often the decision of whether to continue walking or opt for full-time wheelchair use is eventually a matter of choice based on individual ability and lifestyle as the child approaches adolescence. A wheelchair offers advantages of speed, efficiency, and attractiveness (Brown, 2001, Liptak and El Samra, 2010). Although mobility decreases from early childhood to the early teen years, those patients who remain mobile in their teens continue to ambulate the

majority of the time (75–100%) in their young adult years (Bowman et al., 2001)

## Results

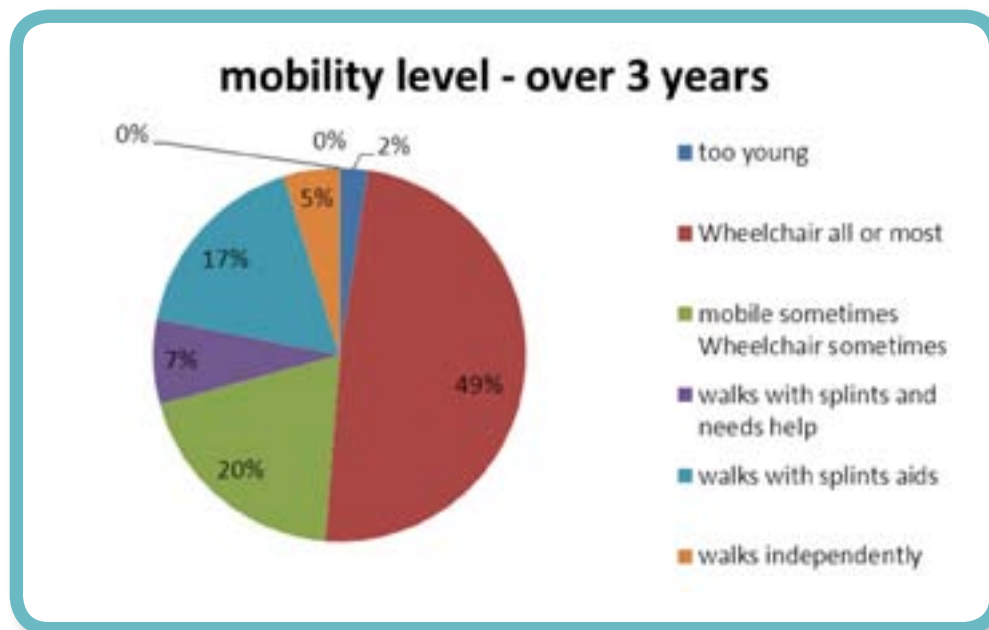
Within our sample, 28% of parents indicated that their child was too young for them to know their mobility potential in the future (Figure 24). 31% of parents noted that their child is a wheelchair user either all or most of the time, and only 5% walked without splints. The remaining 32% have equipment dependent mobility requiring a variety of splints / aids and mobility aids.

Figure 24



By the age of 3 years when the mobility potential is more apparent (Teulier et al., 2009, Bartonek, 2010), 49% were mobile with a variety of ability and equipment needs to achieve this (Figure 25). 69% of children used a wheelchair at least some of the time and 44% required splints or orthoses for mobility. Only 5% were independently mobile without aids.

Figure 25



Qualitatively parents reported that the impact of the disability became more obvious after toddlerhood when motor delays began to set them apart from their peers. Parents expressed aspirations that their child would walk.

*“Because it’s a baby most of the special needs you wouldn’t notice it then because all babies are in nappies and all babies are being carried so you wouldn’t specify” (Parent / Guardian)*

*“His only wish is to walk, if you ask him what his wish he says I wish I could walk” (Parent / Guardian)*

During the focus group when asked about how they feel about how their body works some children reported frustration in relation to the difficulties with their physical abilities.

*“I don’t feel good about it, (how my body works) it doesn’t work all the time, it might work for a few months and then it stops and then it works again” (Focus Group Child)*

## Discussion

A variety of mobility levels are represented within our sample, with the majority requiring a wheelchair for mobility. Our figures for equipment assisted mobility are comparable to a previous study by Johnson et al. (2007) who reported 57% of adolescents and young adults sampled from the children’s hospital, Seattle used a manual or electric wheelchair, 35% used braces, and 23% used walking aid. However, they reported a much higher proportion of independent ambulation without aids or braces outside of the home at 30% compared to 5%

within our sample. Similarly, Bowman et al. (2001) reported 46% to be mobile with or without aids. Although the reasons for this were not explored, potential explanations may be due to unclear proportions of lesions levels as discussed in the previous section. Early access to therapy, equipment and orthopaedic surgery can all influence the mobility potential of these children which will be addressed later in this report.

Parents need to be informed of the rationale and reasoning behind different mobility potential and aids required by assessing the child's muscle function, cognitive ability and motivation; establishing goals and discussing expectations. Merkens (2006) in the guidelines for spina bifida health care services throughout the lifespan recommends annual manual muscle testing to 'individualise interventions and therapy goals as well as to monitor for neurological threat'.

## **Recommendation**

1. It is recommended that annual muscle charting be performed by an adequately trained Physiotherapist to monitor muscle function and document change. Communication between local and specialist Physiotherapists is essential to determine functional ability and results should be discussed with parents.

## **ORTHOPAEDIC NEEDS**

### **Background**

The most common orthopaedic complications associated with spina bifida are hip dysplasia or dislocation, scoliosis, and foot and ankle deformities which are described in the subsequent sections. Other complications discussed in the literature included knee pain, in particular the impact of valgus knee strain which is common due to the weakness of the abductor musculature of the hip which can be a cause of knee pain and limited mobility (Thomson and Segal, 2010). This can be managed by exercise physiotherapy and appropriate orthoses.

### **Foot and Ankle Deformities:**

Up to 68% of children with spina bifida have deformities of the foot and ankle. (Brown, 2001). The most common deformities being equinus contracture, clubfeet, and calcaneal deformities with excessive dorsiflexion (Thomson and Segal, 2010). Most foot deformities although initially treated with the Ponseti method for the treatment of idiopathic clubfeet, will require surgical intervention (Brown, 2001). Surgery is recommended early if Ponseti method is failing irrespective of weight-bearing status.

## **Hip Dislocations:**

There is a lot of debate within the literature in relation to the surgical management of hip dislocations in spina bifida. Thirty per cent of children with spina bifida can present with hip dislocation occurring at birth or throughout childhood. Children with a lesion level of L3 or lower tend to dislocate earlier, whereas thoracic level lesions or lesion level L1/2 can continue to dislocate until the age of 11 due to muscle imbalance, as well as the influence of prolonged sitting or scoliosis (Broughton et al., 1993). Huff and Ramsey (1978) noted that the presence of active quadriceps without active hip abduction was a risk factor for dislocated or subluxed hips.

In a large review of literature in relation to validity of hip and spine surgery in spina bifida Wright (2011) reported that there is a lack of level 1 evidence in this area. The conclusions indicated that hip reduction surgery does not improve function but may be appropriate in low-level unilateral dislocation. Children with lesions above L4 have limited probability of walking beyond adolescence and therefore there was no support for surgical treatment in this case. In low level unilateral dislocation there was support for surgical treatment to prevent leg length discrepancy although the literature did not have functional outcome measures to monitor for improved function. Broughton et al. (1993) conclude that there is no benefit in prophylactic surgery to correct muscle imbalance. The additional risks of surgery must also be considered as relocation of dislocated hips may be complicated by fracture, infection, pain, and stiffness with a high rate of re-dislocation due to poor musculature control (Wright, 2011, Brown, 2001). Gabrieli et al. (2003) suggest the treatment of soft tissue contractures at the hip give better functional outcomes than surgical repair of the dislocation.

The importance of postural management is also highlighted in the management of hip integrity (Pountney et al., 2002). From early in infancy, positioning, stretching and light bracing are recommended to maintain flexibility and decrease the risk of developing contractures (Brown, 2001)

## **Scoliosis:**

Scoliosis has been reported to be present in as many as 52% of children with spina bifida (Ascani et al., 1986, Trivedi et al., 2002). Scoliosis has the potential to cause pelvic obliquity, spinal imbalance, seating difficulties, and in the long-term pulmonary dysfunction. It requires vigilance and monitoring throughout the growing years. The treatment goals for scoliosis in patients with spina bifida include preventing progression of the underlying deformity, achieving a solid fusion, maximizing functional independence, increase sitting tolerance, and achieve a level pelvis with a balanced spine in both coronal and sagittal planes (Thomson and Segal, 2010). In a large review of orthopaedic literature in spina bifida Wright (2011) reported that although scoliosis surgery can improve radiographic parameters, the overall effect on physical function was mixed. Wai et al. (2005) also commented that scoliosis correction did not improve physical functioning and self-perception which are the most important outcomes. The risks for

surgery are also high with complication rates such as infection and non-union.

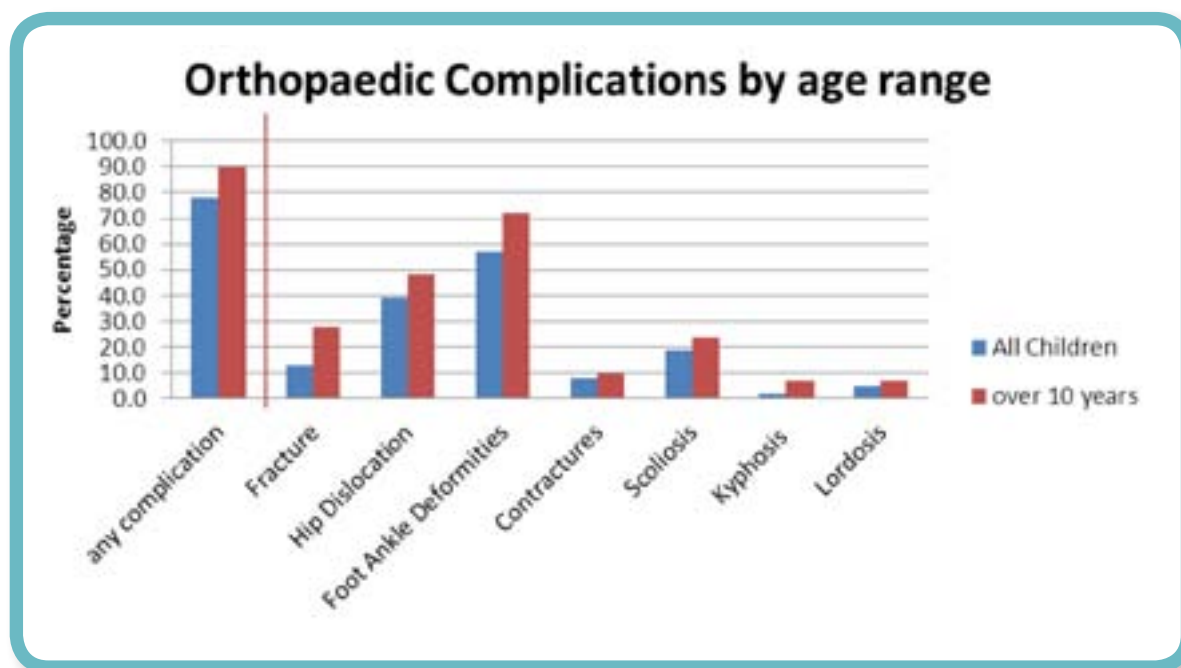
## Fractures:

Children with spina bifida are predisposed to fracture of their lower extremities especially after periods of immobilisation and more common in early adolescence (Dosa et al., 2007). Akbar et al (2010) reported as incidence of 11% of one or more fractures in patients with Myelomeningocele and found that those with thoracic level lesions had a six fold higher fracture risk than those with sacral level lesions. Brown (2001) advocates the need for regular physiotherapy and an early standing programme to promote bone health.

## Results

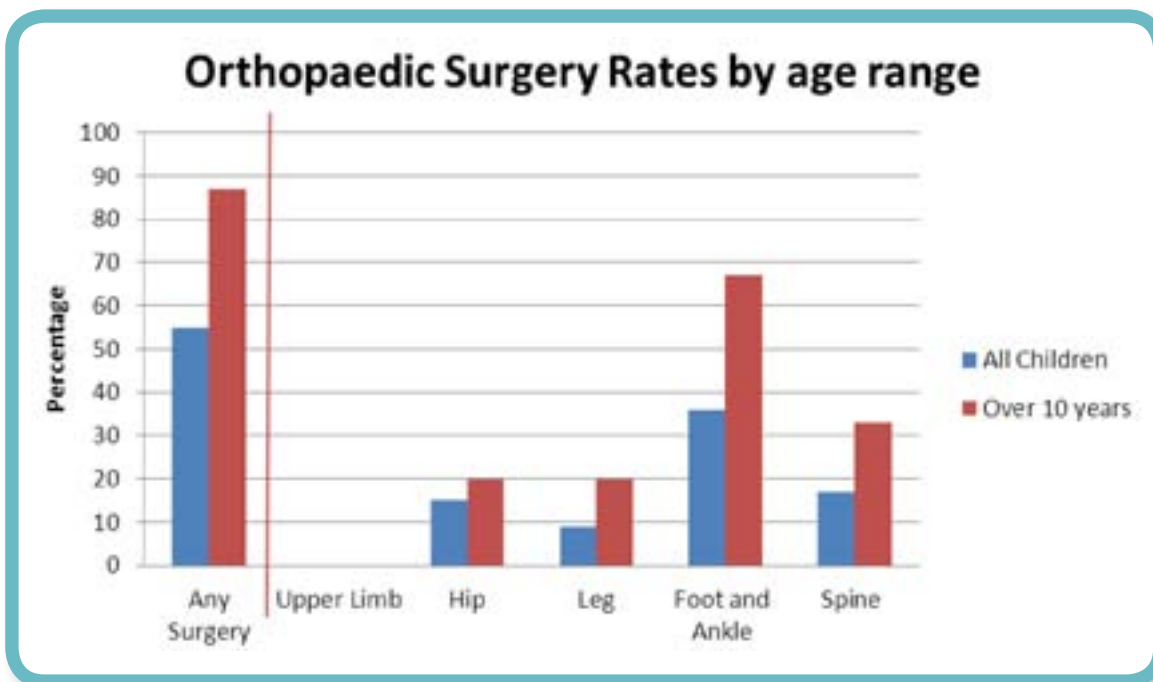
78% of parents indicated that their child had an orthopaedic complication related to their spina bifida. This increased to 90% for parents of children over 10 years (Figure 26). The most common complications were foot and ankle deformities, and all complications were higher in the over 10 year age group.

Figure 26



In relation to the frequency of orthopaedic surgery, 55% of children were reported to have undergone orthopaedic surgery of some kind; however this figure increased to 87% in the over 10 years age group. The most common surgery was related to the foot and ankle with all surgical rates being higher in the over 10 age range (Figure 27).

Figure 27



## Discussion

In conclusion, orthopaedic complication and surgical rates are high in our population. The needs and complexity of surgical intervention increase with age. Orthopaedic evaluation is required for all children with spina bifida to ensure a good functional outcome and quality of life. The most common orthopaedic complications presenting in spina bifida children are foot deformities, hip dislocations, and scoliosis. The debate of if and when to intervene must be considered along with the perceived benefits in functional outcome rather than radiological outcome. Secondary orthopaedic complications such as progressing scoliosis and flexion contractures must also be prevented by regular review and monitoring.

## Recommendations

1. Frequent orthopaedic review is required for children with spina bifida throughout their lifespan.
2. Orthopaedic surgery should only be completed when warranted and for functional gain



## **EQUIPMENT NEEDS**

### **Background**

Assistive devices are used in spina bifida to promote mobility, manipulative skills, personal care and cognitive functioning (Johnson et al., 2007). As health-related quality of life is reported to be significantly associated with independent mobility for children with spina bifida (Mazur and Kyle, 2004) the provision of equipment to support independence is vital. The rationale for the most common pieces of equipment used with children with spina bifida is discussed below.

### **Orthoses:**

Ankle Foot Orthoses (AFOs) are the most frequently used orthoses in children with spina bifida to assist mobility (Vankoski et al., 2000). The use of an appropriate AFO can correct functional deficits, improve alignment of the lower limb, protect from pathological forces, reduce energy costs, increase stride length and walking speed in children with low level lesions (Malas, 2011, Thomson et al., 1999, Bartonek et al., 2007). Bartonek (2010) reported that although orthotic usage was equally common in both ambulatory and non-ambulatory children, the ambulatory children most frequently used an AFO or KAFO (Knee Ankle Foot Orthoses), for at least 5 hours a day whereas the non-ambulatory children most commonly used RGOs (reciprocal gait orthoses) and standing frames.

The use and benefit of achieving ambulation with complex orthoses such as RGOs and HKAFOs (Hip Knee Ankle Foot Orthoses) in children with high level lesions have been questioned in the literature. The financial cost of the orthoses, orthopaedic and physiotherapy input required to achieve mobility must be considered against the patient's potential to achieve long-term mobility, motivation and available parental support (Schoenmakers et al., 2005, Roussos et al., 2001, Mazur and Kyle, 2004). Johnson et al. (2007) demonstrated no benefit of ambulation in children with high level spina bifida lesions in terms of renal health, fracture risk, obesity, participation in sport, incontinence, ADLs or orthopaedic procedures, but suggested the need for further research with functional outcomes. On the other hand Roussos et al. (2001) did suggest financial benefits in maintaining ambulation due to reduced risk of pressure sores and fractures. Mazur et al. (1989) compared 2 similar groups of children with spina bifida, one group who participated in an early walking program, and another group who had a wheelchair prescribed early in life. The patients who walked early had fewer fractures and pressure sores, were more independent, and were better able to transfer than were the patients who had used a wheelchair from early in life. However, during childhood and early adolescence, the patients who had always used a wheelchair had spent fewer days in the hospital than did those who had participated in the walking program. There were no major differences between the two groups with regard to skills of daily living, function of the hands, and frequency and severity of obesity. In a long term follow up of RGO users Roussos et al.

(2001) found that children used these devices for an average of 9.6 years, continuing their ambulation for up to 7.5 years. It is recommended that patients who are candidates for an RGO first undergo an evaluation in a used but adjusted RGO and only those who achieve a minimum of ten strides on repeated occasions receive their own RGO (Katz-Leurer et al., 2004). Success is inversely correlated with the neurological level and correlated directly with parental cooperation.

### **Wheelchairs:**

The importance of appropriate wheelchair and seating cannot be underestimated and must meet the physical and social needs of the child, suit the environment, be safe and durable, and achieve appropriate postural control (WHO 2008). Wheelchair use in children with spina bifida provides appropriate postural support, comfort, skin protection and stability to ensure participation in functional activities at home and school and to facilitate psychosocial and cognitive development (Wright et al., 2010).

Children with mid and low lumbar lesion levels have increased heart rate during walking and higher walking-related energy cost than peers without disability (Bartonek and Saraste, 2001) which supports wheelchair use to enhance participation in society. Bartonek (2010) reported a high acceptance of early wheelchair use, even for children with expected ambulatory function in order to encourage early participation; with manual wheelchairs used more commonly by ambulators and powered wheelchairs more commonly in non-ambulators. The individual needs of each child must be considered when prescribing a wheelchair, and may be very complex depending on context (Batavia et al., 2001).

### **Standing Frames:**

The benefits of standing for children with spina bifida have been reported as increased urinary drainage, improved bowel function and bone health (Mazur et al., 1989). Standers stabilise the joints of the lower limbs with footplates on the bottom of the orthoses providing a sufficient support area within which the centre of gravity of the patient can be located so as to prevent the intrinsically stable patient toppling over (Stallard et al., 2003). The standing frame allows the child to be upright at home and school which enhances social, cognitive and psychological development. They can be used with children with lesions as high as C6 (Stallard et al., 2003) and have been recommended from the age of 12-15 months (Stallard et al., 2003, Brown, 2001).

### **Sleep Systems:**

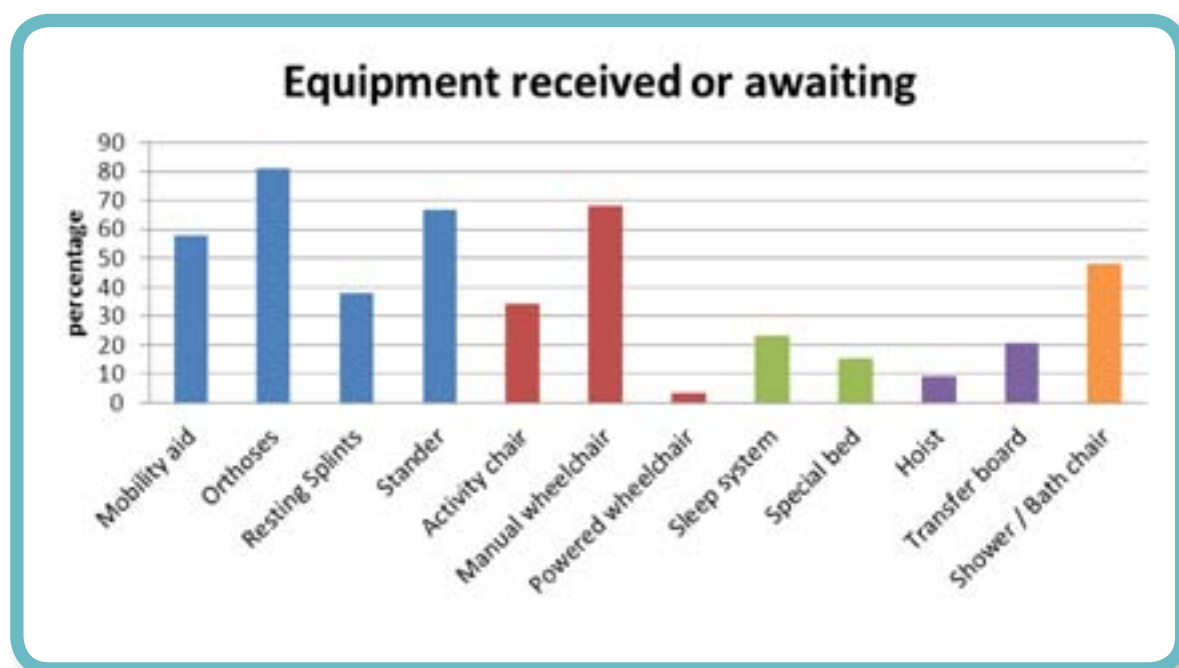
Sleep systems are used as part of a 24 hour postural management programme for certain children with neurological dysfunction to encourage aligned positioning at night time, and to protect the body from further secondary structural deformities such as scoliosis, pelvic obliquity

and lower limb contractures. Day time positioning such as appropriate seating, stretching and strengthening are also very important. Farley et al. (2003) described postural management as “the use of any technique to minimise postural abnormality and enhance function”; Wandel (2000) recommends early implementation to promote development. Postural care relies on the appropriate training of health care providers and parents as well as the availability of appropriate equipment which is often fragmented (Goldsmith, 2000, Humphreys and Pountney, 2006). The long term impact of postural management on outcomes within any of the components of the International Classification of Functioning, Disability and Health (ICF) requires further research (World Health Organisation, 2001) for children with complex movement disorders. However, their potential to improve the quality of life of these children may still be very valuable (Bacon, 2013).

## Results

Eighty three per cent of parents from our sample indicated that they have received equipment of some sort to support their child. The types of equipment that had been provided are listed in the Figure 28 below with the most frequently received pieces of equipment being those relating to assisting mobility - Orthoses (76%), standing frames (64%), manual wheelchairs (61%), and mobility aids (54%). Other frequently in use pieces of equipment are shower/bath chairs (40%) and activity chairs (29%). Only 18% of patients in our sample reported having a sleep system in place.

Figure 28



There were no statistical differences noted in the percentage receipt of equipment by geographical area.

Sixty one per cent of our patient sample reported having a manual wheelchair in place to support ambulation. Only 2% of the sample reported having a powered wheelchair. This may be due to the young age of the sample, poor social acceptance of this equipment, as well as limitations of funding for powered wheelchairs as described by a service provider:

*“I’d love her to have a new power chair, but that will have to be next year, there is somebody else waiting for 7 or 8 months for a power chair” (Service Provider)*

Sixty six per cent of patients within our sample reported having a stander in situ. Service providers commented on the benefits of standers in achieving upright standing as well as preventing contractures.

*‘But prevention is very important as well, so that you can prevent contractures first and then you can put child into the standing frame, or the standing position at the table even, in the age appropriate time. Some of them they are ready when they are aged 10 months, some 1yr and half, some two years. But I would try to follow the milestones.’ (Service Provider)*

In interview, the benefits of various pieces of specialised equipment were discussed and seen as a ‘vital adjuncts’ to meet the specific needs of these children. They also commented on the fact that these children often require specialist individualised equipment

*“I think that orthotics play a huge role in the management of a child with spina bifida and an evolving role as the child grows up so yes, it is a vital adjunct” (Service Provider)*

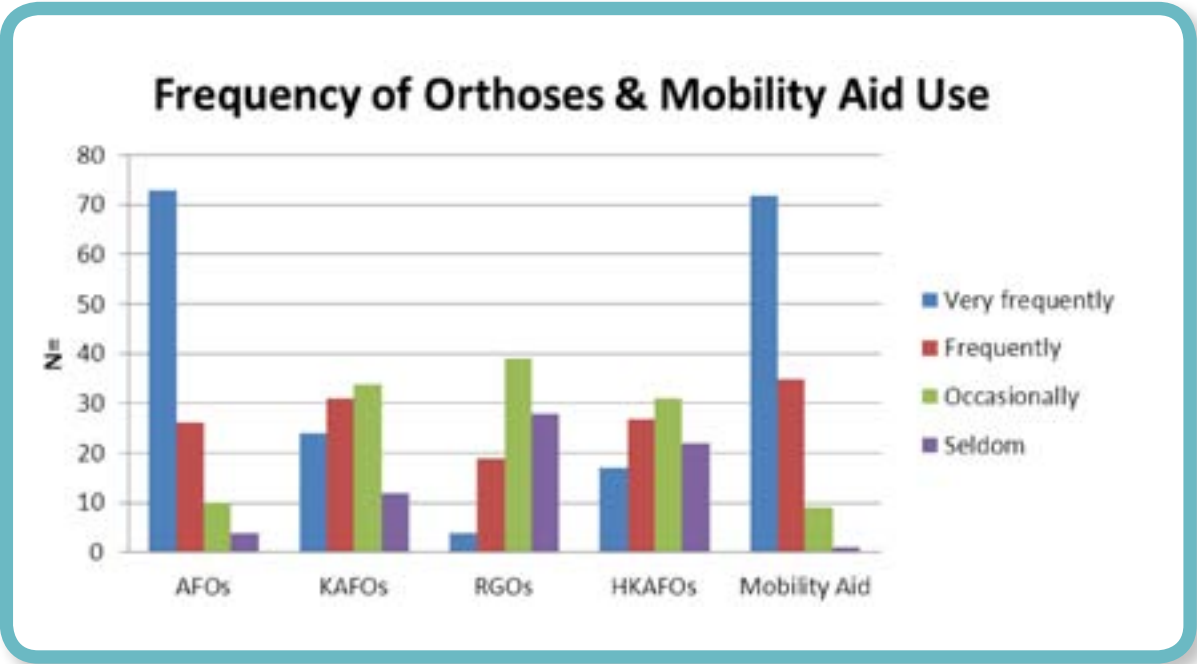
*“Often our children need these devices to meet their potential so definitely and it would be part of our guidelines that children are provided with walkers and standers as required, definitely” (Service Provider)*

*“They are vital, absolutely vital and it’s one of the biggest roles of OT” (Service Provider)*

*“The standardised equipment don’t suit kids with spina bifida”(Service Provider)*

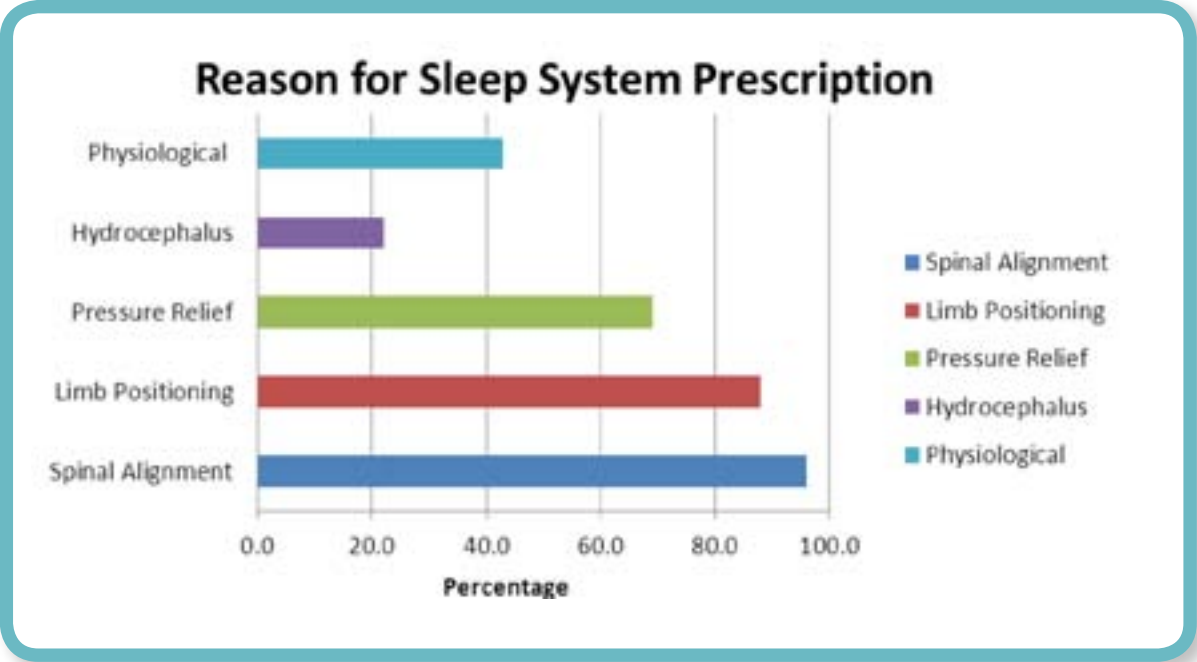
Service providers were asked in their professional opinion which service areas they would prioritise in relation to the management of a child with spina bifida. The most frequently reported area of high priority was mobility and equipment needs (96% of respondents indicated these being high priority areas). Service providers reported that AFOs were the most frequently used type of orthotic in their experience with 57% indicating they are used very frequently and RGOs the least frequently used (3%) (Figure 29). Mobility aids were also very frequently used according to 56% of service providers.

Figure 29



The most frequently reported reasons for prescribing sleep systems by Service Providers are spinal alignment (96%), limb positioning (88%) and pressure relief (69%) (Figure 30).

Figure 30



**Discussion**

The children in this sample had high equipment needs with a wide variation of equipment in place to support their needs. The most frequent pieces of equipment in place for children are orthoses, standing frames and wheelchairs. Service Providers also reported equipment to be a high priority for children with spina bifida. Ankle Foot Orthoses were the most frequently

used orthotic with the benefits of these being well supported in the literature. Thorough assessment based on the child's needs is required for more complex orthotic provision. Although equipment has been demonstrated to improve mobility and independence, the choice of equipment must be appropriate for the level of ability, lesion level and potential functional outcomes. A trained professional must complete a detailed functional and gait assessment including neurological function and orthopaedic deficiencies in order to recognize a child's potential to achieve functional ambulation and set realistic goals (Roussos et al., 2001, Bartonek and Saraste, 2001).

The details regarding the provision of equipment are discussed in Chapter 5.1 'Local Services'.

## **Recommendations**

1. Timely availability of a variety of generic and specialised equipment is required through local services to meet the needs of these children.
2. Experienced staff who are most involved in providing equipment, namely Occupational Therapists, Physiotherapists, Orthotists and Nurses require adequate training.

## ***BLADDER AND BOWEL IMPACT***

### **Background**

The majority of children with spina bifida present with bladder and bowel dysfunction due to incomplete innervation of these organs. This can result in bladder sphincter dysfunction, urinary tract infections, vesico-ureteral reflux, chronic constipation, urine and faecal incontinence and preventable renal damage if not appropriately managed (McDonnell and McCann, 2000, Filler et al., 2012, Dik et al., 2006, Bauer et al., 2012, Zegers et al., 2009). These are major causes of ill health in spina bifida patients (Cahill and Kiely, 2003).

This section discusses the impact of bladder and bowel difficulties on children with spina bifida with results relating to the investigation and interventions which are most frequently in place and also feedback from parents regarding concerns relating to social continence. Access to Urology and other professionals necessary to support bladder and bowel needs will be discussed further in chapter 5.2 Specialist Service (access to urology).

The main aims of bladder and bowel management are to maintain kidney function and promote social urine and faecal continence (Filler et al., 2012, Cahill and Kiely, 2003, Clayton et al., 2010, Zegers et al., 2009, de Jong et al., 2008). Early evaluation and intervention is required

for bladder and renal function, normalisation of bladder pressures to prevent secondary damage, prevention of urinary tract infections (UTIs) and lifelong surveillance (Merkens, 2006, Rawashdeh et al., 2012). Various interventions exist to support social continence for these children including Clean Intermittent Catheterisation (CIC), prophylactic antibiotics and surgery. However the timing and appropriateness of these interventions is debated within the literature (Cahill and Kiely, 2003, Zegers et al., 2009, Dik et al., 2006). CIC is a vital tool in the management of urinary continence with a success rate of up to 90% (Dik et al., 2006). A proactive and retrospective approach are discussed by Joseph (2008) regarding when to commence CIC with his preference being a proactive approach where all neonates are commenced on CIC until investigations are completed to identify those most at risk.

The practice of prophylactic antibiotic treatment for children undergoing clean intermittent catheterisation for neurogenic bladder is controversial in the literature. The International Children's Continence Society recommendations are that the use of prophylactic antibiotics does not reduce the rate of symptomatic UTIs (de Kort et al., 2012). The practice of antibiotic prophylaxis is still well documented in the literature with (Dik et al., 2006, Verhoef et al., 2005c) reporting that 30% of adult spina bifida patients were on continuous antibiotics and 62% had taken high dose antibiotics within the last year to manage a UTI.

Filler et al (2012) have recommended that routine early and repeated urodynamics (UDS) should form the basis of urological management of the neurogenic bladder. In a review on behalf of the International Children's Continence Society, Bauer et al. (2012) made the following four recommendations:

- Early evaluation during infancy by UDS to identify children with high bladder pressures.
- Renal ultrasound to identify the presence of hydronephrosis, ureteral dilation, a discrepancy in renal size or increased bladder wall thickness.
- As the child enters periods of rapid growth in the toddler and adolescent years, the risk of spinal cord tethering is high and therefore UDS should be repeated yearly or more frequently, especially if changes such as increased need for CIC, new onset wetting or recurrent UTIs occur.
- Once growth is completed ultrasound is recommended every 3 years and UDS only as required. In adolescent years into adulthood preservation of the upper urinary tract and maintaining renal function remains a priority.

(Bauer et al. 2012)

In a study by Verhoef et al. (2005c) reviewing incontinence in spina bifida patients aged 16-25 years, the prevalence of faecal incontinence was reported as 34% with double incontinence noted in 31% of patients. Risk factors for incontinence were presence of hydrocephalus and a lesion of L5 or above and the majority of patients with urinary or faecal incontinence perceived this as a problem impacting on their quality of life (Verhoef et al 2005c).

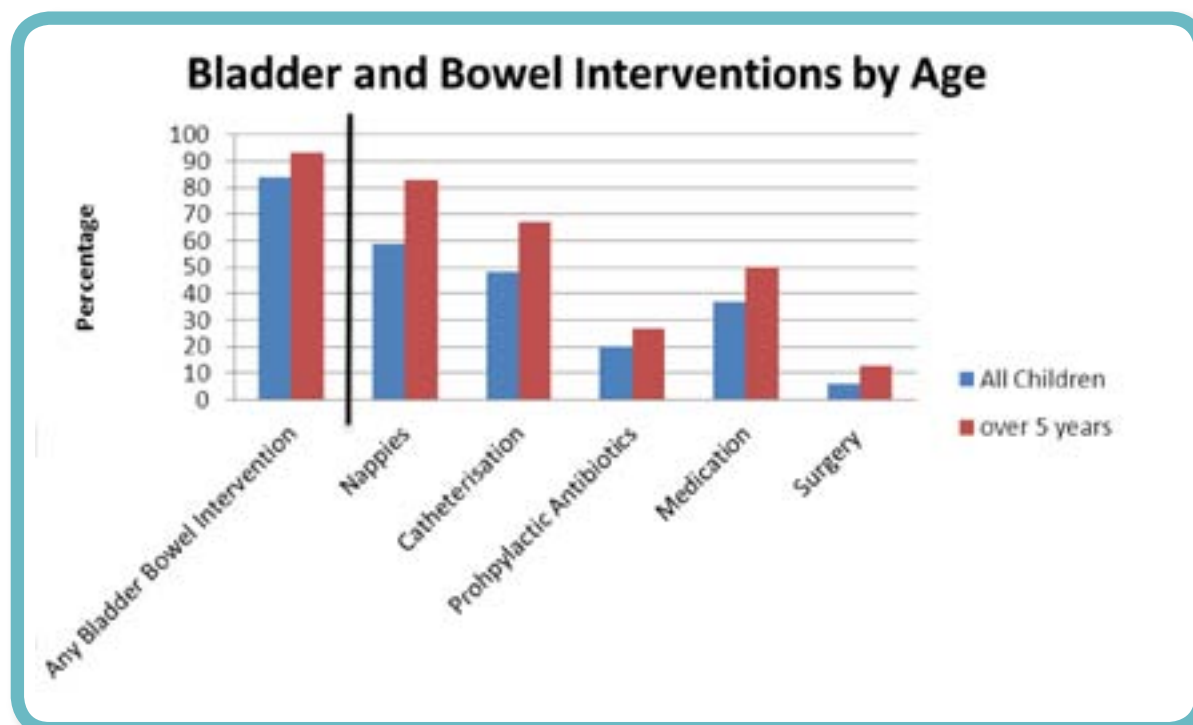
## Results

### Investigations and Interventions

In the current study 84% of all children required interventions to support their bladder and bowel needs, which increased to 93% for school age children (over 5 years). Overall 20% of children were taking prophylactic antibiotics, 48% were using CIC, and only 6% had undergone bladder or bowel surgery (Figure 31). The need for nappies to manage continence was high at 59% for all children.

As expected, the continence needs increase with age for all interventions. 67% of children were using CIC, 27% were on prophylactic antibiotics, 50% were taking medication such as anti-muscarinic medication, 13% had undergone surgery and 83% were still requiring nappies by school age to manage continence (Figure 31).

Figure 31



Service providers were asked which (if any) bladder or bowel continence management services they personally provide. Of those that responded, Paediatricians reported to have a role in the management of bladder and bowel needs, specifically, 67% were involved in ultrasounds, 60 % were involved in prescriptions for Urinary Tract Infections (UTIs), and



25% were involved in catheterisation along with support from clinical nurse specialists (25%) or other community nursing staff. Occupational Therapists were reported to be the most frequently involved professional regarding toilet adaptation advice (74%) and providing practical independent toilet training advice (48%). Clinical nurse specialists were most frequently involved in bowel washouts (29%)

Significant psychosocial impacts of bladder/bowel incontinence were raised by both parents and service providers as this can impact on participation in daily life and social interaction if not appropriately managed. The high prevalence of incontinence in the spina bifida patient group can have significant impact for both the family and child in terms of dependence and psychological well-being. Reported difficulties are embarrassment, social isolation, and impairment of activities of daily living. In school age children, when social continence is desirable, 83% still wear nappies / continence pads. Parents and service providers commented on the need for earlier bladder/bowel investigations and surgeries to ensure the bladder and bowel needs of the children are addressed in a timely manner. The significant impact of incontinence on social interactions and the child's self-esteem were also expressed.

*“It’s really the continence and bowel issues that really take over as the main problem” (Service Provider)*

*“My thing is now, is getting her ready for secondary school, I know it’s 2 years away but I want her dry and I want her clean, I don’t want her going in smelly and I mean children can be very hurtful” (Parent / Guardian)*

*“The problems for us are, the lack of access to specialists urology clinic for advice. And that particularly relates to children where you are concerned with regard to early intervention and want to address the problems quickly” (Service Provider)*

*“My aim is to get him off nappies. I thought I’d have him dry for school” (Parent / Guardian)*

## **Discussion**

The results indicate that the children with spina bifida have considerable bladder and bowel needs which require specialist professionals throughout their lifespan to evaluate and provide interventions to manage. The rates of bladder and bowel interventions are in line with international practice, however, earlier evaluation and intervention by specialist professionals is required to preserve kidneys and promote social continence. The rates of catheterisation compare to previous studies. Clayton et al. (2010) and Verhoef et al. (2005c) reported that

65% of spina bifida patients in a large study were performing CIC which is comparable to our rates of 67% in over 5 year olds. Although early commencement of CIC during infancy is recommended (Dik et al., 2006, de Kort et al., 2012), our rates increased with age which does not reflect early implementation. As mentioned above, the use of prophylactic antibiotics to prevent UTIs are debated in the literature, and our use of antibiotics is lower than previous studies. The very small numbers of children who have undergone surgery may reflect the limited access to specialist Urological services which are discussed in chapter 5.2 'Specialist Services'. Overall, there remains a high number of children using nappies / continence pads to manage continence needs which has impacts in terms of social participation and self-esteem for older children. The impact of incontinence on self-esteem and quality of life has been reported in the literature (Verhoef et al 2005c), this study identified similar negative impacts of incontinence of childrens lives. Improved consultation with parents, children and professionals is required to achieve satisfactory outcomes for social continence for children with spina bifida.

## **Recommendation**

1. Appropriate interventions, implemented earlier in the child's life, need to be in place to support difficulties, protect renal function and to assist with achieving social continence.

## **GROWTH AND NUTRITION**

### **Background**

Concerns regarding the significance of obesity in the spina bifida population have been reported as early as the 1970s by Hayes-Allen and Tring (1973) who noted significantly increased skin fold thickness measurements. The prevalence of obesity in spina bifida has been reported to range from 18% to as high as 58% (Dosa et al., 2009a, Dosa et al., 2009b, Mita et al., 1993, Buffart et al., 2008a). Compared with typically developing peers, children with disabilities are up to three times more likely to be overweight and up six times more likely to be obese (Neter et al., 2011).

### **The physical and psychosocial effects of obesity in children with spina bifida:**

The impact of obesity on this already vulnerable population is significant. As children born with spina bifida can now expect to survive well into adulthood (Bowman et al., 2001), health promotion is important to optimise health in adult life. Inactivity, obesity and reduced physical fitness can increase the likelihood of secondary conditions including hypertension and depression (Buffart et al., 2009 Reinehr et al., 2010). The combination of these factors can

compound the difficulties that these children already face, placing an unnecessary additional burden which is largely avoidable if detected early. As well as physical complications of obesity, there is also a significant impact of obesity reported on the emotional wellbeing and quality of life of children and adolescents (Reinehr et al., 2010). Able bodied adolescents with obesity have reported quality of life lower than that of children with complex medical conditions (cancer, Juvenile Rheumatoid Arthritis, Diabetes Mellitus, and congenital heart disease). Children with spina bifida reported a negative quality of life compared with healthy peers, however the added presence of obesity did not further impact their low quality of life (Abresch et al., 2007). A disease specific QOL tool may be necessary for this population to determine the specific impact of obesity.

### **Aetiology of obesity in children with spina bifida:**

The reasons for high rates of obesity in the spina bifida population have been explained in relation to activity, lifestyle and diet. Children with spina bifida participate significantly less in physical activity than healthy comparison peers (van den Berg-Emons et al., 2001, Dosa et al., 2009b) with up 70% not partaking in any organised sports (Neter et al., 2011). Buffart et al. (2008a) analysed physical activity participation among adolescents with spina bifida using activity monitors and reported that only 81 minutes per 24 hours was spent on physical activity which is significantly less than the time spent by able-bodied peers (163 mins). Persons with higher level of ambulation were more likely to be physically active, have higher aerobic fitness and less body fat. van den Berg-Emons et al. (2003) agreed that aerobic fitness was 20-35% lower in a spina bifida group than healthy peers. Daily wheelchair propulsion is insufficiently strenuous to elicit cardiovascular or muscle adaption in adolescents (Widman et al., 2007). Barriers and facilitators to participation in physical activity were explored by Buffart et al. (2009) who identified barriers such as motivation similar to able-bodied peers, however fatigue, lack of energy, injuries, complications associated with disability and fear of injury were identified as specific barriers for adolescents and young adults with spina bifida to participate in physical activity. A lack of information and professional support were also discussed. As well as less physical activity, children with spina bifida lead a more sedentary lifestyle and spend more time watching TV or playing computers than their able bodied peers (Reinehr et al., 2010).

Diet is also an area of concern and significant differences have been noted in the dietary habits of children with disability. Neter et al (2011) found that children with disabilities are significantly more likely to miss breakfast, consume more sugary drinks and less likely to eat fruit than peers who are non-disabled. Vega-Sanchez et al (2012) highly recommends personalised nutritional counselling as one of the first strategies in the management of children with neuromuscular disorders due to their known risks for weight gain. Buffart et al. (2008a) did not find any correlation between physical fitness or body fat to participation in physical activity, therefore suggesting that strategies such as reducing energy intake may be more important in preventing obesity in this population group than increasing physical activity or

fitness.

Some studies have noted that obesity increases with age and female gender, and can be influenced by non-ambulatory status and reduced physical activity (Dosa et al., 2009, CDC 2005, Buffart et al., 2008a, Roberts et al., 1991). Inactivity due to motor impairment can have an impact on weight gain and obesity. However, in a review of 203 patients with spina bifida (including 34 children) Dosa et al., (2009) noted the highest rates of obesity occur in sacral level lesions, however this result was not statistically significant. High obesity levels can however occur even in the most physically able category. Their study saw only a slightly higher prevalence of obesity in spina bifida compared to typical population, suggesting that the increasing rate of obesity in the US population is 'catching up' with high obesity rates in disability.

### **Classifying obesity in children with spina bifida**

When calculating BMI both the weight and height are considered. The specific profile and body composition of children with spina bifida pose some challenges to the accuracy of measurement of height and weight (Shepherd et al., 1991). Numerous studies have demonstrated the significant short stature that exists in the spina bifida population which needs special consideration (Widman et al., 2007, Roberts et al., 1991). Charney et al. (1981) noted that 51% of all children with myelomeningocele had a body length of less than the 3rd percentile for their age. This rose to 77% in children with lesions above L3, whereas no children with sacral lesions were of short stature. The reasons for short stature include less developed lower limbs and associated musculoskeletal deformities of the spine, hips, and knees and occasionally as an impact of hydrocephalus and growth hormone secretion from the Pituitary gland. Duval-Beaupère et al. (1987) agreed that children with myelomeningocele can have growth disturbances with the higher the level of the lesion, the greater the expected growth defects. The use of height as an indicator of growth is therefore questioned in spina bifida and many researchers have proposed the use of arm span as a valuable growth parameter, (Shurtleff et al., 2010, Roberts et al., 1991). Accurately measuring height in children with high-level lesions with severe scoliosis may be challenging and result in artificially excessive BMI. Such issues should be considered when interpreting growth development in such children. However, Shurtleff et al. (2010), concluded that the BMI Centre of Disease Control percentiles are appropriate for use in the spina bifida population.

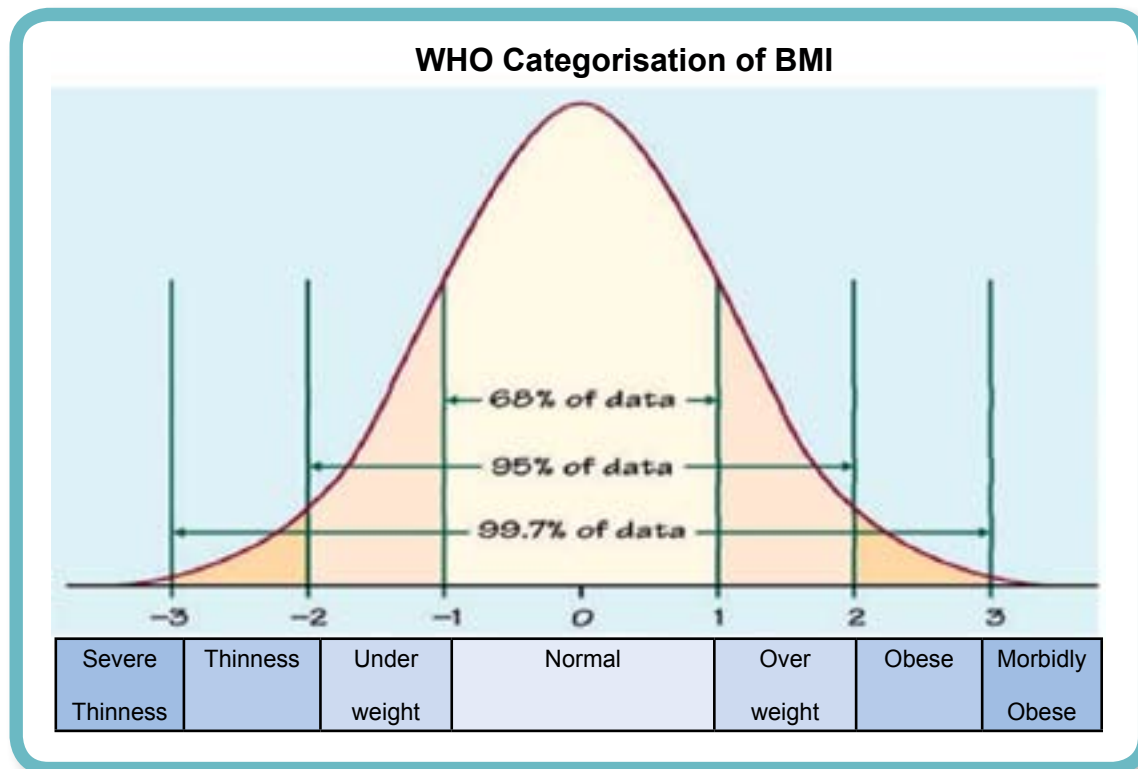
## **Results**

### **Anthropometric data**

Parent / Guardians reported their child's height and weight within the questionnaire. The weight, height, BMI, BMI centiles and BMI Z-scores were calculated using LMS growth software. UK growth curves classify a child as overweight if BMI is >91st centile or if BMI

z-score is between 1 and 1.9. Obesity is classified as a BMI >97th centile and a BMI z-score >2.0 (Figure 32). The BMI z score is a measure of relative weight adjusted for child age and sex, and is a recommended outcome measure used to estimate and monitor overweight and obesity over time (Rolland-Cachera, 2011).

Figure 32



There was complete data available for 79 children to calculate BMI. The mean BMI was 19.8 kg / m<sup>2</sup>. The mean BMI centile was 73.6 with a mean standard deviation score of 1.2.

Table 4

#### Anthropometric Data

	N	Minimum	Maximum	Mean	Std. Deviation
Age Total Years	153	0.2	18.0	5.7	4.7
Weight (Zimmet et al.)	97	5.2	80.0	22.2	15.7
Height (cm)	86	56.0	180.3	102.3	30.1
BMI	79	11.3	39.7	19.8	5.0
Weight centile	94	0.0	100.0	50.9	37.4
Height centile	82	0.0	100.0	27.6	33.4
BMI Z Score	77	-4.8	5.2	1.2	1.8
BMI Centile	77	0.0	100.00	73.6	31.5

## Classification of obesity categories:

When classified according to z-scores, 30% of our sample population was obese, with 6.5% of these being morbidly obese. A total of 56% were either overweight or obese. When compared to the Growing up in Ireland study (Department of Health and Children, 2011) which is following the progress of almost 20,000 children across Ireland, there are significant differences. The 9 year old cohort was chosen as comparison as it best matched our samples age range. In the typically developing child sample in Ireland, 6% presented as obese with 25% of children being either overweight or obese. Therefore our sample were more than twice as likely to be overweight or obese, and 5 times more likely to be obese than typically developing children.

Figure 33

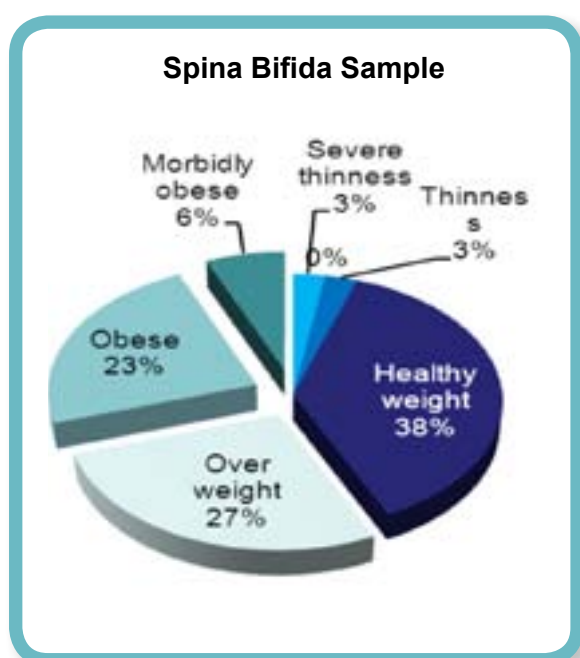
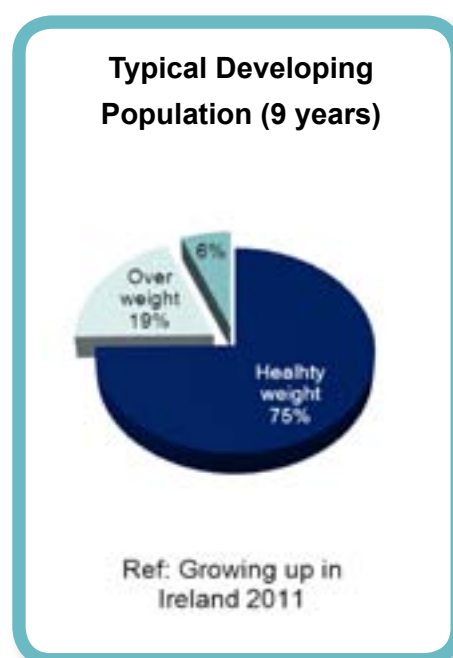


Figure 34

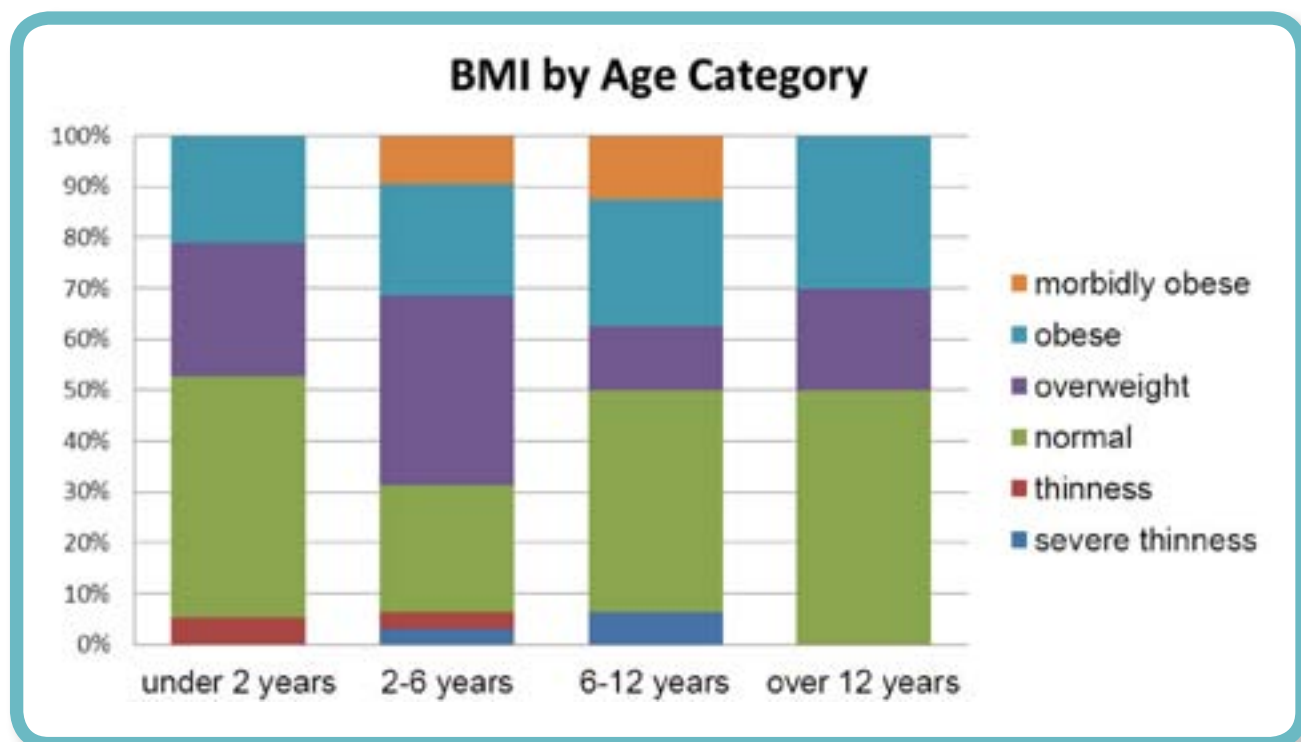


## BMI by age category:

The sample in this study was divided by age categories (under 2 years, 2-6 years, 6-12 years and over 12 years) to determine differences between age ranges. No significant difference was noted in the BMI z-scores across age ranges. The majority (69%) of 2-6 year olds were overweight, obese or morbidly obese. A large proportion of children aged 6-12 years (12.5%) fell into the morbidly obese range (figure 35).

Raw height and weight increased significantly across all age groups ( $p=0.00$ ) as expected in growing children. However, of note, there was a less significant increase in height noted between the 6-12 age group and the 12+ age group ( $p=0.027$ ) indicating a potential height plateau for children over 12 years which is in line with the onset of puberty.

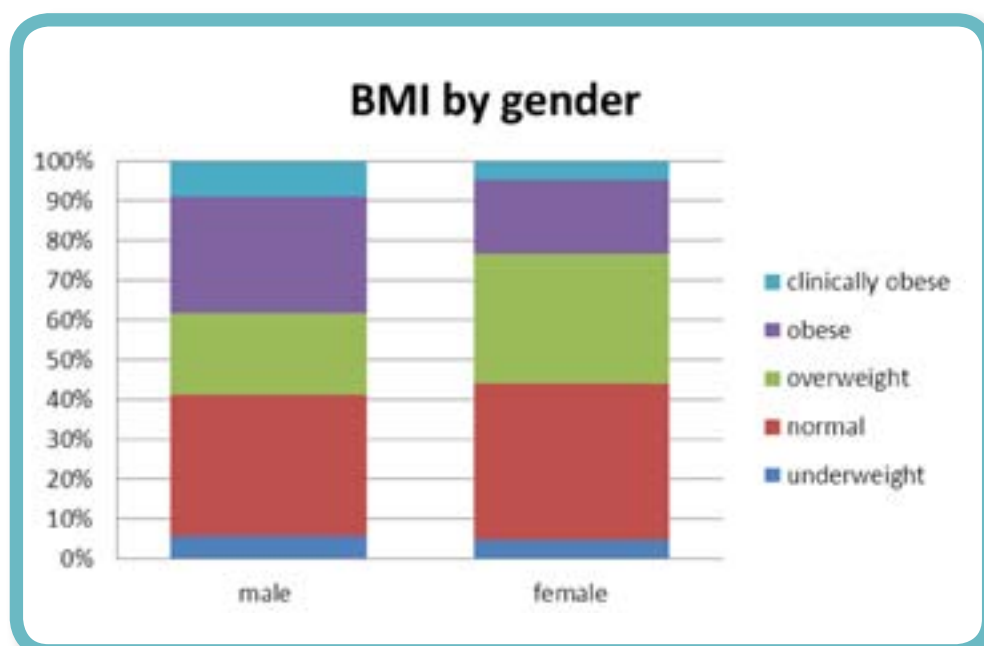
Figure 35



### BMI by gender:

No significant differences were noted in the BMI z-score categories by gender suggesting there is no increased risk of obesity by gender.

Figure 36

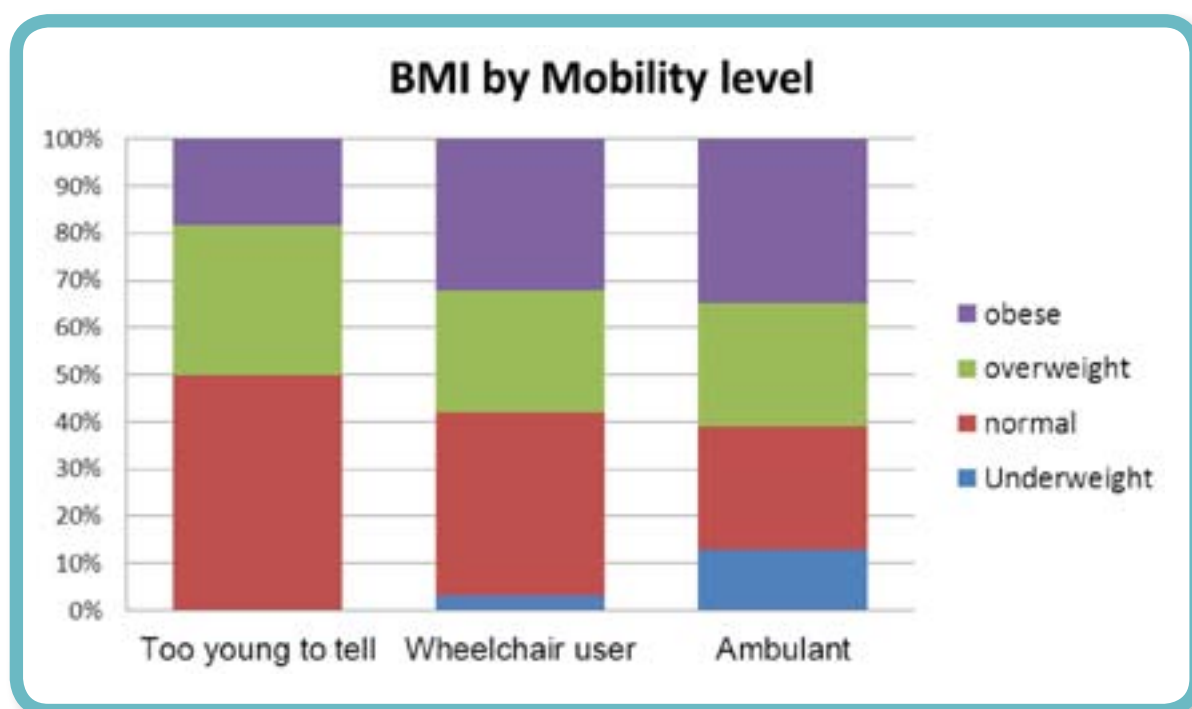


### BMI by mobility level:

Children in our sample were categorised by mobility level (wheelchair user, ambulant and those that were too young to determine their mobility level) and BMI z-scores were compared across groups to determine correlation.

Over 50% of all groups were in the overweight / obese range. Although no significant differences were noted between the ambulant and non-ambulant group, the BMI in wheelchair users (21.46) was higher than the ambulators (18.86) indicating a trend towards a higher BMI in wheelchair users than in the ambulant category.

Figure 37

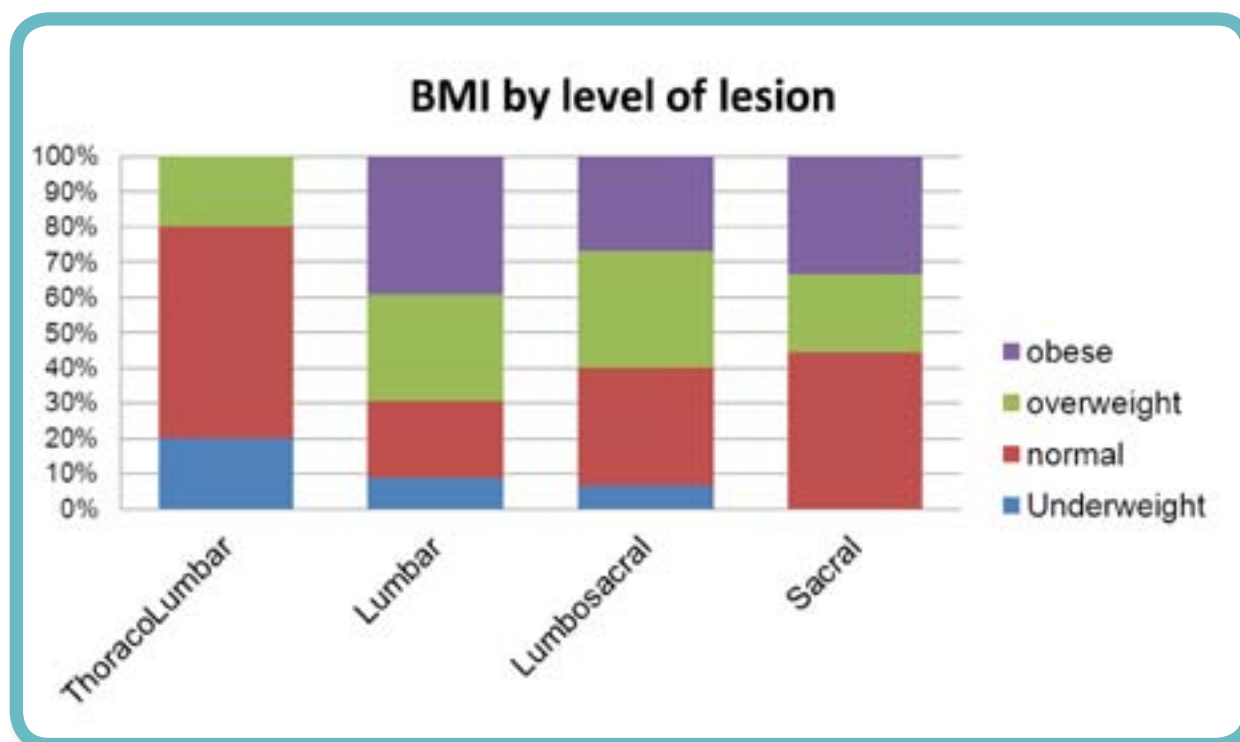


### BMI by level of lesion:

When children were divided into levels of lesion to determine correlations, again we saw no significant differences between groups. Of note, however nearly 70% of children with lumbar lesions were overweight or obese. It was surprising that there were no children who were obese in the thoracolumbar population; reasons for this are unclear however the small sample numbers in this group may have influenced results.



Figure 38



### Surveillance and Intervention

Due to the high prevalence of overweight and obesity within our sample, the surveillance and interventions were investigated. Parents were asked how often their child's height and weight were being monitored. They reported that although height and weight were measured relatively frequently when they were infants and preschool aged, the height and weight for children over 2 years was most frequently measured 56% of the time annually or less than once a year. Fifty six per cent of children were measured for height once a year or less, and 49% of children were measured for weight once a year or less.

Only 16% of the service providers questioned reported being involved in the monitoring of height and weight of children with spina bifida with Paediatricians and Physiotherapists being those who reported being involved most frequently. Of those who were involved with height and weight monitoring, height appears to be most frequently measured between every 6 months and 1 year whereas weight appears to be measured less than once a year, which correlates with the parent reporting as outlined below. Unfortunately the current service provider sample did not consist of many nursing professionals who would also be expected to be involved in this process.

Figure 39

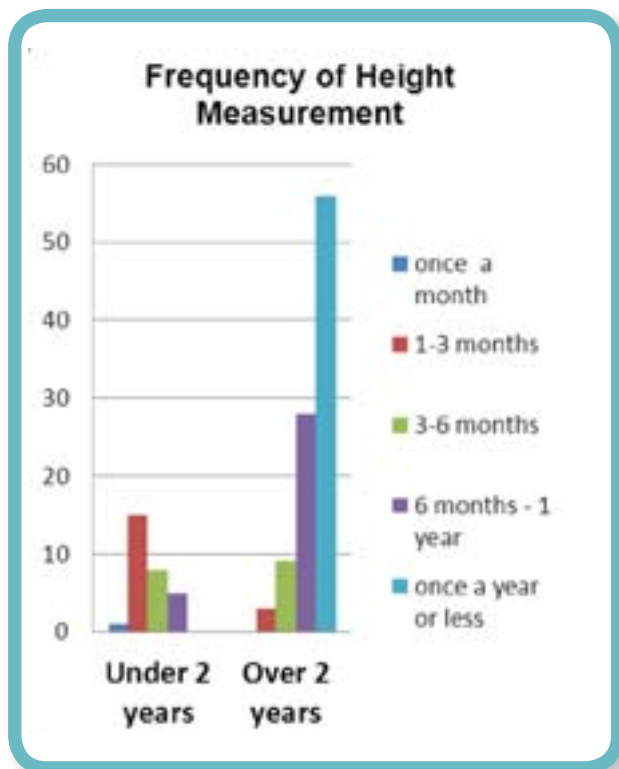
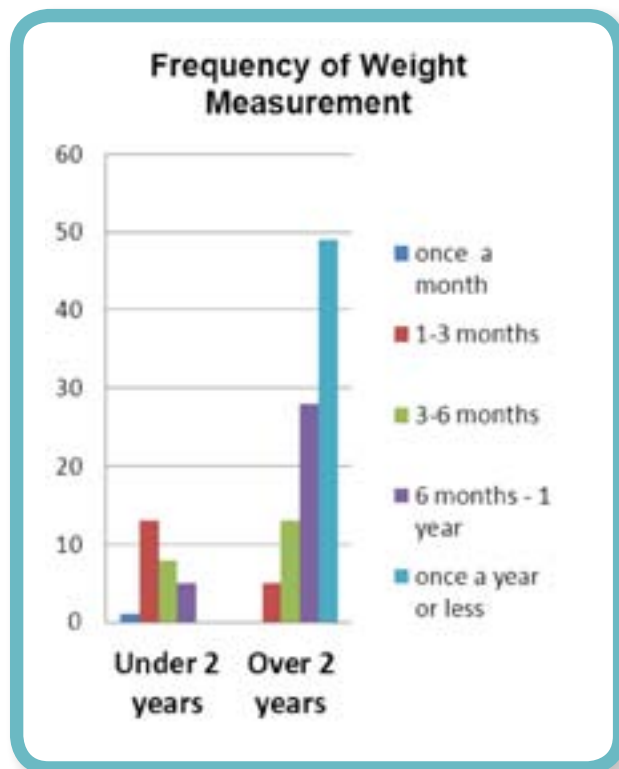
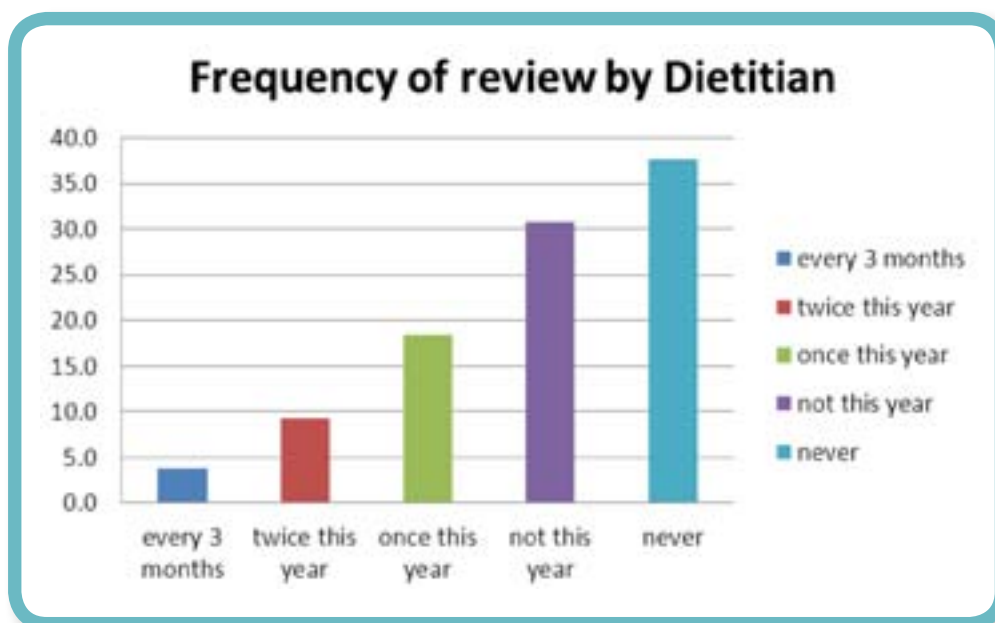


Figure 40



Dietitians are key professionals necessary to be involved for weight or diet related issues. Despite the fact that children with disabilities including spina bifida can be seen to be at high risk for obesity in the overall sample 68% (n=89) of parents reported that their child had either never been reviewed by a dietitian or had not seen one within the previous year.

Figure 41



Parents of overweight, obese and morbidly obese children reported poor access to regular dietitian review with nearly a third of overweight or obese children never accessing a dietitian (see table 5).

Table 5

Frequency of dietitian review by weight category

	Normal	Overweight (%)	Obese and morbidly obese (%)
Every 3 months	0.0	5.3	0.0
Twice this year	0.0	5.3	23.5
Once this year	25.9	10.5	23.5
Not this year	18.5	47.4	23.5
Never	55.6	31.6	29.4

Many additional diet and bowel specific difficulties for children with spina bifida exist such as reduced gastrointestinal motility, reduced fibre intake secondary to swallowing difficulties, sphincter dysfunction, abnormal rectal and anal sensation, faecal incontinence, and altered hormones (e.g., precocious puberty and decreased growth hormone) (Liptak and El Samra, 2010). All of these factors increase the risk for developing weight and nutrition related difficulties which require specialist diet advice to manage.

### Qualitative:

Service providers reported on weight gain as being a barrier for participation as well as having a negative impact on the child's self-esteem.

*“Another thing I think is a big impact is the weight gain on self-esteem, but also on ability to engage with activities. And that's one of the biggest issues that need to be addressed when the kids are very young.” (Service Provider)*

Connor-Kuntz et al. (1995) reviewed the participation of 133 youth with spina bifida in sporting activities. He reported that 90% of subjects received physical education at school, and that 83% participated in sport. Of note, children who walked without assistive devices were least likely to participate in non-school sports. The extremely positive impact of sports and extracurricular activities was reported by parents and service providers. Service providers felt that the benefits of sport and exercise need to be emphasised to families to help avoid future mental health problems. Issues in relation to access and availability of suitable exercise facilities, funding cuts and restrictions for previous existing facilities and lack of acceptance / inclusion into mainstream sports were the main barriers for participation.

*“I suppose medically things have improved a lot and educationally things have improved a lot. I suppose just looking at community involvements, sports and recreational activities, the whole kind of confidence building for a child and the alternatives for them, But we would find that it’s a challenge”. (Service Provider)*

*“He was thrilled to find something that he was good at because school, you know, he was never up there with school”. (Parent / Guardian)*

*“It’s a challenge to find a team or a club or something for him that he can be a part of or accepted as part of”. (Parent / Guardian)*

*“He is very good at racing and he likes wheelchair basketball and he likes wheelchair racing and hockey but we don’t have the services locally”. (Service Provider)*

*“Sometimes, she’s not able to do it, and I see her getting down, mammy I can’t do it. But you talk her through it and say, god you might not be able to do this, but you can do that.” (Parent / Guardian)*

*“The school runs summer camp and there is Gaelic, hurling and soccer, but she is still stuck in the middle. And they do put in, they have great time for her, they spend a lot of time with her, and they get her involved. Gone are the days that they were left behind the door not seen, and now they are out. Then she’d ask, would I be able to do it, I say, why not? I said it’s just going to take a different way around it”. (Parent / Guardian)*

*“If the kids are involved in activities that they can master and they can enjoy, then you avoid some of the mental health issues and then you address some of the weight issues as well”. (Service Provider)*

## Discussion

Overall, high levels of obesity were noted in spina bifida which may be due to a combination of disease specific, physical and social factors. Obesity further impacts the physical and emotional wellbeing of a child with spina bifida and complicates their medical care. Obesity levels were not related to age, gender, mobility status or level of lesion in this group. There were small numbers of children aged over 12 years when height plateaued. It would therefore be worth repeating analysis with an older cohort to determine whether this influences BMI and obesity. Although wheelchair users did not have a statistically greater BMI as noted in previous literature (van den Berg-Emons et al., 2003), there was a trend to suggest this may be an influencing factor. The mobility level as determined by the questionnaire made it difficult to divide the sample into 'ambulant' and 'wheelchair user', therefore all children who mobilized to some extent were categorized as 'ambulant' even if this was non-community ambulation. This may have impacted the results and would warrant further investigation with more structured mobility categories.

The regular monitoring of height, weight, BMI and physical engagement by medical professionals has been highlighted as infrequent but extremely necessary (Dosa et al., 2009b). If feasible, professionals most frequently involved with these children should be trained to measure height and weight and calculate BMI Centiles and Z scores. This could include Paediatricians, Physiotherapists, Occupational Therapists, and primary care physicians including GPs and Public Health Nurses. To reduce excessive burden on services, parents should be educated on how to appropriately measure their children and to record information in a patient passport which will be brought to each appointment. Alternatively electronic growth monitoring tools would be useful to parents, patients and care providers. Such a tool could feed into a national growth monitoring initiative as recommended by The Report of the National Taskforce on Obesity (2005) to develop a national database of growth measurements.

It is also recommended that support be given to parents and children to overcome barriers to participation in sport (McPherson et al., 2013). Savoye et al. (2011) reviewed the benefits of a weight management programme in complex paediatric populations and results indicated that education, behaviour modification and exercise training had a long lasting effect on the food choices and physical activity at 12 month and 24 month follow up. Increased access to and availability of accessible sports and opportunities for aerobic exercise for children with limited mobility is required.

Access to dietitian was highlighted as an area of concern with a third of overweight and obese children having never seen a dietitian. Due to limitations in availability of dietitians in local teams, access to dietitians on a regional basis should be made available for children with spina bifida to access. Our results highlight that all professionals within the multidisciplinary teams must be aware of the increased risk of obesity in children with spina bifida and that these issues be discussed with and educated to the children and their families.

## Recommendations

1. It is recommended that height, weight and BMI (Centiles and Z score) of these children are measured at a minimum every 6 months by local professionals most frequently involved in the care of the child.
2. A multidisciplinary approach to prevention and management of increased weight is required specifically for children with spina bifida due to their increased risk. Parents, families and professionals should be educated about specific nutritional difficulties of children with spina bifida, be given advice on healthy eating, active living and promoting a positive self-image. This should be discussed at annual review at a spina bifida MDT Clinic.
3. Children with spina bifida who are overweight / obese should have equitable access nationally to a dietitian for assessment and intervention.

## ***VISUAL DIFFICULTIES***

### **Background:**

Children with spina bifida and hydrocephalus have been reported to have difficulties with visual skills and in particular visual perceptual skills (Dennis et al., 2001). People use vision in their day-to-day lives for three principal activities: visually guided movement, access to information in their environment, and social interaction.

Visual perception is an intricate system that is concerned with both object identity and localization in space, and is closely connected with action systems (Jeannerod, 2006). Visual perceptual difficulties can impact on visual discrimination (differentiating between shape, orientation, size and colour), visual memory and visual sequential memory (remembering the characteristics of a picture over time and recalling them as seen), visual spatial relations (determining where things are in space), visual form constancy (recognising shapes, sizes, textures, and their positions or orientation in space), visual figure ground (distinguishing an object from irrelevant background information) and visual closure (recognising an incomplete object) (Gardner, 1996)

As children with spina bifida have limited mobility, they don't have a chance to explore their environment in the early years in the same way as a typically developing child. The visual perceptual difficulties experienced by children with spina bifida have been reported to be due

to limited upper limb skills as well as limited opportunities for visuo-motor learning due to restricted mobility (Hetherington et al., 2006, Simms, 1987). While much is understood about the basic aspects of visual function, such as visual acuity, visual fields, and colour vision, the nature and development of higher visual function and its disorders are more enigmatic, particularly in the developing child (Houliston et al., 1999). Visual difficulties can be noted as early as 18 months of age (Taylor et al., 2010).

Cortical Visual Impairment (CVI) has been described as one of the most common causes of visual impairment in children in the developed world. Philip and Dutton (2014) described CVI as 'Visual brain damage', impairment of the visual pathways and the pathways serving the higher visual functions. Visual impairment arising from perceptual and cognitive visual dysfunction is due to damage to the areas of the brain that are responsible for higher visual processing (Lam et al., 2010). Children with CVI often have typically functioning optic tracts, but they have damage to areas of the brain related to vision which can negatively affect their sense of safety, initiation, engagement in the environment, feelings of competence, satisfaction and overall development (Shaman, 2009). There are multiple causes of CVI including prematurity, hypoxic–ischemic damage, neonatal hypoglycaemia, metabolic disease, central nervous system infection, stroke, traumatic brain injury, structural brain anomalies, hydrocephalus, chromosomal aberrations and seizures (Lehman, 2012). Within spina bifida populations, about 25 per cent have mildly impaired vision, while 10 to 15 per cent have low visual acuities (Philip and Dutton, 2014). These children have difficulty extracting information from crowded visual scenes, lack visual attention, and have inaccurate visual guidance of motion of their lower and upper limbs (Dutton et al., 2004). In a large study of over 3000 children referred to the Brazilian Association for the Visually Impaired People, cortical visual impairment accounted for 20% of those with multiple disabilities and was attributed to perinatal hypoxic–ischemic disturbances (46.2%), malformation of the CNS (12.8%), and meningitis (11.0%) (Haddad et al., 2007).

Each child with CVI is unique but Shaman (2009) highlighted two main types of CVI including:

1. Lower level CVI which is caused by damage to the visual pathways up to and including the striate cortex. This affects acuity, understanding of the visual image and ability to see various parts of the visual field, this can vary in severity.
2. Higher level CVI (sometimes called cognitive visual impairment) which refers to damage occurring beyond the striate cortex. This is a less severe disruption of specific functions of vision (movement awareness, shape or colour recognition) but not of visual field or acuity.

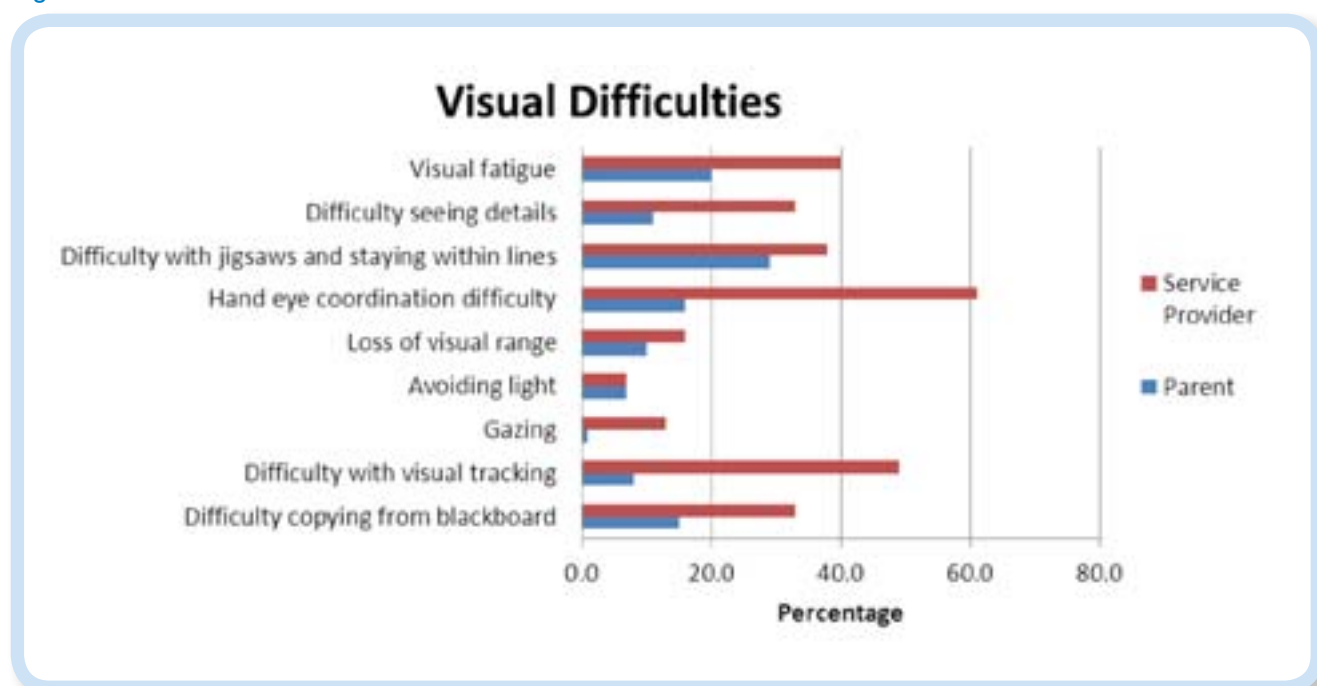
Houliston et al. (1999) reported that half of the children with spina bifida and shunted hydrocephalus had symptoms of higher level CVI including problems with shape recognition, simultaneous perception, perception of movement, colour, shape, object and face recognition and orientation difficulties.

Assessment and interventions for CVI are complex and evolving. It is vital that the evaluation focuses on the interaction between the child, the activity and their environment and team collaboration is essential to develop a programme (Shaman, 2009). Members of the team working with children with CVI could include parents, ophthalmologists, Occupational Therapists, neuropsychologists, specialist teachers, paediatricians and nurses (Philip and Dutton, 2014, Shaman, 2009). Haddad et al. (2007) report that children with a CVI require a long, complex, and costly process of rehabilitation.

## Results:

A third of parents (36%) indicated that their child presented with a visual difficulty, although this figure increased to 55% of parents of children 5 years or older. The ranges of visual difficulties are represented in the chart below, with the most common difficulty reported by parents being with jigsaws and staying within lines (reported by 29% of parents), followed by visual fatigue (20%), hand-eye coordination difficulties (16%) and difficulties copying from the blackboard (15%).

Figure 42



A higher proportion of service providers (52%) reported the presence some kind of visual difficulty among the children they review which is comparable to the parents of children over 5 years. Among those that were questioned, hand-eye co-ordination was the most frequently reported difficulty (61% of service providers felt this was an area of difficulty), followed by difficulties with visual tracking (49%). They reported a much higher prevalence of all difficulties than parents, agreeing that children had difficulties completing jigsaws or staying inside the lines (38%) and suffered with visual fatigue (40%)



Qualitatively, service providers also frequently commented on this being a part of their intervention and difficulties may be explained by the delay in motor milestones

*“My main area visual motor development” (Service Provider)*

*“These children don’t really do a lot of their earlier milestones like crawling and everything which kind of impacts on visual perception” (Service Provider)*

The area of visual assessment and diagnosis particularly for more significant difficulties such as cortical visual impairment is an area of concern for local service providers who do not have access to the appropriate expertise to diagnose and recommend strategies to manage this condition. Specialists with a knowledge in diagnoses as well as guidance in the management of these difficulties was sought frequently by professionals.

*“I suppose CVI is a particularly difficult one, I think that’s a gap...the whole management of CVI I’m finding difficult to get to negotiate children through that process. I’ve actually asked on a number of occasions could we have a functional visual assessment done and I’m not able to get that locally so again some either training for the local services if they could do it” (Service Provider)*

*“CVI is definitely something we are looking into more and it’s coming more prevalent” (Service Provider)*

*“I would love to learn more about (visual difficulties) and to find out if there is somewhere that we can refer them onto, where these kind of kids can be helped” (Service Provider)*

*“It is a minefield, when assessing a child, to see could they actually have something like CVI, or any other visual impairment” (Service Provider)*

## Discussion

Overall, service providers reported visual difficulties as a more prevalent difficulty than the parents themselves. Although these are reported to be relevant in many children according to the service providers, there are limited specialists who can provide assessments and interventions. The parents may also benefit from education and awareness of this aspect of physical need. Houliston et al. (1999) and Philip and Dutton, (2014) developed a structured history taking questionnaire to aid with the diagnosis of visual difficulties and recommend referral to an interdisciplinary team for assessment. The earlier that deficits can be detected, and measures implemented to circumvent them, the greater the potential opportunity for functional improvement. Lam et al. (2010) described a model developed in Glasgow to assist children with CVI. Practical adaptations to everyday life such as using tactile stimuli, verbal and visual cues maximize the use of intact alternative sensory pathways. Simplifying the environment, avoiding clutter and change, using large simple fonts and introducing yourself by name can all help to reduce the difficulties facing these children.

There is a need for a specialist assessment, diagnostics and support service for children with corticovisual impairment within an Irish context. Intervention programmes can be performed by local therapists to help improve understanding of this complex problem and to assist with improve functioning of these children. This could consist of a Neuropsychologist, Occupational Therapist, Ophthalmologist and /or Paediatrician

## Recommendations

1. A comprehensive functional visual assessment should be available to all children with spina bifida who present with visual difficulties.
2. This could occur in conjunction with the TSCUH MDT spina bifida service but would require extra resources and staff in the area of ophthalmology, occupational therapy and neuropsychology.

## ***SPEECH AND LANGUAGE DIFFICULTIES***

### **Background:**

Children with spina bifida may present with speech and language that on the surface appears to be intact. Fletcher et al. (2002) noted that at the deeper level children with spina bifida present with language difficulties that can combine to cause difficulties in the academic setting and social difficulties. Poorer outcomes in adulthood have been reported if the aforementioned difficulties are not pre-empted or mitigated by early intervention. Economic disadvantage has significant negative impact on language function of children with spina bifida. Children with spina bifida present with specific speech and language difficulties which can impact on function, these difficulties can be related to both congenital malformations of the cerebellum and corpus callosum and are also influenced by the presence of hydrocephalus (Dennis et al., 2008, Huber-Okrainec et al., 2002, Fletcher et al., 2002).

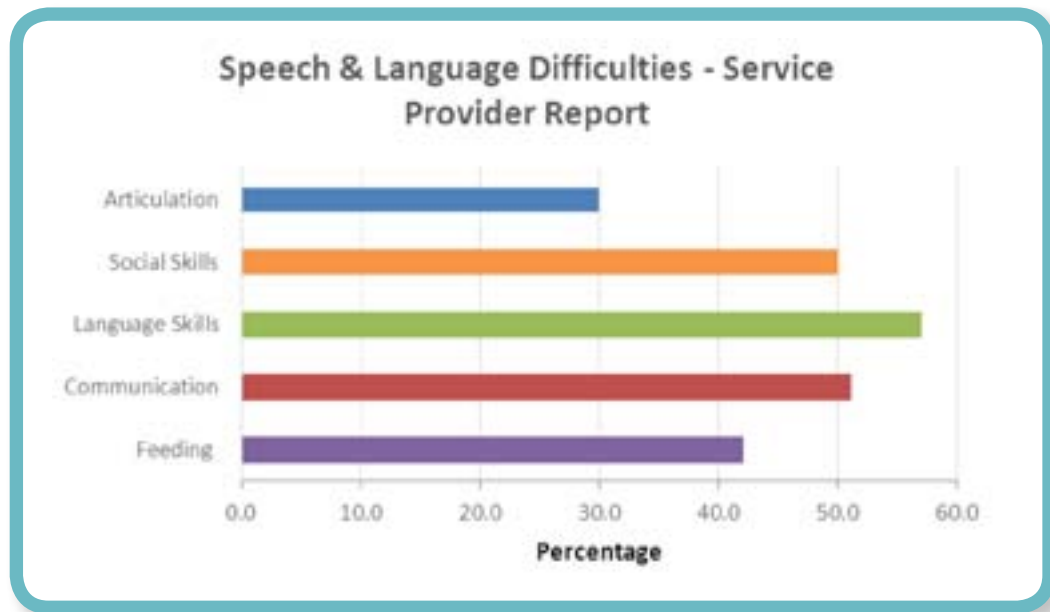
Although verbal IQ can be high, performance IQ tends to be low indicating difficulties with comprehension of language (Fletcher et al., 2002). Huber-Okrainec et al. (2002) concluded that individuals with spina bifida were more dysfluent, had more ataxic dysarthric features, and had a slower speech rate than aged matched peers, which, singly or in combination, have a negative impact on speech intelligibility. Adequate performance may be identified on specific vocabulary measures but deficiencies may be present on less structured language tasks. Children with spina bifida may find it challenging to make sense of language and the social use of language. Fletcher et al. (2002) also reported that children with spina bifida demonstrate more pervasive impairments of language.

Lomax-Bream et al. (2007) indicated that the kinds of cognitive (inclusive of language) difficulties that children with spina bifida exhibit by school age can be documented and monitored as early as infancy. Therefore, an intervention programme is warranted at the earliest stage, to provide support to families as to how they can stimulate speech and language development, and to provide a specialist link with local services. In the case of older, school age children, the Speech and Language Therapist and other members of the team, such as the Neuropsychologist, should provide baseline assessments to identify social and educational needs that become evident as social and academic demands increase beyond the early years (Lomax-Bream et al., 2007).

### **Results:**

90% of service providers reported that some form of speech, language or swallowing needs were prevalent with children with spina bifida. Language, communication and social skills difficulties were the most frequently reported.

Figure 43



Qualitatively service providers reported a delay in early communication with receptive language and social skills becoming more of a concern as the child develops.

*"I suppose because of the hydrocephalus a lot of those children, their receptive language would develop at a slower rate than their expressive language....they can be slow to start off. But once they do, then they get a burst of talking".(Service Provider)*

*"So their needs change a lot I think. I suppose with the younger ones, it's early communication. Then for school age, it can be kind of a specific thing like language or a speech thing. And as they get older, it tends to be the whole social communication".(Service Provider)*

## Discussion

Children with spina bifida present with a range of speech and language needs, with known concerns regarding receptive language, social development and executive functioning. Nearly all service providers reported concerns regarding their patient's language development. The importance of surveillance by a speech and language therapist is highlighted. Speech and Language intervention should commence at infancy to support language development with specific assessments at school age to examine social skills and higher level executive functioning difficulties. Strategies should be implemented with cooperation of parents and support from specialist centre where required. Access to Speech and Language Therapists are discussed in more detail in chapter 5.1

## Recommendation

1. Access to a Speech and Language Therapist should be should be available to all children with spina bifida for assessment of language skills and appropriate interventions as required.

## ***TISSUE VIABILITY***

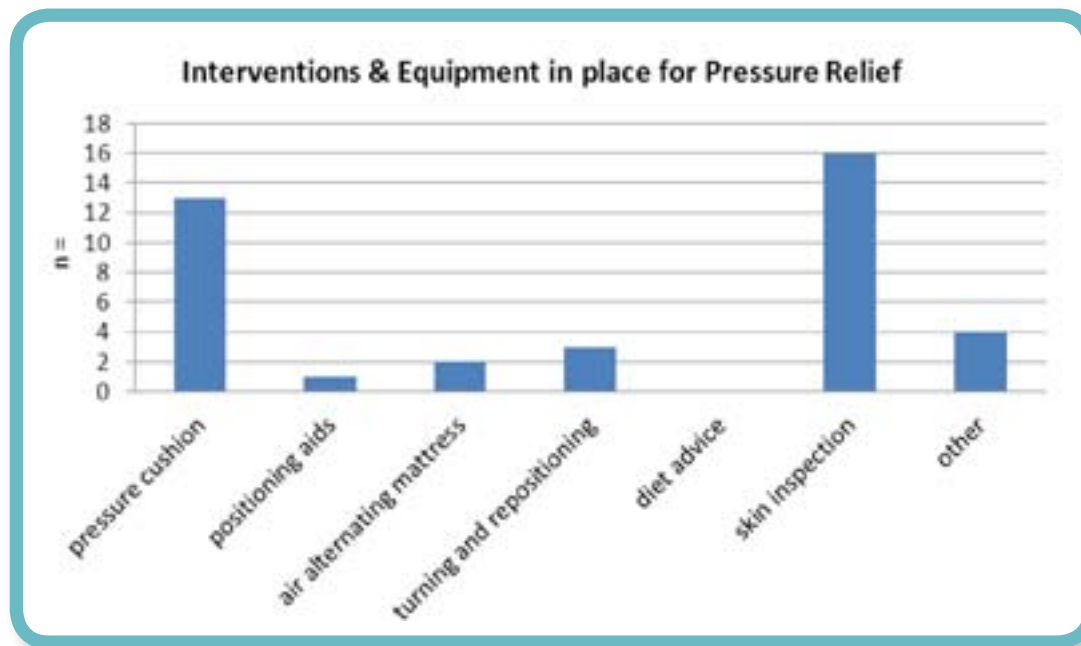
### **Background**

Skin integrity and pressure areas are a significant cause for concern in the child with spina bifida and there can be detrimental effects once a pressure area presents. Due to nerve impairments associated with spina bifida, individuals have diminished or absent skin sensation below the level of their lesion. As a result, they may not feel pressure, pain, or touch and may not respond to hot or cold touch. This, along with immobility, makes them readily susceptible to pressure sores and skin breakdown (America., 2005, Pallija et al., 1999). In addition, the effect of spina bifida on autonomic nerves may affect the vascular supply to insensate areas, and sores that are present may require extensive periods to heal (Liptak and El Samra, 2010). In an older review of inpatient admissions of chronically-ill patients (Pallija et al., 1999) could attribute 994 of 4533 or 22% of hospital days of children with spina bifida to be due to loss of skin integrity with significant cost implications for healthcare facilities and health implications for children. From the child's perspective skin breakdown can affect their body image leading to grief, anxiety and/or depression (Magnan, 1996). Pressure sores have been reported to be a current issue in 4% and a previous problem in 38% of adult with spina bifida (Liptak and El Samra, 2010). These figures suggest that the incidence of pressure areas is reducing. Possible explanation for this are increased vigilance from the neonatal stage and parental education, improved nutrition and wound care. Recent advances in moulded equipment will also have a part to play in reducing pressure areas.

### **Results:**

Nineteen per cent of parents in our sample noted that there was a history of pressure sores with their children, with 96% of those parents indicating that a measure had been put in place to manage this. The most common measures in place are regular skin inspection and pressure relieving mattress.

Figure 44



One parent commented on the significant impact a pressure sore had on her son which occurred following an operative procedure and a short period of immobility in hospital which should have been avoidable with appropriate pressure-relieving supports and education:

*"I just I never realised how bad a pressure sore could be, it was shocking, I was really taken aback... I mean they noticed it at lunchtime on Thursday and two hours later he had an air mattress, he could have had that on the Monday and it needn't have happened. I just felt that was something I couldn't control and he couldn't control it and the people who should have known better should have controlled it". (Parent / Guardian)*

Another described a pressure sore due to incorrectly fitting equipment:

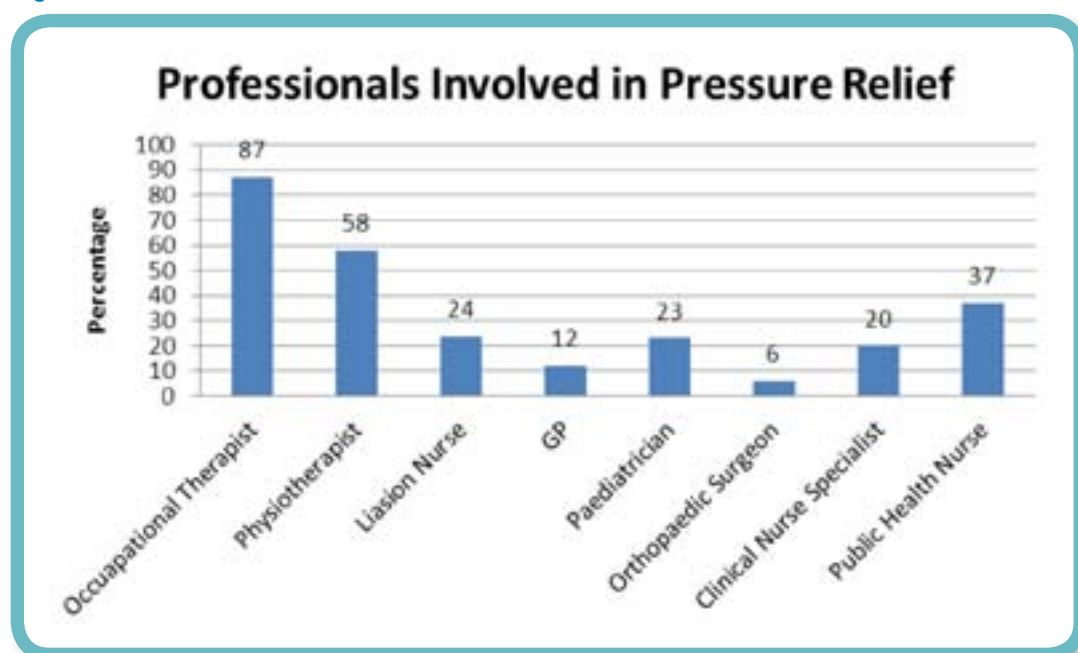
*"A wheelchair that doesn't suit him so he had, he was sitting on his coccyx bone which is always grinding against the cushion so it was, and he had a pressure sore and that lasted for like 2 years, and always bleeding". (Parent / Guardian)*

The impact of a pressure sore can result in further physical immobility while that pressure area heals.

*"There was a break down in his foot, it seemed like not a big thing. But it meant that he spent his first year in primary school in a wheelchair". (Service Provider)*

In relation to pressure relief, Occupational Therapists were identified as the professional most involved in the management of pressure relief (87% of service providers). Nursing staff (a combination of liaison nurses, clinical nurse specialists and public health nurses) were the profession next most frequently involved (combined 84% of service providers). Physiotherapists were reported to be involved by 58% of service providers. Paediatricians were reported to be involved by 23% of service providers. Paediatricians were reported to be involved by only 23% of service providers and GPs by only 12% of service providers.

Figure 45



In relation to assessment and intervention, 59% of service providers reported not having access to pressure mapping equipment and an additional 16% report this assessment is completed by another service. This equipment is essential for detailed monitoring of children's pressure sensitive areas in seating, but is rarely available for use with these high risk children.

## Discussion

Although the risk for pressure areas is high for those with impaired sensation, and the impact is devastating for the children and families who have suffered pressure sore, the numbers reported here are relatively low at 19% of parents indicating that their child had a pressure area in the past. This rate is higher than Liptak and El Samra (2010) reports as a current rate, but lower than their historical figures. Of note, nearly all parents who reported that their child had a pressure area had an intervention in place to manage this. This suggests increased vigilance and awareness of this preventable complication. Pressure mapping for children with limited mobility can be vital to prevent pressure areas, although the availability of this equipment is very limited. Children when identified as high risk should have access to pressure mapping assessment.

## Recommendations:

1. It is the responsibility of all health care professionals to be vigilant of pressure areas and to act on concerns when raised.
2. Children and parents require education regarding the risks of pressure sores and appropriate equipment needs to be in place to prevent the development of pressure areas.
3. Those at high risk of pressure sores, particularly those who are immobile, should have access to pressure mapping assessments regionally to determine appropriate preventative measures which need to be provided (i.e. pressure cushions or supportive equipment).

## 4.2 COGNITIVE AND PSYCHOSOCIAL IMPACT

### ***Neuropsychological profile***

Children with spina bifida and hydrocephalus present with a complex neuropsychological profile. They can often display average intelligence. However this condition can impact on children's cognition, psychological wellbeing, and ultimately on participation and engagement in activities in daily life, education, employment and independence (Dennis and Barnes, 2010, Holmbeck and Devine, 2010, Northrup and Volcik, 2000).

Difficulties related to cognitive functioning can present in areas such as hand-eye coordination (perceptual-motor), comprehension, distractibility and attention difficulties, restlessness or hyperactivity, memory or organisational skills difficulties, difficulties with sequencing, decision making and solving problems (Lollar, 1994). Dennis and Barnes (2010) identified two main cognitive phenotypes of spina bifida to include: domain general core assets and deficits which are related to brain dysmorphologies (e.g. Chiari II malformations) and domain specific functional deficits. Domain general core assets include issues related to timing and rhythm, attentional difficulties related to orienting and detaching and movement difficulties reducing dynamic motor control. The domain specific functional deficits relate more to coordinate perception, language in relation to rhetoric and semantics, literacy particularly when inferencing is required and numeracy in the areas of estimation, problem solving and mental calculation (Dennis and Barnes, 2010). Executive functioning has also been reported to be



impaired specifically relating to working memory (Burmeister et al., 2005).

Australian research collecting data between 1980 and 1999 identified that 19% of children with spina bifida received a diagnosis of intellectual disability (ID= IQ <70) compared with only 1% of individuals born during the same period without spina bifida who presented with an ID (Pettersen et al 2007). Those who do not have a shunt in situ were reported to have a much better outlook in terms of intelligence (Shaer et al., 2007).

In this study, information gathered from questionnaires and interviews with parents and service providers aimed to highlight needs related to cognitive and psychosocial functioning. The psychosocial impact from the perspective of children and adolescents themselves is required to understand their perspective however these have been seen as lacking in the literature (Kinavey, 2007).

Data will also be presented relating to cognitive and psychosocial functioning as highlighted in the child focus group from the perspective of children aged over 8 years with spina bifida.

## **Results:**

In relation to cognitive ability, 50% of parents reported that their child had undergone a cognitive assessment with the mean age of these children being 7.9years. The mean age of the children who had not had a cognitive assessment completed was 3.6years and a Mann-Whitney U Test confirmed that there was a significant difference between the age of children who had completed a cognitive assessment to those who had not ( $p=0.000$ ) suggesting children tend to be older when completing an assessment.

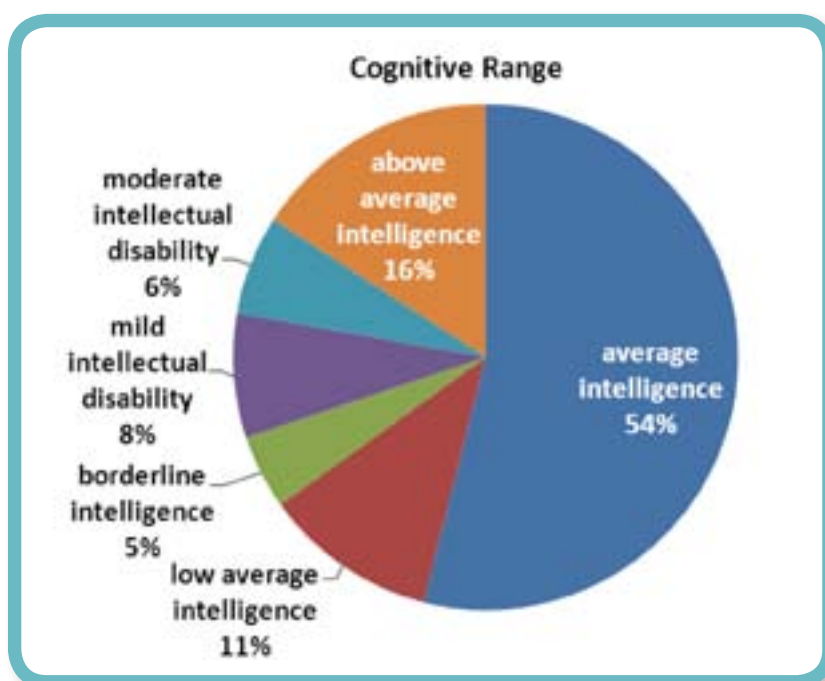
Of those that had not completed a cognitive assessment, 73% of parents reported that a cognitive assessment had not been required or requested to date and their childrens mean age was 2.9years suggesting these children were too young to complete a formal standardised cognitive assessment. A further 6% of parents report their child was waiting on a cognitive assessment and these children had a mean age of 5.5years. Finally 4% of parents indicated that a cognitive assessment was not available to their child however these children tended to be older with a mean age of 14.4years (range 10years to 16.8years). See Figure 35 for clarity. There was a significant difference in age for children who did not require/request an assessment to date, were waiting on an assessment or to whom it was not available ( $p=0.003$ ). The remaining 17% noted other reasons for their child not completing a cognitive assessment with comments including that their child was too young to complete an assessment or they were not aware or informed about the availability of this service. Access to the cognitive assessment appears adequate with the majority of parents who had not received a cognitive assessment highlighting this was not requested or required to date. However older children and adolescents need access to cognitive assessments if required.

Table 6

Cognitive Assessment Not Completed		
Reason	Percentage	Mean Age (years)
Not required/requested to date	73	2.9
Waiting on cognitive assessment	6	5.5
Not available to their child	4	14.4(10-16.8)
Note 17% noted 'other' reasons see comments above		

Of children who had completed a cognitive assessment, results suggest that children in the current sample display the full range of cognitive abilities. More than half of children (54%) fell into the average intellectual ability range (Figure 46). Overall 14% of parents report their child had received a diagnosis of intellectual disability (either mild 8%, or moderate, 6%).

Figure 46



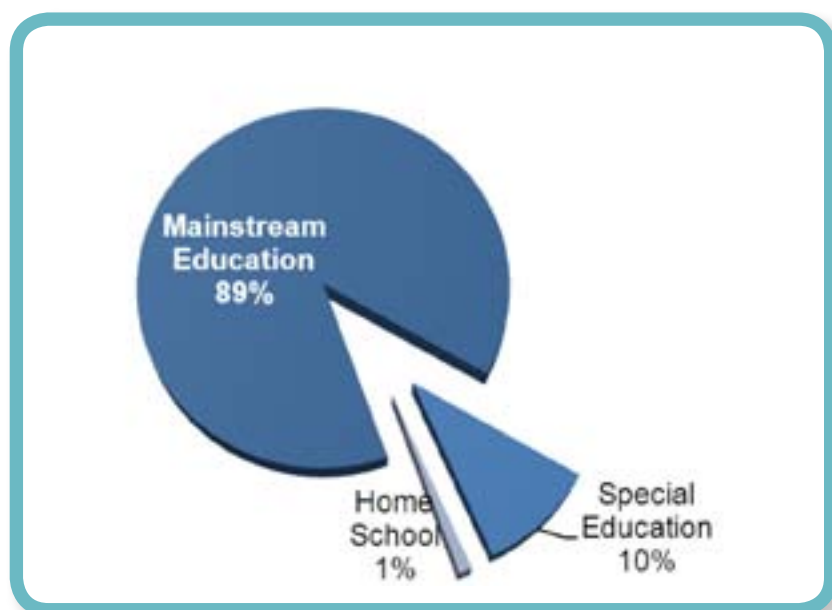
Service providers also commented on the cognitive functioning of children with spina bifida on their caseloads. 44% of service providers highlighted concerns in the area of attention/concentration and 39% felt there were also difficulties with learning and attainment within the classroom environment. These attentional and learning and attainment difficulties raised by service providers also require monitoring. Attentional difficulties in the form of Attention-Deficit Disorder and Hyperactivity Disorders are common with children with spina bifida however these difficulties often relate specifically to orientation of attention and difficulties with switching attention once engaged (Burmeister et al., 2005).

## Educational Setting

In relation to educational setting a large portion of the sample were too young to attend formal education (41%). Of the children old enough to attend school the majority (89%) attended mainstream education (18 children in preschool, 47 in primary school and 14 in secondary school) (Figure 47). For those who were in attendance many parents in interviews reported being satisfied with their progress in mainstream school.

Figure 47

## Educational Setting



For children who were accessing mainstream education with appropriate supports many parents highlighted positive outcomes with some noting good supports and inclusion:

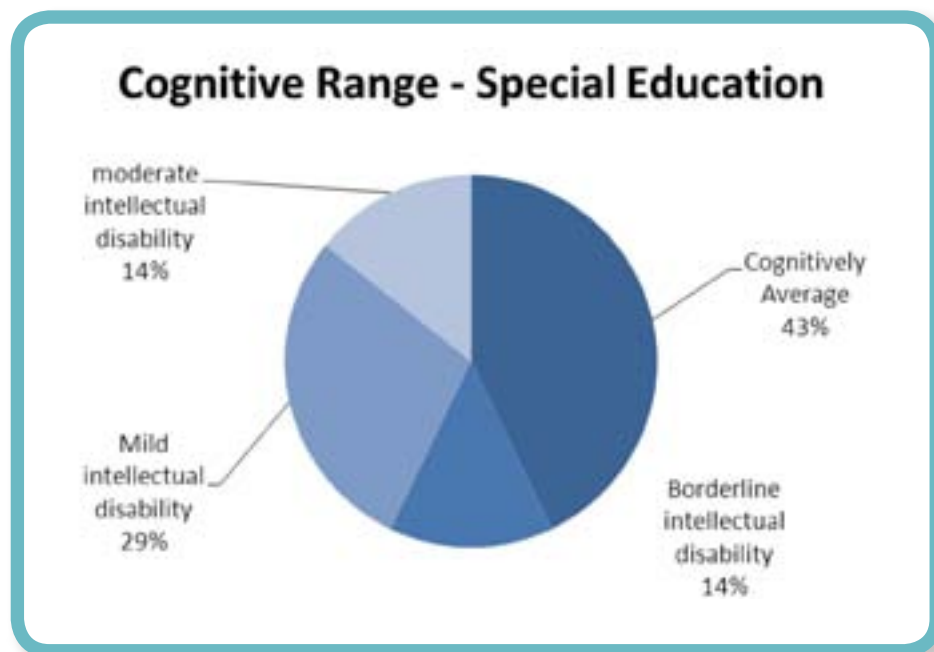
*“Everyone understands and likes him for who he is so he’s accepted and that’s why I like the school it’s inclusive” (Parent / Guardian)*

*“Academically he’s in mainstream school he has an SNA and he’s in 4th class. He’s average and I give him extra work myself to get him over but its good and he’s happy” (Parent / Guardian)*

Of those attending school, 9 children (10%) were in special education. The mean age of these children was 7 years with a range from 2.4 years to 16.3 years. Only 2 of the children were in special education secondary school, and 2 in special education primary school the remaining 5 were in special education preschools.

Of those attending special education, 7 had completed a cognitive assessment, the 2 who did not have cognitive assessments completed may have been too severe for standardised assessment but reasons were not provided. For those who had completed a cognitive assessment, the range is displayed below:

Figure 48



Of note, 43% of children attending special education presented within the average cognitive range however it is important to note this was only out of 9 children. Appropriate school placement and supports are of vital importance to enable cognitive and psychosocial functioning as studies have shown that positive experiences in school contexts can contribute to psychological outcomes (Essner and Holmbeck, 2010).

The determining factors as to whether a child will attend mainstream or special education school setting were multifactorial and individual with reasons including how motivated the child and parent are, their level of socio-economic status (SES), and the child's level of dependence for toileting tasks (catheterisation/bowel washouts).

*"It's a combination of parental interest and motivation and drive and belief in the child. And also, socio-economic background and the type of schooling that the parent chooses for the child, whether it's special or mainstream" (Service Provider)*

*"I think the parents who are more able will send the kids to mainstream. And they will have a more challenging educational experience" (Service Provider)*

Difficulties with accessing mainstream school were frequently noted by parents and service providers to be dependent on the availability of supports to assist with supervision or

catheterisation/toileting. This was most frequently provided by a special needs assistant (SNA), or occasionally by the parent themselves, with one reporting she visited her child 3-4 times daily in school to complete clean intermittent catheterisation (CIC) as the SNA was not allowed or not willing to complete this task. The impact of social continence and the important role of school-based nursing support is discussed by Katrancha (2008) who concluded that clean intermittent catheterization (CIC) with nursing support helps build the child with spina bifida's self-esteem, and allows the student greater independence in school. Service providers also highlighted dissatisfaction with the inequity of this.

*“Some of the children in our (special) school could go to mainstream if there was support around the catheterisation” (Service Provider)*

*“I don't believe that their toileting and their other issues should prevent them going to mainstream” (Service Provider)*

Concerns were often raised by parents in relation to inequity of access to school supports, and fears regarding further cuts to these which would affect child's ability to partake. Parents were often required to coordinate school transitions and planning for full participation in the school system.

*“And I think what's happening with resources being cut, particularly around special needs support in schools and the special needs assistants and social resources that need to be put into schools, particularly around toileting or bowel management, we have a lot more to do there” (Service Provider)*

*“He's just done his Junior Cert and he passed everything by the skin of his teeth. But he did which is great because he doesn't, he's border line on all his assessment so he didn't qualify for any resource help or anything like that so that's hard” (Parent / Guardian)*

In particular physical accessibility of schools was generally seen as poor. Service providers and parents commented on the difficulty of physically moving class, particularly in secondary school and also difficulties with access in the yard, wheelchair-accessible toilets/changing areas and the need for frequent equipment provision to schools.

*“Trying to move from class to class and trying to find a toilet, disability toilet that's sometimes the other thing” (Service Provider)*

*"I think it would be great if schools were more accessible as standard" (Service Provider)*

*"Her school yard is a huge problem, just with access, because the entire school is on a hill, so she won't walk with the walker. And it's just not suitable for her really. And we are working with the school constantly" (Parent / Guardian)*

*"Getting ready for school would be a huge area of intervention, doing the school visit, prior to them starting school, doing any applications for equipment. So a changing bench, looking at turning space, access, all environmental stuff. So there is a lot of work around that age, that transition into primary school" (Service Provider)*

Real inclusion in schools is vital to prevent children from becoming isolated, having reduced confidence or lack of friendship reciprocation either through physical or psychological barriers.

*"We've often had that if he goes on a school tour I have to drive behind the bus cos they can never get a wheelchair bus for the school tour" (Parent / Guardian)*

*"I think just sometimes out the yard, she does get a little left" (Parent / Guardian)*

*"I think there is more of an understanding now, the child is a whole child and whatever needs the child has, needs to be addressed in school, in order that their educational needs can be met, so there isn't that division" (Service Provider)*

*"The other issue is how the child is supported with school with bowel and bladder issues, and that's obviously a major problem if the staff within the school or the children don't really accept and help the child integrate and allay the embarrassment then it can be a big problem for many children" (Service Provider)*

*"So it's trying to get that balance of, they don't become isolated in the community and the sense of themselves and their sense of being involved in the community" (Service Provider)*

## Discussion

Results suggest that although the majority of children present within the average cognitive range and attend mainstream school they face difficulties with accessing appropriate supports to assist with attention and concentration difficulties and meeting continence needs. Peny-Dahlstrand et al (2013) found that children with spina bifida require more support to be actively involved in school, particularly during unstructured activities. Their participation was also seen to be directly correlated to their physical ability (Peny-Dahlstrand et al 2013). The United Nations Convention on the Rights of Persons with Disabilities states that all countries should provide full accessibility and that all necessary measures should be taken to ensure the full enjoyment by children with disabilities of all human rights and fundamental freedoms on an equal basis with other children (UN, 2014). In spite of this, physical access issues and issues with inclusion in the school environment were raised by both parents and service providers. Many children in the current study who attend special education schools were reported to be in the average intellectual ability range with qualitative comments suggesting that access to mainstream school can be dependent on access to appropriate supports to meet bladder and bowel needs (i.e. SNA/Assistants).

## Recommendation

1. Mainstream education, along with necessary supports, should be encouraged for children with spina bifida, when this is appropriate to their learning needs. Access to mainstreams school should not be influenced by physical or continence needs.
2. Supports are required to assist with physical or continence needs might include SNAs, classroom assistants, resource teaching or school nursing.
3. Improvements in physical accessibility of schools and communities are necessary to increase independence and active participation in daily life.

## Psychosocial Functioning

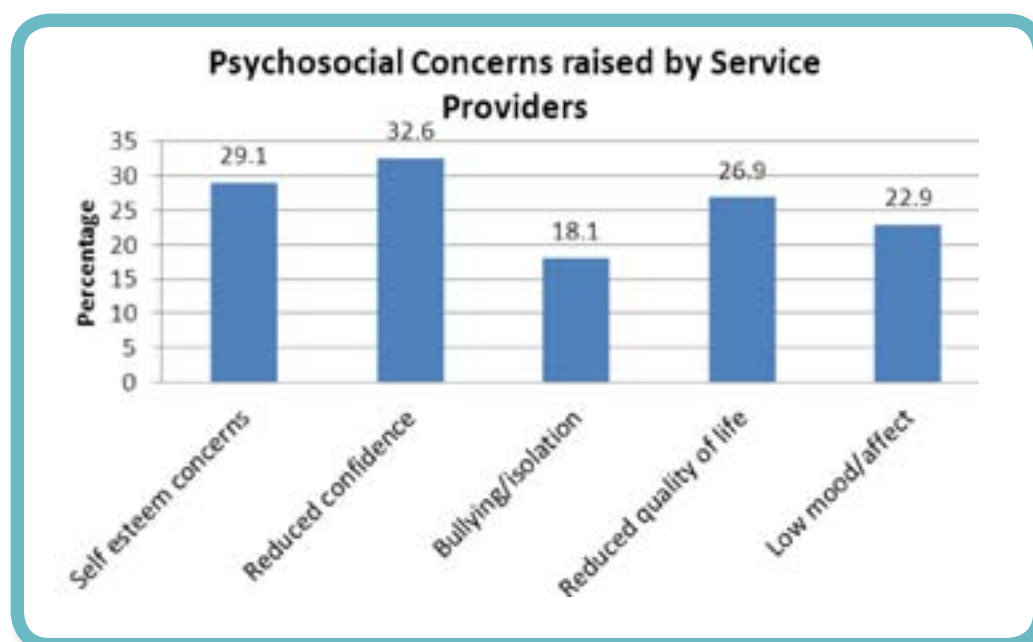
### Background:

The psychosocial functioning relates to the social, emotional and psychological functioning. Many aspects of spina bifida including personal, psychological, social and sexual issues can impact on the psychosocial functioning of these children (Crosthwaite et al., 2001). Examples of psychosocial challenges for which they are at risk include difficulties with depression and anxiety, reduced confidence and self-esteem, poor body image and stigma (Bowman et al., 2001, Kinavey, 2007, Merkens, 2006). One of the functional outcomes for spina bifida treatment should be to achieve developmental competence and positive psychosocial functioning (Merkens, 2006). Supporting psychosocial functioning should include maximising development within supportive environments while fostering positive identities (Bellin et al., 2010, Kinavey, 2007).

### Results:

Fifty six per cent of service providers reported that psychosocial concerns are prevalent for children with spina bifida. Out of those who highlighted a concern in the area of psychosocial functioning, professionals identified issues to include reduced confidence (n=74, 25%), reduced self-esteem (n=66, 22%) bullying/isolation (n=41, 14%), reduced quality of life (n=61, 21%) and low mood/affect (n=52, 18%) (see Figure 48).

Figure 49



25% of all parents advised that spina bifida had impacted on their child's self-esteem.

By school-age (5 years) this rate had increased to 50% and remained at this level at aged 12 years.

When comparing across age groups there was a significant increase in self-esteem difficulties with increasing age ( $p=0.01$ ). Results indicated a significant increase from the under 2s, to the 2-6 years and between the 2-6 years and the 6-12 year age groups. This indicates that self-esteem difficulties can present as early as 2-6 years. Given the lack of difference between



the 6-12 year age group and the over 12s, it would appear that self-esteem issues are well established by the 6-12 age range.

Correlation analysis was performed between self-esteem and mobility level, presence of hydrocephalus, obesity, bladder and bowel interventions, and age to determine which factors were most related to reduced self-esteem. There was no difference noted in self-esteem between those that were ambulant and those that were wheelchair users; neither was there a difference between those that had hydrocephalus compared to those that did not.

Self-esteem was also impacted by obesity, but only those children in the morbidly obese category were significantly more likely to have self-esteem issues than the normal weight ( $p=0.013$ ), overweight ( $p=0.008$ ), and obese ( $p=0.014$ ) groups. In summary there was a significant relationship for children who were morbidly obese and self-esteem issues when compared with other groups.

The presence of bladder and bowel interventions significantly affected self-esteem ( $p=0.016$ ) (of note the question asked related to interventions not difficulties). The impact of continence and bladder and bowel interventions and its significant impact on self-esteem requires further investigation.

In interviews, service providers commented on the negative impact of weight gain and difficulties with bladder and bowel management on self-esteem and confidence:

*“A big impact is the weight gain on self-esteem, but also on ability to engage with activities. And that’s something I think that and the toileting issues are to me are the biggest issues that need to be addressed when the kids are very young” (Service Provider)*

*“They become a lot more conscious of that (bladder bowel difficulties) and it impacts hugely on their confidence” (Service Provider)*

*“He is very aware that he wears nappies and socially he feels different” (Parent / Guardian)*

*“She is very self-conscious about her splints, does not want anyone to see them. Has suffered the embarrassment of toileting accidents in school. She is also very conscious of the scar on her back” (Parent / Guardian)*

Some service providers commented during interviews about difficulties with motivation or indifference and also on the negative impact of conflicting child and parental aspirations:

*“Huge apathy in terms of their learning... not being bothered, no motivation, very low motivation to learn” (Service Provider)*

Both parents and service providers recognised that some children demonstrated excessive sociability or disinhibition without regard for social consequences or cues. While this was seen by parents as positive, some service providers felt this did not accurately demonstrate their actual social abilities and felt it could be “deceptive”.

*“He is super confident even to the point of being inappropriate sometimes but at least he’s not hiding in the corner” (Parent / Guardian)*

*“I think some of the parental aspirations can be low, and that has a huge impact on the kids. And some of the kids’ aspirations can be ridiculously high, not in line with how they are actually doing” (Service Provider)*

*“Cocktail party syndrome and she falls into that category...And behind it all that then, if she asked you what did you have for your breakfast this morning, she mightn’t have a clue really not a clue” (Parent / Guardian)*

When speaking with the children themselves during the child focus group some inappropriate perceptions regarding friendships and relationships were also raised. For example after just meeting the interviewer that day one participant invited the interviewer to her upcoming house-warming and another participant commented that she has a boyfriend but then went on to explain he was a famous rugby player. Others seemed idealistic with regards friendship groups with comments including:

*“The whole school is my friend up to 6th class, and 5th class” (Focus Group)*

Finally, the detrimental effect of long term mental health difficulties which may present if the social, emotional and psychological needs of children are monitored reactively as opposed to proactively was highlighted by many of the service providers.

*“A lot of our kids end up not that happy with mental health issues. And there is a lot of services that go into the lives of these children and it’s at the end, we really haven’t succeeded if they are in a bad place at the end of all of this medical intervention. So I think all the other stuff outside of the medical thing is so important” (Service Provider)*

## Discussion:

Results of this study highlight a high prevalence of difficulties with psychosocial functioning from the perspective of parents and service providers with difficulties highlighted relating to reduced confidence, reduced self-esteem, reduced quality of life, mild issues relating to bullying and low mood/affect. This is supported in the literature with studies reporting a high risk of reduced quality of life due to their physical and psychological profile and supporting and fostering independence to support quality of life is of great importance (Padua et al., 2002, Cornegé-Blokland et al., 2011). A higher incidence of bullying can be evident for children with disabilities and therefore this risk needs to be appropriately monitored (Bell Carter & Spencer, 2006). These complexities further highlights the importance of early intervention with a full multidisciplinary team to support the psychosocial needs of these complex children (Carroll et al., 2013).

Overall, self-esteem was not affected by ability to walk or by the presence of hydrocephalus. Results suggest it is however affected by the presence of bladder and bowel interventions, being morbidly obese, and increases with age until 12 years. Verhoef et al. (2005c) similarly found that urinary and faecal incontinence can impact on the quality of life of patients with spina bifida. It is important that the development of self-management skills, fostering independence, ensuring a positive body image, managing excessive weight gain and ensuring social continence are encouraged as these could all help to reduce the significant psychosocial impact of spina bifida on the child.

Children in the current study were noted and observed to be overtly sociable at times. Although underresearched this phenomenon of inappropriate sociability, excessive verbosity, social immaturity and other difficulties with social skills have been described in the literature (Burmeister et al., 2005, Holmbeck et al., 2003) and may be associated with other aspects of the cognitive profile, support in these areas is required.

## Recommendations

1. All team members should be aware of and responsive to the child's psychosocial needs and there is a need for on-going psychological interventions when cognitive or psychosocial issues present.
2. Exposure of children to peer support activities for example extra-curricular activities, sports or social events, is essential.

## **Services to support Cognitive and Psychosocial Functioning**

As a result of the complex and varied cognitive and psychosocial needs of children with spina bifida various local and specialist services are required to support their social, emotional and psychological needs and to support engagement in education, employment and independence (Holmbeck and Devine, 2010, Northrup and Volcik, 2000). This section describes access to supports necessary to assist with cognitive and psychosocial functioning.

### **Results**

During interviews service providers highlighted the reactive rather than proactive approach to supporting psychosocial functioning with only the “tip of the iceberg” (SP) being met but both parents and service providers highlighted the need for appropriate supports and professionals to assist with psychological functioning.

*“Psychology here is always when there is a crisis and when we need them in for support or if things are falling apart ... it’s fire fighting” (Service Provider)*

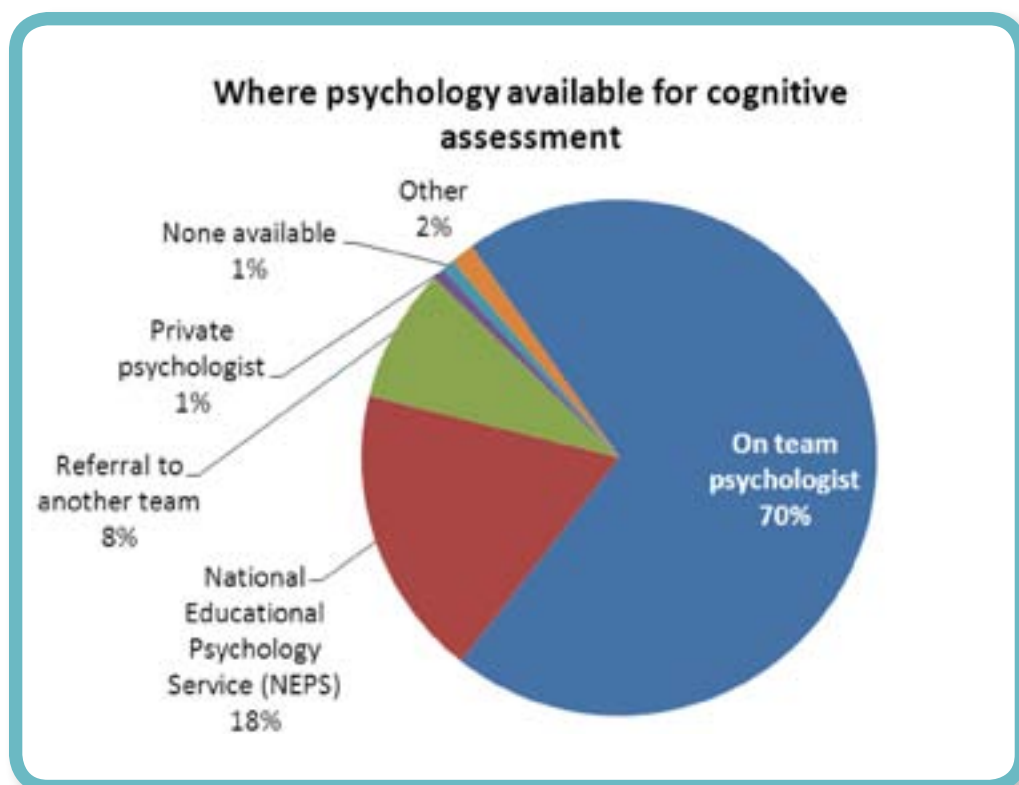
*“They are often children who will to some degree try and cover up those needs by just putting on very brave face, and I think it’s up to us to really recognise that they have very significant needs in that direction. So I would strongly recommend that any team looking after children with spina bifida would have good psychological support” (Service Provider)*

*“A huge gap of psychology I think that’s huge, for the parents” (Service Provider)*

*“Parents need an awful lot of psychological support ... and that wouldn’t have been available to them” (Service Provider)*

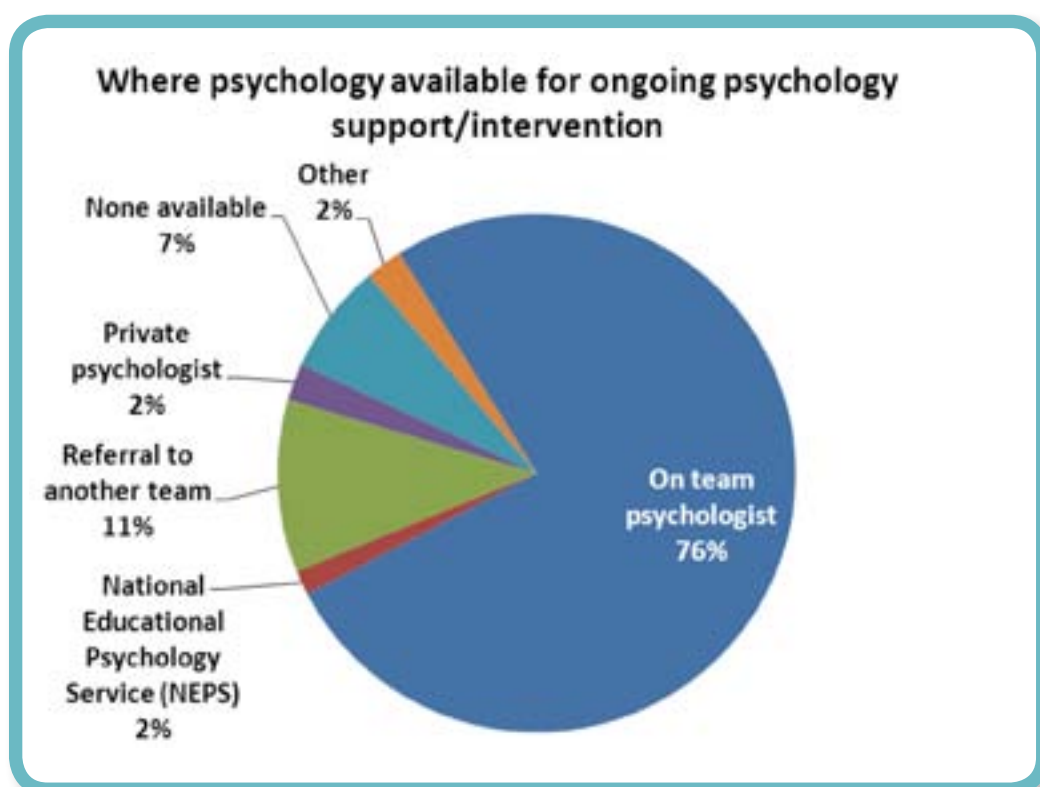
Service providers were asked where they could access psychology for cognitive assessments and also where they could access psychology for on-going support or intervention. Seventy per cent of service provider report they could access a cognitive assessment through their on-team psychologist, 18% through the National Educational Psychology Service (NEPS) and 8% required a referral to another team (Figure 49).

Figure 50



For on-going psychological supports or interventions 76% reported this could be accessed via their on-team psychologist and 11% required referral to another team (Figure 50).

Figure 51



The need for early psychosocial supports and counselling was also highlighted and also difficulties with accessing psychology supports when required.

*“We referred him on to the psychologist but there was a year and a half waiting list and by the time we had overcome the problem” (Parent / Guardian)*

*“The most important thing is just to get in there early” (Parent / Guardian)*

The psychologists who responded to the questionnaire (n=10) were asked which service they feel they personally provide. All psychologists reported they were involved in the development of learning and attainment 7 reported being involved in self-care, and 3 were involved in interventions related to sexual functioning. The majority of service providers highlighted the importance of having a psychologist present at a Multidisciplinary Spina Bifida Clinic (52% felt this was very important and 34% felt it was important). In contrast less parents placed the same importance on their presence (19% felt this was very important and 14% felt it was important). This response from parents may be due to the young sample age with many psychosocial needs only becoming more apparent with age. Education for parents regarding the potential long-term social, emotional and psychological difficulties which can present in spina bifida is required. Overall the psychosocial well-being of children with spina bifida was seen as high priority by 85% of service providers justifying the need for service providers and parents to be appropriately educated about supporting children in relation to these needs.

Finally, in relation to services to support cognitive and psychosocial needs, results suggest that children’s cognitive ability is currently being monitored but less proactive interventions to support social emotional and psychological needs were reported. Monitoring and actively supporting psychosocial needs is vital in order to ameliorate for the increased risks for mental health problems which can present into adulthood (Sawyer and Macnee, 2010, Holmbeck et al., 2010).

## **Discussion**

Merkens (2006) guidelines recommend the monitoring of cognitive and social development. It is specified that these children should undergo a developmental assessment of their psychological and educational skills during pre-school years (aged 3-5 years). Attentional or concentration difficulties should be monitored if present, with referral to psychology if difficulties arise.

## Recommendations

1. Children with spina bifida should have frequent monitoring of their cognitive and psychosocial needs with access to a cognitive assessment and interventions to support their abilities when necessary.

## *Child's Strengths and Quality of Life*

Parents were asked to describe the good things about their child with spina bifida or their child's strengths. Most parents highlighted a number of positive attributes. Many highlighted their good, funny or determined nature and sociability, happiness and independence were aspects highly rated where available. For a visual of the most frequently represented words describing the children's strengths please see the figure below.

Figure 52

### Childrens Strenghts





The child focus group session highlighted some difficulties with inclusion and acceptance from the child's perspective. Overall results of the KINDL questionnaire, Life Ladder and Happiness in School scales (which were used to prompt further discussion regarding quality of life) highlighted that the older girls appear to have lower health related quality of life than the boys. It is important to bear in mind the very small sample size of 4 children and also to consider that the girls, being slightly older than the boys may have simply demonstrated an increased level of insight into their difficulties with maturity.

In relation to specific questions on the KINDL questionnaire results, children were asked to rate on a 5 point Likert scale how often during the past week they felt about a list of items (e.g. afraid my illness might get worse, sad because of my illness). The 5 point scale ranges from 1= all the time, 2=often, 3= sometimes, 4= seldom and 5=never. The highest rated was that none of the children reported feeling sad about their illness that week however the lowest rated answer was 12/20 in the area of "I wanted nobody to notice my illness" with the most frequent answer being 'sometimes I didn't want this to happen' (Figure 52 and 53).

Figure 53

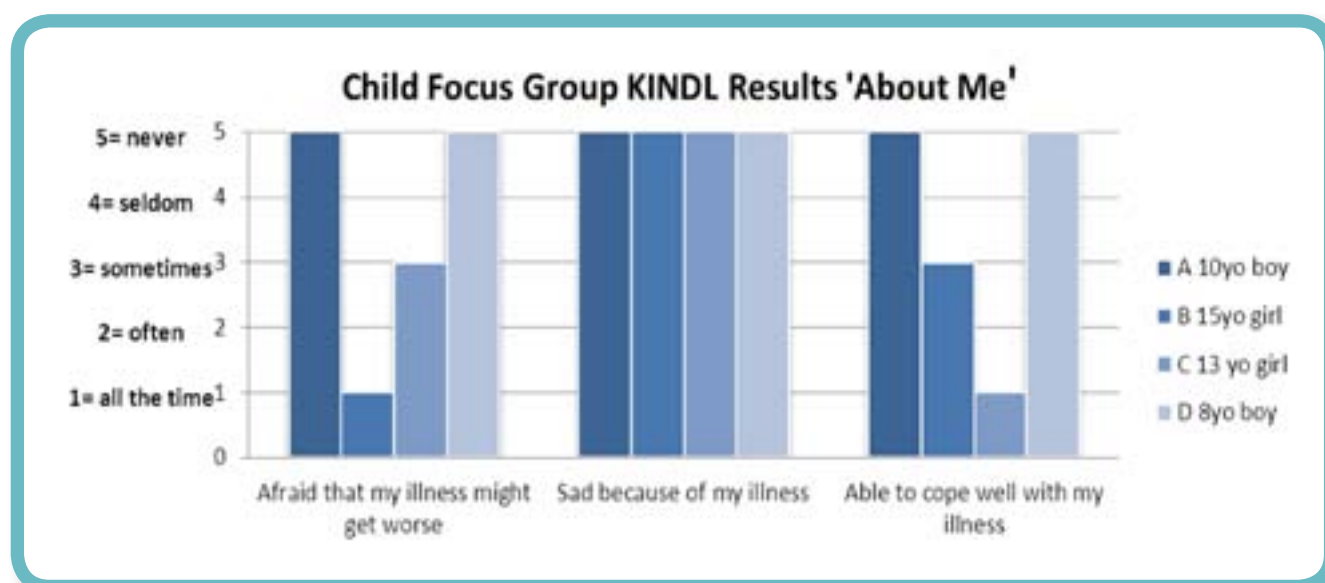
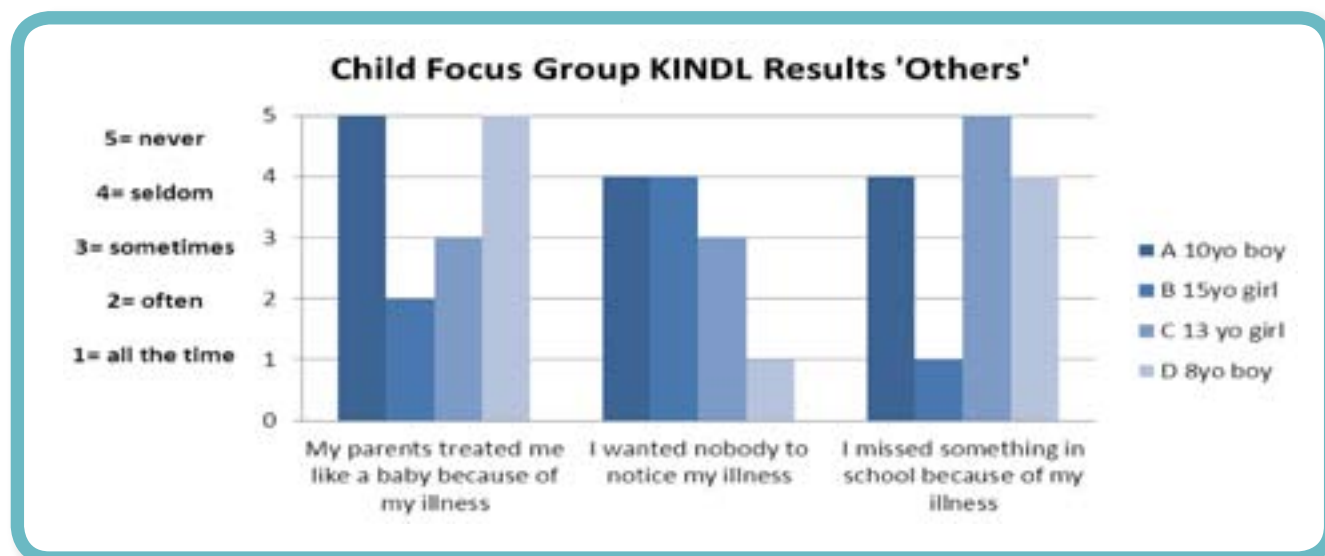


Figure 54





## 4.3 FAMILY IMPACT

### Background

The complex nature of spina bifida includes various life events including neurosurgical procedures and repetitive daily hassles which can have chronic impact including managing incontinence and ambulation and have the potential to have a drastic impact on the family (Vermaes et al., 2008). Ignanace Vermaes from the Netherlands has described the impact of spina bifida on the family with one study suggesting that parenting a child with spina bifida can impact psychological functioning and can commonly result in stress (Vermaes et al., 2005). This study also highlighted that having a child with spina bifida in a family can be a significant challenge to parental psychological well-being, especially for mothers. In a later study (Vermaes et al 2008) reported that stress is particularly common with more severe physical disability and attributes which help parents to cope with stress include extraversion, emotional stability and agreeableness. Cleve (1989) reported a significant positive relationship between parental coping and marital satisfaction, the quality of the relationship and the attendance at a spina bifida support as well as the fact that higher income and increased age of the parent contributed to parental coping.

Having a child with spina bifida within a family can impact on work-family divisions and parents can feel more socially restricted than other parents (Vermaes et al 2008). Adzick et al. (2011) also commented on the enormity of the emotional and financial impact of having a child with spina bifida on the family. Loebig (1990) described the greatest impacts of children with spina bifida in the family as financial, family conflicts and sibling over protectiveness. As a result of the above Vermaes et al. (2005) therefore recommends parental psychological assistance which should be part of the care of the child with spina bifida to ensure family wellbeing

### Results

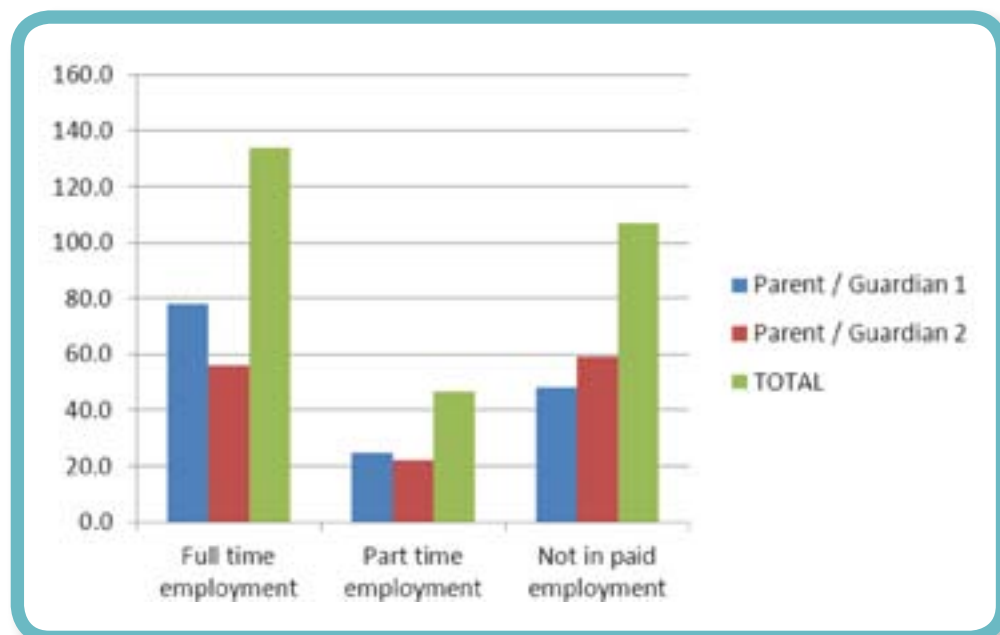
#### Family Dynamics and Financial Impact

In relation to family dynamics, from parent guardian questionnaire respondents the mean number of children per family was 2.72, on average the child with spina bifida was the 2nd child of the family and over 92% of families had 2 guardians. In interviews, some parents highlighted that having a child with spina bifida had affected their decision regarding extending their family, or the timing of same. One parent who subsequently had another child and saw positive impact of this explained the reason for delaying doing this:

*"I think the main delay was...after her surgery, she's in cast quite a bit, that we could concentrate on her fully. I suppose we were just able to spend a lot more time with (child with spina bifida). Now in hindsight, I think that if we realised how good things would be, it probably would have gone sooner"*  
(Parent / Guardian)

Figure 55

## Parent / Guardian Employment



47% of parent / guardians reported to be in full-time employment; 16% were in part-time employment and 37% were not in paid employment (Figure 58).

In interviews many parents commented negatively on their need to reduce time in paid employment due to the demands of their carer role. 54% of parents in the questionnaire highlighting that their employment status had changed as a result of having a child with spina bifida. Examples of comments from parents include:

*"I am unable to work as I am a full time carer to my son"*  
(Parent / Guardian)

*"Cannot go back to work because crèches won't take her and I have no one to mind her"* (Parent / Guardian)

*"Because of bladder/bowel continence and mobility issues, I could not expect a family member or fully rely on a crèche/school in later years to fully tend my sons' needs to my satisfaction"*  
(Parent / Guardian)

While some financial compensation exists, significant extra costs were also highlighted in caring for a child with spina bifida. A parent described the financial impact:

*“It’s just everything is just money draining...there’s extra costs because our electricity bill won’t be the same if we didn’t have (him), he has an airbed and that has to be on throughout the night...and there’s appointments to go to so there’s petrol” (Parent / Guardian).*

Some parents also highlighted the change in family dynamics of less time for siblings and extra planning required for day to day life, particularly in relation to identifying wheelchair accessible holidays or venues for family outings:

*“It does impact really on family life probably more than you think it will. Regular days out have to be thought out a lot more and a lot farther in advance. If you didn’t have a child with a disability you could go on a whim but you really can’t go off on a whim, you have to plan, everything has to be planned really” (Parent / Guardian)*

*“If you’re going out for dinner or anything I’d always go for a ground floor”  
(Parent / Guardian)*

There is limited research regarding the impact of spina bifida on siblings but the importance of including the siblings’ perspective in family-centred practice has been highlighted due to the risk of stress and challenges for siblings (Bellin and Rice, 2009). Parents in the current study highlighted both the positive and negative impact on siblings with more responsibility expected of siblings and at times less parental availability. However many described an increased awareness and understanding of disabilities.

*“It does impact on family life and siblings are affected as well”  
(Parent / Guardian)*

*“Now they see what different looks like and how you could live with other people with disabilities” (Parent / Guardian)*

Some parents also highlighted new diverse social opportunities which they established through peer support, with the positive impact of new friendships which can assist with coping with having a child with spina bifida.

*“I think the connections we have made with other families, through the association it’s a different style of friendship because, we have something much more significant shared” (Parent / Guardian)*

*“So those kind of connections are very deep. And I’m very grateful for them”  
(Parent / Guardian)*

However, coping can depend on family circumstances / supports as suggested by one service provider below:

*“We have found that the level of disability is not the indicator of the family distress, you could have a single parent with a very disabled child, may need a lot of support...sorry also you could have a single parent with a very mild child but their whole lives have been turned upside down, they mightn’t be able to return to work and may need a lot of support. So it’s not so much the diagnosis, it’s the family circumstances really is what we will be dealing with” (Service Provider)*

## **Discussion**

Results highlighted the substantial impact of spina bifida on the family in relation to changes in family dynamics, financial situation and siblings. There was a significant impact on the extended family and having a child with spina bifida resulted in reduced time for siblings and extra planning required for family activities and outings. Financially, the impact was twofold due to increased costs in caring for a child with spina bifida and often the forced change in parental employment status. These implications have been reported previously in relation to family impact (Loebig, 1990) and Yi et al (2011) similarly discussed the reduction in paid work for parents of children with spina bifida. These impacts on the family of a child with spina bifida can be stressful and challenging and adequate supports are necessary to ensure a good quality of life.

## **Recommendation**

1. On-going family, peer and sibling support should be available to all families wishing to avail of this

## Diagnosis

### Results

Qualitative interviews highlighted persistent upset and anger and negative emotional reactions to their child receiving a diagnosis of spina bifida, particularly when this was late or not diagnosed antenatally. Parents reported for example

*“Despite having numerous scans, it wasn’t picked up...  
I was very angry about that in the beginning”(Parent / Guardian)*

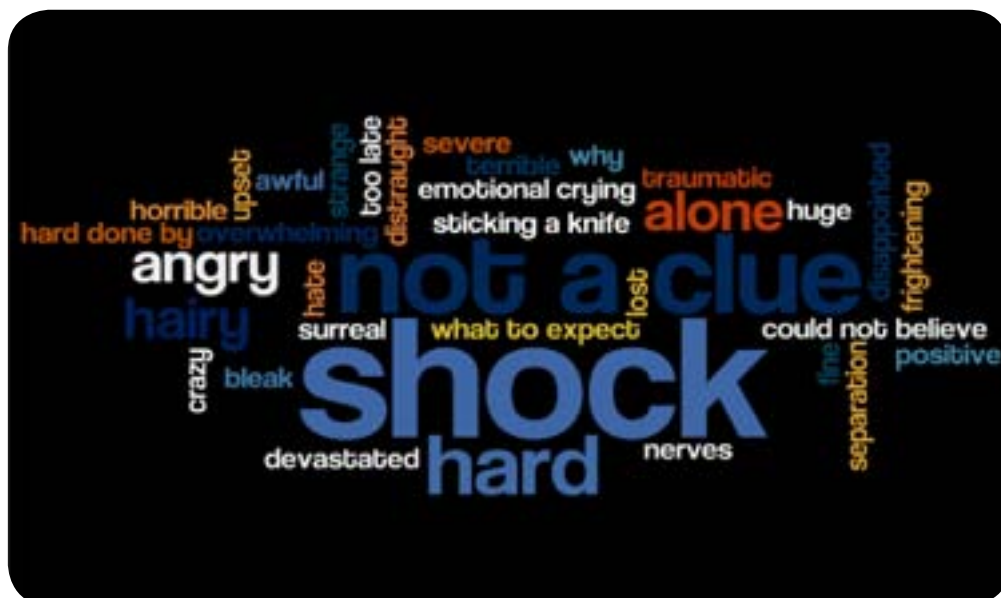
*“We only discovered that she was going to have  
spina bifida the night before she was born”  
(Parent / Guardian)*

Parents reported mostly feelings of shock as their emotional reaction to obtaining the diagnosis, with fears about what to expect and concerns about future prognosis. Not being aware of spina bifida as a condition prior to the diagnosis was also reported to be an extra stressor for parents during this difficult time.

*“The only way I can describe it was that someone was just  
after sticking a knife in us. Because it was a fair shock now,  
she was our first child and we knew nothing about it beforehand”  
(Parent / Guardian)*

Figure 56

Parent / Guardian emotions of diagnosis



## Discussion

Parents reported a mainly negative initial emotional reaction to diagnosis and birth, and this emotional reaction was still apparent at time of interview, in most cases many years subsequently. During interviews most parents spent a large proportion of the time discussing their initial reaction to the diagnosis and the stress, anxiety and upset during this time. A surprisingly large number of parents who were interviewed obtained no antenatal diagnosis or a diagnosis very late in their pregnancy, it is important to note however that this sample was self-selecting and may not be representative of all parents' experiences.

## Recommendation

1. Parents and families require earlier support, practical and emotional assistance following the antenatal diagnosis of spina bifida.

## *Folic Acid and Prevention*

As discussed in the background chapter, women of childbearing age should take 400 µg of folic acid three months before becoming pregnant and in Ireland there is voluntary fortification of folic acid in the food supply. To ensure sufficient intake of folic acid by women of childbearing age, 47 countries internationally have introduced mandatory folic acid food fortification (usually in cereal grain products) for the prevention of NTDs including the United States, Canada, Australia and New Zealand (EUROCAT, 2014). In the UK, the Scientific Advisory Committee on Nutrition met in June 2009 and supported a recommendation for mandatory fortification with folic acid with controls on voluntary fortification (Scientific Advisory Committee on Nutrition, 2009). The European Surveillance of Congenital Anomalies (EUROCAT, 2005) recommends that European countries should review their policies regarding folic acid supplementation by additionally introducing fortification of a staple food with folic acid. In 2006 the Food Safety Authority of Ireland published the 'Report of the National Committee on Folic Acid Food Fortification' which recommended the mandatory fortification of all bread in Ireland with folic acid noting that this would require legislation, regulation and monitoring and continued education and health promotion (FSAI 2006). In spite of this mandatory folic acid food fortification has still not been introduced in Ireland.

A large systematic review of the literature regarding the impact of folic acid fortification of flour on the prevalence of NTDs highlighted that fortification has had a major impact on NTD in all countries where implemented with examples including a 60% reduction in Costa

Rica and 55% in Chile (Castillo-Lancellotti et al., 2013). The impact of mandatory folic acid fortification of cereal grain products in the United States which commenced in 1998 has further decreased the prevalence of NTDs by 26% (CDC, 1995, Au et al., 2010). In September 2013 however, the Food Safety Authority of Ireland (FSAI, 2013) released a press release advising against mandatory folic acid food fortification in Ireland. This was following testing of blood samples between 2005-2007 and 2010-2012 which indicated evidence of high intakes of folic acid mainly due to voluntary food fortification by industry however the FSAI reported they will continue to monitor the situation (FSAI, 2013). Conversely a recent review of NTDs in Ireland (McDonnell et al., 2014) highlighted an increasing incidence of NTDs in Ireland and recommended an urgent review of public health policy on folic acid fortification, folic acid supplementation and pre-conceptional care. In the United States where mandatory fortification is in place the Spina Bifida Association of America still highlight the vital importance of ensuring education about folic acid and supplementation for future pregnancies for parents and children/adolescents with spina bifida (Merkins 2006). Yunni et al. (2011) completed an economic evaluation and demonstrated that folic acid fortification in food and preconception folic acid consumption are cost-effective ways to reduce the incidence and prevalence of NTDs when considering the significant cost burden of NTDs on healthcare systems and society concluding that the benefits of prevention of NTDs with folic acid far outweigh the cost.

Reluctance to introduce mandatory fortification of folic acid has been due to fears of adverse effects and the fact that by targeting the food supply all members of the population, and not just women of child bearing age, would receive extra folic acid. Taking folic acid may reduce the risk of certain cancers, with moderate evidence to support the protective effect of dietary folate in colorectal cancer (Scientific Advisory Committee on Nutrition, 2005). However one study suggested that if the amount of folic acid greatly exceeds normal requirements then this may be cancer-promoting (Kim, 2004) and while there is no direct evidence or randomised controlled trials to support this, it is important to closely monitor the fortification and ensure appropriate and safe dosage. Taking too much folic acid can mask a vitamin B12 deficiency by correcting the associated anaemia and thus hiding the deficiency which can cause other potential neurological problems, therefore the FSAI calculated the appropriate level of folic acid suitable for food fortification to inform their decision making. (FSAI, 2006).

Folic acid supplementation may yield other health benefits with some research suggesting it may reduce cardiovascular disease risk but this requires further confirmation (Food Safety Authority of Ireland, 2008). Yunni et al. (2011) completed an economic evaluation and demonstrated that folic acid fortification in food and preconception folic acid consumption are cost-effective ways to reduce the incidence and prevalence of NTDs when considering the significant cost burden of NTDs on healthcare systems and society concluding that the benefits of prevention of NTDs with folic acid far outweigh the cost.

As discussed previously in chapter 4.1 Neurosurgical Needs section, results indicated a very strong family history of NTDs in the current sample. During interviews some parents discussed their history of periconceptual folic acid intake and public perception and knowledge about folic acid suggesting that increased awareness is required.

*“I think she said, I’m very sorry your baby has spina bifida and the next words out of her mouth were, so next time take more folic acid!.”*  
(Parent / Guardian)

*“My brother has a girlfriend at the moment who’s moved in and all I want to do is tell her, take folic acid and it’s too forward you know what I mean!”* (Parent / Guardian)

*“One of the local (professionals) said well in the future you can get an increased dose of folic acid, you should be taking 20 times the amount so I took her advice, a young trainee therapist on board and all these professionals never said it.”* (Parent / Guardian)

## Discussion

All women of childbearing age should be taking 400 µg of folic acid periconceptually however compliance with supplemental folic acid intake in Ireland is poor and voluntary food fortification can be hard to monitor. Mandatory food fortification with folic acid is common in other countries (EUROCAT 2014).

## Recommendation:

1. There is a need for increased public awareness about spina bifida, pre-conceptional care, folic acid fortification and supplementation in Ireland to reduce anxiety on receiving a diagnosis of spina bifida and to support the families and individuals to be seen as part of their community.



## ***Stigma and Participation***

Difficulties with public awareness and attitudes regarding disabilities have often been discussed in the literature. Spina bifida is a complex physical disability which is often misunderstood by the general population. Gannon and Nolan (2005 p.12) define social inclusion as “being in a position to participate fully in the life of the society one lives in”. Access and inclusion in society is of vital importance to ensure active participation in life which is essential to ensure a good quality of life (WHO, 2001). The Education for Persons with Special Educational Needs (EPSEN) Act (Government of Ireland, 2004 p. 6) defines Special Educational Needs (SEN) as a restriction in ones capacity to “participate in and benefit from education” on account of an enduring disability which results in them learning differently. National and international legislation support the inclusion of pupils with SEN in mainstream schools (DES, 2007).

Most people acknowledged that while public attitudes towards disability, including spina bifida, has improved in recent years, there is still a long way to go to reduce stigma, ignorance, nervousness and to increase public awareness and understanding of the condition and its prevalence.

*“Things have come a long way... but there’s still a long way to go, there’s still a lot more, just the whole area of disability to be recognised and that whole being a second class citizen is still out there and that has to be removed just through more awareness of people with disability and their acceptance”*  
(Parent Guardian)

*“I think a lot of it is nervousness in people and I don’t know it’s very difficult to know or to break it down. If people look a bit different they’re seen as being different, people have to get it into their head that they are just people like you and I who want to do the things”*  
(Parent Guardian)

*“Maybe it is ignorance, maybe people don’t want to know until it comes to their own door, I don’t know but, you know, they are just people who are the same as us who want to do as much” (PG)*

*“I suppose, people’s attitudes are a barrier, I mean unreal, like the attitude of some people”. (Parent Guardian)*

Education and increased inclusiveness were highlighted as potential ways in which the stigma may be reduced for these children and to allow them to be accepted and partake more fully in society

*“I suppose maybe information sessions for parents and children  
how to normalise it really” (Service Provider)*

*“And I think that the profile of these children needs to be raised really  
in terms of planning for the future - yes definitely” (Service Provider)*

*“And I think one of the things that I have always felt is the whole  
concept of looking after these children in the context of their educational  
and social systems as well, that they have to be joined. And in a  
way, we have to work a lot harder in terms of integrating these children  
and making them feel that because, even if they have a disability that  
is actually not going to cause them to feel any different or to feel in  
anyway ostracised in their own community because I have seen that  
many many times” (Service Provider)*

*“A child with a physical and sensory disability needs to be part of their  
community and needs to be supported in the community”  
(Service Provider)*

## **Discussion**

Parents, children and families still face difficulties with stigma, exclusion and prejudice due to their physical disability. Irish law states that children with special educational needs should be able to actively participate in education and where appropriate they should be included in mainstream school (DES, 2007, Government of Ireland, 2004). It is of vital importance that all service providers and the general public help support families become fully integrated within their community and society. Education of parents about the full spectrum of spina bifida as a condition to the general public is necessary.

## **Fighting for Services and the future**

Parents and service providers in interview both noted the significant parental burden to fight for services, for appropriate accessible housing and the coordination of communication between service providers.

*“You have got to realise that your child is your priority and theirs is theirs and you have to fight your own battle, because if you don’t, they’ll just take it off you” (Parent Guardian)*

*“Everybody has to fight for their own and if you don’t fight you get nothing, it’s given to those who do”(Parent Guardian)*

*“Families are having to fight so so hard for medical cards, for grants, for wheelchairs, I just think life is hard enough for them without that battle as well”  
(Service Provider)*

## **Discussion**

It is important that parents’ psychological well-being be prioritised as part of the care of a child with spina bifida from the early stages to assist with their coping (Vermaes et al., 2005, Cleve, 1989). Benefits of parental support from spina bifida groups have been reported (Cleve, 1989) which is also supported by parent if this study who reported a positive and strong social impact of peer support groups for their families. This support network can provide opportunities for friendship, burden sharing and may help parents acknowledging their child’s future needs

Caregivers can experience high levels of objective burden (assisting with daily living, transportation etc.) and subjective burden (worry, reduced care giver well-being) (Angold et al., 1998). Additional and significant parental burden were described in relation to having to fight for services as well as coordinating and communication between service providers. Improved co-ordination of care and communication pathways requires development to ease this additional avoidable parental burden.

## **Recommendation**

1. Improved coordination of care and communication is required to reduce parental burden.

# **CHAPTER FIVE**

## **HEALTH SERVICES TO**

### **SUPPORT CHILDREN**

#### **WITH SPINA BIFIDA**



The health care needs of children with spina bifida are complex. They need specialists, generalists, and an integrated system to deliver this complex care (Liptak and El Samra, 2010).

The local service plays a vital role in the lives of children with spina bifida, providing medical, therapy, and community support at a local level. The value of having a primary provider who coordinates and assures quality of care for children is viewed as a central component of health care, particularly for children with special health care needs (Christakis et al., 2003).

The historical establishment of primary care paediatric teams and where these services are accessed are described in the Background chapter 1. Children with spina bifida are referred to these local primary health services within their area following discharge from the tertiary hospital. Professionals may work individually or as part of local multidisciplinary teams (MDTs).

Children and adults with spina bifida also require frequent access to specialist tertiary services and this need continues into adulthood with hospital admission rates being 9 times higher for adults with spina bifida than the general population (Young et al., 2005). Since 2008 TSCUH has been the national neurosurgical centre for children aged 0-6 years and it also provides

on-going multidisciplinary care for children with spina bifida aged 0-16 years (Committee to Review Neurosurgical Services, 2005). The fact that all children with spina bifida born since 2008 in Ireland now attend one national centre for inpatient and outpatient specialist care is unique in northern Europe.

A comprehensive, integrated and appropriately-staffed specialist service is required to meet the needs of children with spina bifida due to complications specific to spina bifida ranging from shunt complications, renal impairment, and postural deformities to obesity. Specialist disciplines such as Neurosurgery, Urology and Orthopaedics monitor complex medical needs, ideally as part of a specialised multidisciplinary team (MDT). The latter is required to coordinate and manage the care of these children with efficient communication and appropriate transition planning at major transition stages (Merkens, 2006). Pathways of care should be established between specialist and local services in a shared care approach to try and provide as much care close to the child's home as possible with support from specialist centre (NHS., 2011).

## **5.1 LOCAL SERVICE**

### ***Teams and Access***

#### **Background:**

The Spina Bifida Association of America (SBA) set out guidelines for the care of individuals with spina bifida. They stipulate that all persons with spina bifida should receive on-going, comprehensive, coordinated care throughout the lifespan that are easily accessed (Merkens, 2006). The Neurological Association of Ireland's standards of care for people with neurodisability in hospital and community state "Following diagnosis people with neurological conditions should be put in contact with a specialist multidisciplinary team in their area" (Neurological Alliance of Ireland, 2002).

The aim of a local primary service is to have a service that is readily available to all regardless of location or diagnosis. It should be staffed with multidisciplinary professionals to address assessment, diagnosis and intervention at a local level. Previous studies have highlighted concerns in disparities of access to services based on the geographic area, the age of the child, and types of disabilities or developmental concerns. Regional and county variations in referral/access criteria currently exist (Carroll et al., 2013).

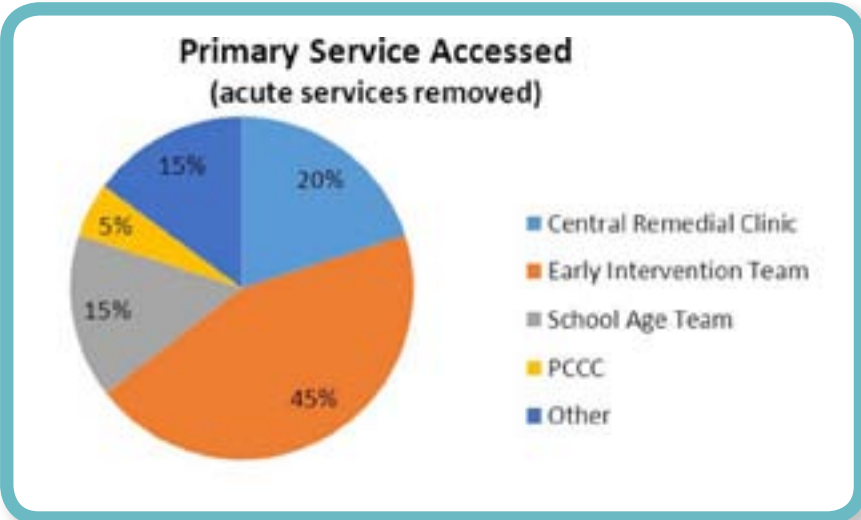
This section aims to investigate the availability of current services for children with spina bifida at a local level from the perspectives of families and service providers nationally, as well as to gather the experience and recommendations of service providers working with this client group within the community as well as parent / guardian expectations regarding their child's local

health and therapy needs.

Results

Team Configuration

Figure 57



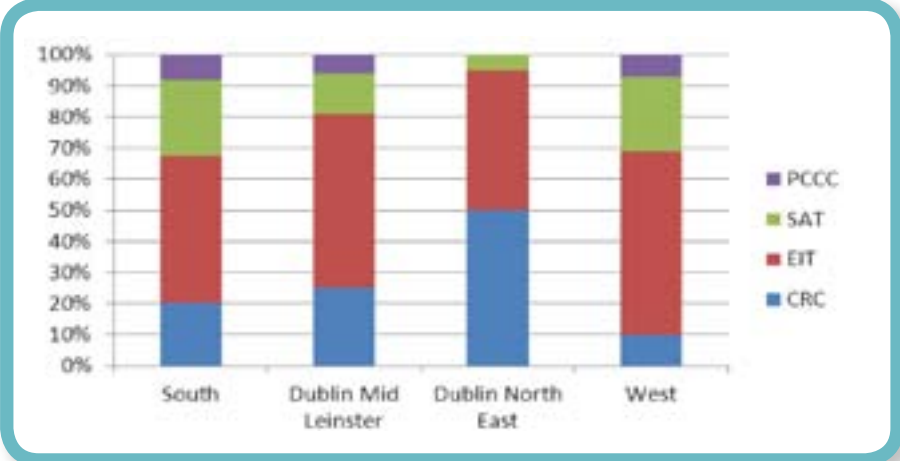
Parents were asked to indicate the primary service from which their child accessed services locally. The largest proportion of children were attending Early Intervention Teams, (45%) which generally provide services for children under 6 years. The Central Remedial Clinic was the primary service for 20% of the sample, followed by school aged teams (15%) who provided

services for children of school age (5/6 – 18 years) throughout the communities. ‘Other’ was representative of charitable services such as the Midwestern Spina Bifida Association. A large proportion of children were accessing early intervention teams which is reflective of the young age of our sample (mean age 5.7 years).

Geographically, there were differences noted in where children access services by address; Children in Dublin North East more likely to attend CRC, with no child accessing the PCCC in this area.

Figure 58

Geographical differences in type of team attended



In interview, both parents and service providers agreed on the benefits of early intervention and the intensive input required during this stage. The availability of staffing can impact on the

service delivery. Although parents expressed disappointment when the frequency of therapy reduced for older children, service providers feel this reduction was based on needs.

*“She was seen very very regularly at the start, but because she’s doing so well, she hasn’t got an actual physio session in months and I mean it could be 8 months” (Parent / Guardian)*

*“A lot of the intervention is up till they start school and I would see them less and less then. So it’s all that intervention until they are settled in school” (Service Provider)*

Nearly all service providers and parents commented on the positive benefits of having a multidisciplinary therapy team/service to aid team decision-making, joint working, better communication, ease of access, better relationships and peer supports. Difficulties were raised when there were separate teams or uni-disciplinary services which were not co-located impacting on liaison/coordination. When MDTs were not fully staffed or missing integral team members this caused frustration for both parents and service providers.

*‘Well clearly it’s a fantastic advantage to have your team on site with you for informed discussion as well as formal meetings’ (Service Provider)*

*“You have to work as a team. And you are expert in your field, I’m expert in my field, you are in yours. And if you put all the three things together, you get much better results”  
(Service Provider)*

*“What we do is, we have the assessments in the morning. And then the afternoon we have the multi-disciplinary team so we all sit down and discuss the case” (Service Provider)*

Parents and service providers both discussed the value of specialist services specific to physical disability or spina bifida rather than generic geographical teams. Parents expressed anxiety, upset and confusion regarding the reconfiguring of services in some areas in line with the ‘Progressing Disability’ strategy as set out in the National Service Plan (HSE., 2010). This strategy aims to reconfigure existing children’s disability therapy resources to generic



geographically-based teams. Services will no longer be 'diagnosis-driven' and clients will be reviewed by need 'whether they have autism, spina bifida, cerebral palsy'. This process was underway during the interview stages, and both service providers and parents expressed concern regarding the impact of this on their frequency of access to therapies. Service providers reported concerns about the future lack of specialism, possible increased waiting lists and changing roles required to work with various disabilities (e.g. ASD).

*"I'm a bit scared now at the moment the way they are amalgamating services and thinning out the services, I think that's going to be a disaster."  
(Parent / Guardian)*

*"I'll be seeing a lot less children with spina bifida [following amalgamation of service]. It will be hard I suppose initially to see where a child with spina bifida will rank in prioritisation." (Service Provider)*

*"I think is gone backwards. I mean we need centres of excellence, we need therapies of excellence, we need better, I mean you cannot be expert on all fields...and parents, I mean I don't mind to travel to get the right guy."(Service Provider)*

Results indicate that therapists locally do not have many children with spina bifida on their active caseloads. On average the physiotherapists had 4.3 children with spina bifida per caseload, Occupational Therapists had 2.8 children with spina bifida per caseload, Speech and Language therapists with 3.5 children with spina bifida per caseload. Some therapists may have had higher numbers on their caseloads as they worked in specialist or spina bifida specific services. Although variations exist in caseloads, in order to get an example of caseload size, the College of Occupational Therapists Consultancy Service 2013 recommends having no more than 50 children on an active caseload. Therefore the children with spina bifida represent about 5% of a therapist's active caseload. This demonstrates that children with spina bifida are only a very small proportion of their large overall caseloads, and some comment this can impact on level of specialisation and amount of priority they may get of these therapists caseloads.

## Access to Professionals and Frequency of Review

The availability of professionals within local teams can vary. As set out in the Health Strategy of 2001 (DoHC., 2001) children should have access to the required professionals close to home in order to provide all services for the child. Service providers were asked to indicate the availability of professionals within their teams. The most frequently available team member was an Occupational Therapist (98% of teams had an OT present), followed a Physiotherapist (97%). Ninety four per cent had a Speech and Language Therapist, 88% had a Psychologist, 83% had a Paediatrician, and 86% had a Social Worker (see figure 58). There appears to be six professionals more frequently available within local teams as represented in the chart below left of the red line, nurses, dietitians, family support workers and orthotists were less frequently available on community teams. Although psychology was present to some capacity on most teams, comments related to busy assessment caseloads with limited clinical psychology intervention.

Figure 59



Forty three per cent of service providers report that there is no waiting list for the first appointment following referral with an additional 43% reporting the wait is between 0 and 3 months. Therefore only 14% of children are waiting more than 3 months for initial access to service. Service providers did not report any significant difference in waiting times based on HSE region

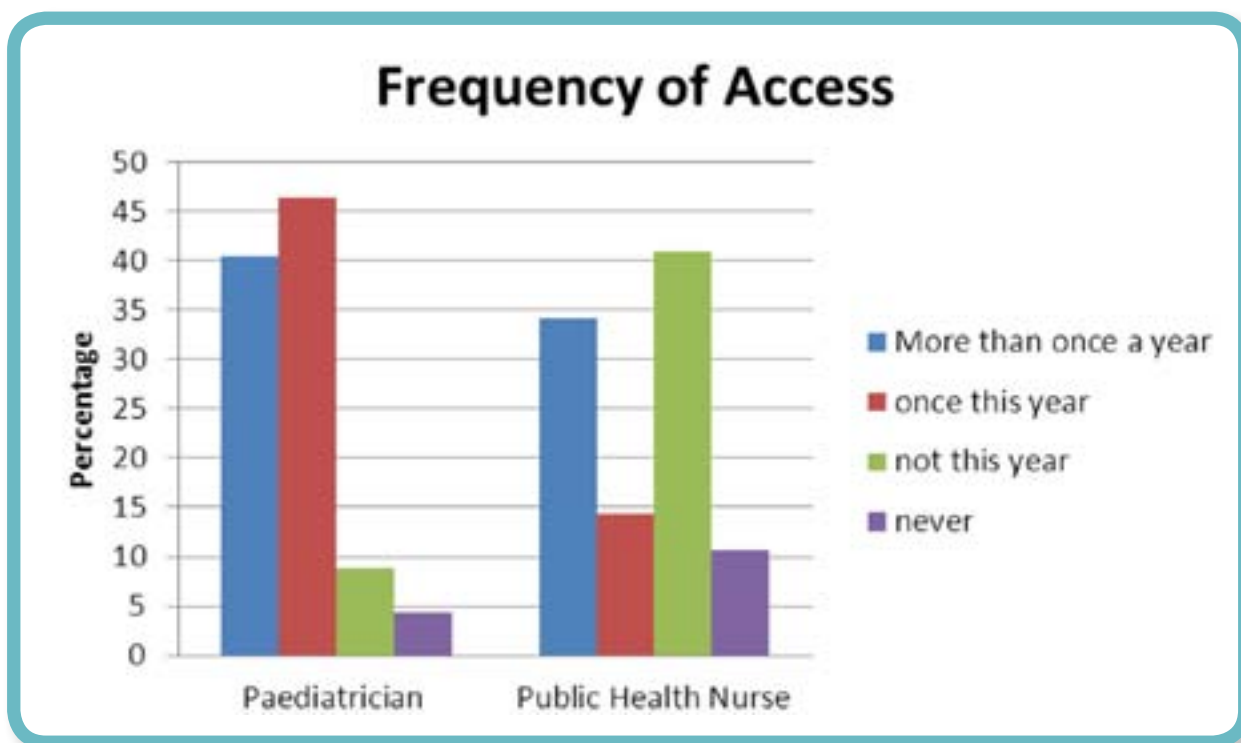
Parents indicated the frequency at which their children accessed a range of local professionals, namely paediatricians, nursing, social work, family support worker, orthotist, physiotherapist, occupational therapist, speech and language therapist, dietitian and psychologist. They were asked the frequency their child had seen that professional within the

last year or to indicate never if their child had never been reviewed by that professional.

## Paediatrician and Nursing

From a medical perspective, a very high proportion (87%) had seen a paediatrician in the previous year, and only 4% had never accessed a paediatrician. Nursing professionals within the local community setting are frequently in place to support families and children with spina bifida. The Public Health Nurse (PHN) was the most frequently accessed nursing support at a local level. 48% of parents report they had seen a PHN at least once in the last year, however 11% indicated that they had never seen a PHN. Some early intervention teams had nursing support within their teams to support early child development; however this was inconsistent across areas and not reported on.

Figure 60



Parents reported that access to a team and nursing support locally was most needed in the period following the initial discharge from tertiary hospital. Parents and service providers indicated that children were accessing and seeing these local professionals in a very timely manner post-discharge from hospital, even some being aware of a referral before the child was born.

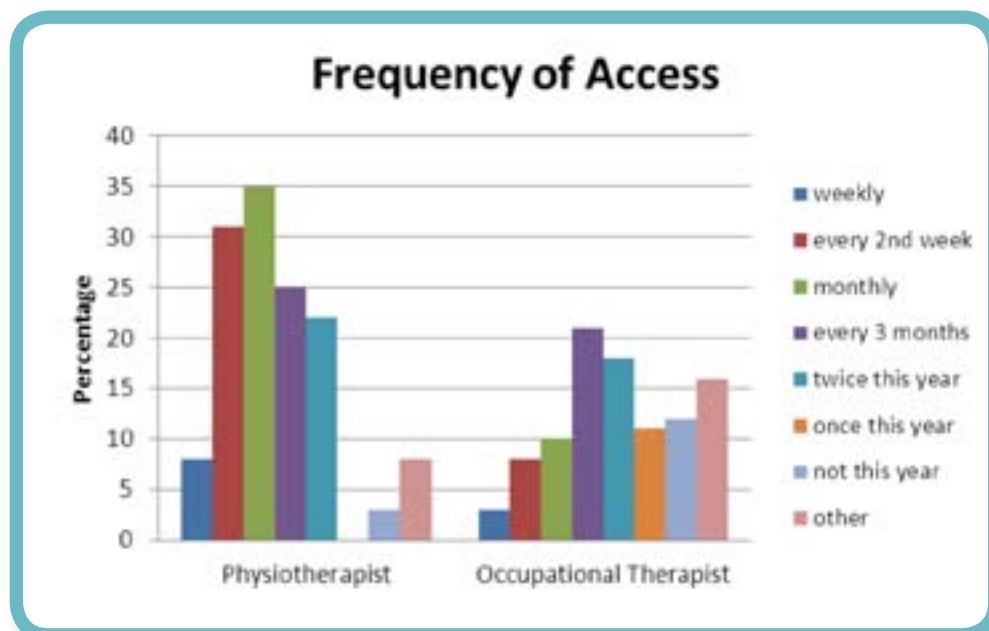
*“She was lucky, she was seen quite early, she was on a team at an early stage.” (Parent / Guardian)*

*“In the recent years we could even have a referral before they are born.” (Service Provider)*

## Physiotherapy and Occupational Therapy

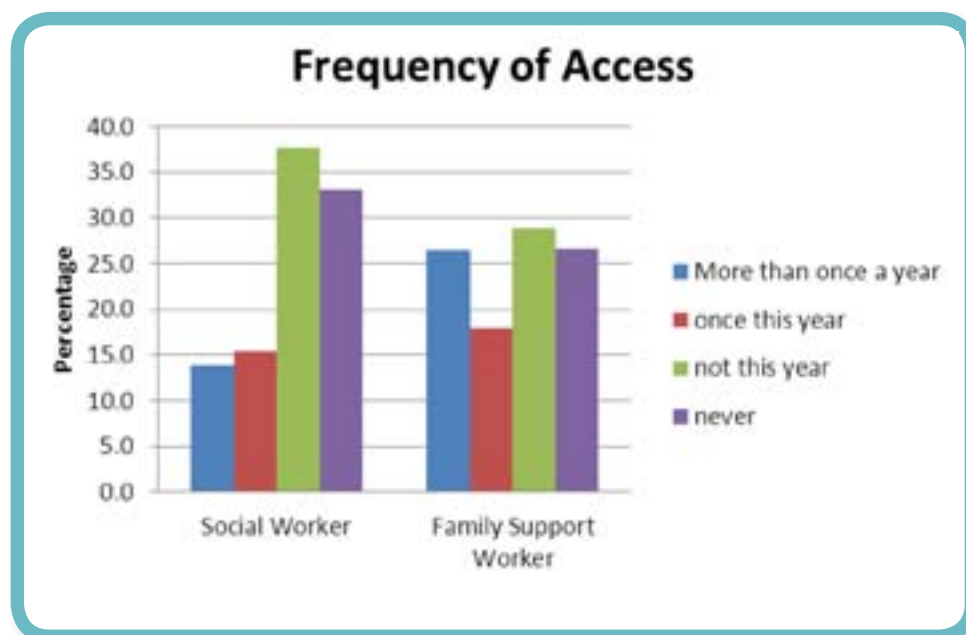
Therapy plays a vital role in the management of children with spina bifida in order to in order to maximize function, minimize complications and improve participation and quality of life (Brown, 2001, Parkes et al., 2004, Bax, 2001, Cottalorda et al., 2012). The therapists mostly frequently accessed are Physiotherapist (96% of children accessed Physiotherapy with the highest portion of children seeing physiotherapy monthly; Occupational Therapist (87% of children accessed Occupational Therapy with the highest portion of children seeing Occupational Therapy every 3 months). Of note, many parents reported accessing Physiotherapy and Occupational therapy in block sessions (4-6 appointments in succession, followed by a break) which is represented as ‘Other’ in Figure 60. Further detail in relation to physiotherapy and occupational therapy services and interventions will follow in the occupational therapy and physiotherapy sections.

Figure 61



## Social Worker and Family Support Worker

Figure 62



Due to the complexity of these children, social support is often required to support the family's needs (Merkens, 2006). Large numbers of families within the current study were not receiving on-going support from social work (70%) or family support workers (56%) who can provide a vital support role. Family support

workers are available through the spina bifida associations; however parents are required to initiate this contact.

Despite timely access to services, parents reported negative feelings of fear and lack of support during that period following initial discharge home from hospital. Early support by a local team was deemed very important. However many reported limited practical and emotional supports to manage at home in the early stages, requesting further support from nursing and medical professionals.

*"I suppose it was crisis management the first two months or whatever, settled down a bit." (Parent / Guardian)*

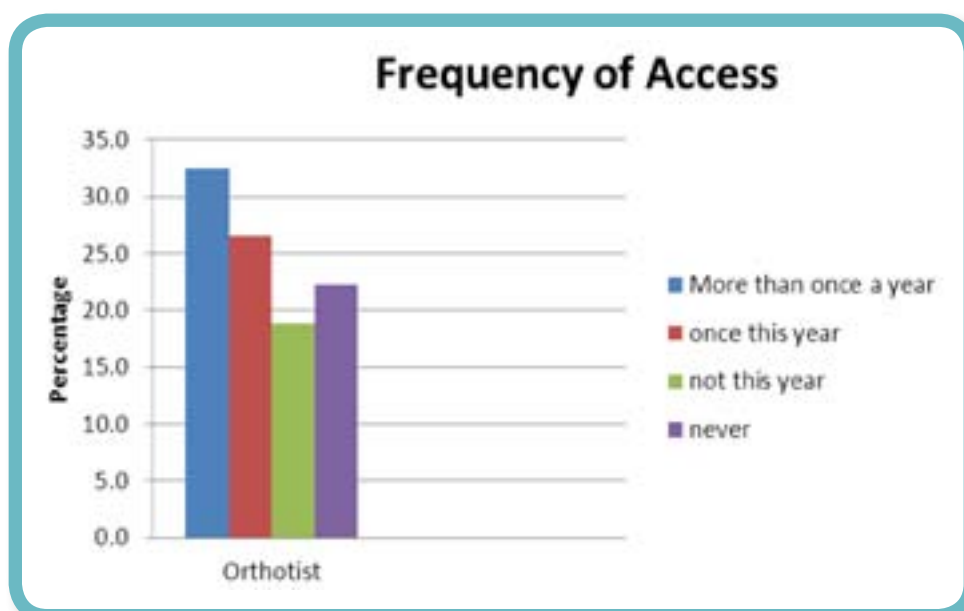
*"Support wise, when in the hospital the support was as good as possible. But when you went home, there wasn't support." (Parent / Guardian)*

*"I just think we had enough to be dealing with besides having to do all of this (organising nursing care at home). And it would have been lovely for somebody to say, look you don't have to worry about that, we'll look after that end of things." (Parent / Guardian)*

## Orthotist

Due to the complexity of the physical needs of children with spina bifida, the orthotist also plays a vital role in assessing and providing suitable orthotic support to support standing and mobilizing (Brown, 2001, Carroll, 1987). As mobility has been linked to improved quality of life for children with spina bifida (Mazur and Kyle, 2004), the provision of equipment to support independence is vital. While there is evidence to support the use of orthotics to reduce energy expenditure and improve mobility, gait pattern and access to environment (Malas, 2011, Thomson et al., 1999, Bartonek et al., 2007), there is debate in relation to the benefits of complex orthotics for high level lesions. This was discussed in more detail in chapter 4.1 in the physical impact on the child with spina bifida.

Figure 63



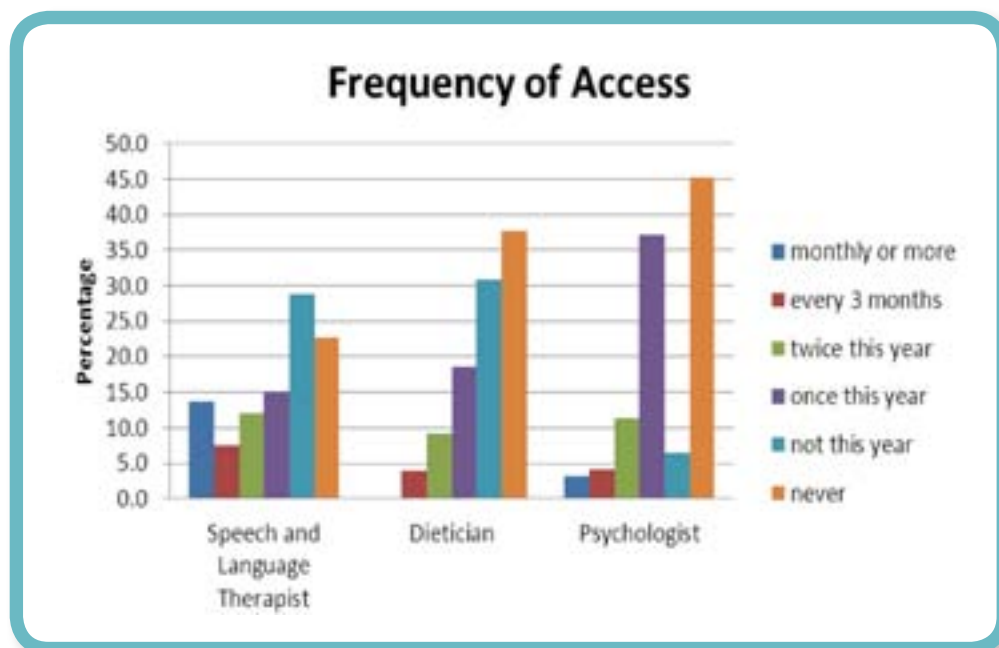
Surprisingly, 22% of parents in the current sample reported that they had never accessed an orthotist. Only 59% reported seeing an orthotist at least once a year (see Figure below). Concerns were raised by some parents that orthotists are not always part of the team, and that services are outsourced to private orthotists.

During interview one orthotist expressed concern that the HSE is cost-conscious and tendering for business is often based on cost rather than quality, suggesting that a fixed price system would allow for quality to prevail. This requires orthotic staff who are adequately experienced and trained to provide a quality service or product.

## Speech and Language Therapy, Dietitian and Psychology

Speech and Language therapy is recommended in spina bifida due to specific difficulties (Dennis et al., 2008, Huber-Okraïne et al., 2002) these have already been detailed in chapter 4.1 'Physical Impact of spina bifida on the Child'. Limited access to Speech and Language therapists was noted by parents in our sample, with 22% of the children having never accessed Speech and Language therapy and an additional 29% having not been reviewed within the previous year (Figure 63).

Figure 64



Children with spina bifida have increased risks of obesity as discussed in chapter 4.1 'Physical Impact of spina bifida on the Child' (Dosa et al., 2009a, Dosa et al., 2009b, Mita et al., 1993, Buffart et al., 2008a). They also present with additional diet and bowel-specific

difficulties leading to both incontinence and constipation (Liptak and El Samra, 2010) that would benefit from the review of a dietitian. There was poor access to dietitian noted by parents. Only 32.5% of parents reported seeing a dietitian within the last year, and 38% of children had never accessed a dietitian.

The psychologist can provide both assessments of cognitive profile as well as interventions for psychosocial / mental health difficulties. The specific cognitive and psychosocial profile of children with spina bifida will be discussed in more detail in section 4.2 'cognitive and psychosocial impact of spina bifida'. Forty five per cent of children had never accessed a psychologist, and if involved was mainly for an individual session on a once a year basis for assessment rather than on-going intervention.

*“One young boy we referred him on to the psychologist but there was a year and a half waiting list and by the time we had overcome the problem.”  
(Service Provider)*

*“[Psychologists] have a bigger caseload and then they are kind of tied up with assessments.” (Service Provider)*



## Access to Professionals by HSE Region

Next comparisons were made between the accessibility of therapy professionals in each of the four HSE Regions to see if differences occurred. Dietitians were more frequently accessed in the HSE South and HSE Dublin North East, Speech and Language therapists were more frequently accessed in the HSE South, and Physiotherapy and Occupational Therapy were most frequently accessed in the HSE Dublin Mid Leinster area. The HSE West had the least frequent access to all therapy professionals.

Figure 65

### Access to professionals by HSE region

Key	least frequently seen by	HSE South	HSE Dublin North East	HSE Dublin Mid Leinster	HSE West
	most frequently seen by				
Dietitian					
Speech and Language Therapy					
Physiotherapy					
Occupational Therapy					

Many services in Ireland provide specific blocks of therapy interventions as a means of managing large caseloads. Parents commented that the frequency of therapy was not adequate to meet the needs of their child, and expressed concern that it is delivered in blocks of interventions resulting in periods of time where there is limited review by professionals and therapy is completed at home by parents themselves:



*“You’d get a block where you’d see somebody, it might be weekly or fortnightly for those 5 or 6 sessions. And then after that, you are sort of on your own”  
(Parent / Guardian)*

*“He use to get physio every week, we are down to now, kind of twice a month. So we get an hour twice a month. And OT maybe twice a month, and then speech and language therapy maybe once a month or once every two months. So I mean I certainly don’t feel it’s enough, I don’t think an hour every two weeks is anywhere near what we need”  
(Parent / Guardian)*

*“In regards to hands on physio and OT and actually therapy for the child its very very poor”  
(Parent / Guardian)*

*“You’d be given a home programme to do which you will do at home but I mean these children deserve a better service which is to be seen weekly or fortnightly not just under review every so months to see how they’re doing, It’s not good enough.” (Parent / Guardian)*

Service providers were asked how often they currently see each of the groups of children (if at all) and how often they would recommend that they should be seen by their profession. Children were divided into 7 categories (infants, too young to tell mobility level, wheelchair most of the time, uses splints & aids sometimes wheelchair sometimes, uses splints and sometimes needs help, walks with splints and aids and independent walkers).

Overall, younger children and those with equipment dependent mobility were seen more frequently with the increased independence resulting in service providers seeing those groups of children less frequently. The table below indicates the current and recommended timeframe of the highest proportion of service providers.

Table 7

Frequency of review by age, current and recommended

	Service Providers: Current Frequency of Review	Service Providers: Recommended Frequency of Review
Infants	Every 2 weeks (34%)	Weekly (51%)
Too young to tell mobility	Every 2 weeks (44%)	Weekly (34%) - Every 2 weeks (46%)
Uses Wheelchair most of the time	Monthly (47%)	Monthly (47%)
Use splints & aids sometimes and wheelchair sometimes	Monthly (48%)	Monthly (53%)
Uses Splints and sometimes needs help	Monthly (39%)	Monthly (50%)
walk with splints and aids	Every 3 months (36%)	Every 3 months (49%)

Recommendations by the highest proportion of professionals, while not statistically significant, suggested that they would like to see infants and those children whose mobility potential had not yet been determined more frequently than is currently occurring. The literature supports intensive early therapeutic intervention to improve outcome and minimize disability in spina bifida (Shaer, 1997, Merkens, 2006).

Service providers comment that services are provided according to need, with younger children being reviewed more frequently. Service providers also commented on the fact that therapy should be available to support a specific need, not on an on-going basis.

*"I tend to meet those kids in the first year, two years of development quite often, because everything is changing so quickly."  
(Service Provider)*

*"The frequency, duration everything is based on their needs and what age they come in" (Service Provider)*

*“We’re not restricted on how much therapy we can provide from a physio point of view, it’s based on the clients needs.”  
(Service Provider)*

A large variation in wait-times, service delivery method and frequency of therapy at were commented on. These are influenced by geographic location, age, staffing levels, child’s needs, communication between clinicians and parental pressure. This can result in inequities for families and extra stress or guilt for service providers who are constantly trying to reprioritise caseloads. Increased transparency, organisation and communication with parents and a more standard method for therapy service delivery nationally were recommended.

*“There is such demand for the services that are there. And with the embargo with taking on staff, it’s difficult” (Parent / Guardian)*

*“It’s terrible that they tell you are on a waiting list, and we found out we weren’t on it at all, two and a half years later...there should be transparency.” (Parent / Guardian)*

*“It not equal and it’s meant to be equal.” (Service Provider)*

*“What would help me out is if there was more standard therapy service.”  
(Service Provider)*

## **Discussion**

Primary care which is local, accessible, multidisciplinary and coordinated is a priority for healthcare provision in Ireland (HSE 2001). The management of spina bifida requires local life-long access to services as outlined in the SBA guidelines (Merkens, 2006). Additional support in the period following initial discharge could be provided by local teams through social work, nursing professionals, special needs counsellors or by spina bifida family support workers.

The early intervention teams are the most frequently accessed for services which can be explained in part by the young sample age represented (mean age 5.7 years), and also by the agreement that therapeutic intervention in the early years is most beneficial (Shaer, 1997). Key professionals present in local teams include Physiotherapists, Occupational Therapists, Speech and Language Therapists, Psychologists, Paediatricians and Social Workers. These findings are supported by other research recommending the need for multidisciplinary teams locally

(Carroll et al., 2013). The access to these professionals however was infrequent particularly for access to speech and language therapy, social support, nursing, dietitian and orthotist and psychology general on an assessment only basis. Geographical differences were also noted with less frequent access to professionals in the HSE West regions.

Service providers have small numbers of children with spina bifida on large mixed caseloads supporting the need for education and advice from adequately resourced specialist therapists and counterparts at the national neurosurgical centre.

Parents would like more frequent access to therapy, and service providers would also like to see younger children more frequently, however, service providers did not recommend seeing other groups of children on a more frequent basis unless determined by a specific need. Multidisciplinary physical disability teams are preferred by both parents and service providers with a concern regarding the change in services towards geographical based paediatric teams as small numbers of children with spina bifida on caseloads may reduce specialism.

## **Recommendations**

1. Local multidisciplinary teams (MDTs) should be available for children with spina bifida across the lifespan and they should consist of at least physiotherapy, occupational therapy, speech and language therapy, psychology, social work, and paediatrics. Improved access to speech and language therapy, social work, family support workers, nursing, orthotists, psychology and dietitian is required.
2. Education and support for local MDT professionals should be readily accessible from the national specialist centre when required.
3. Improved practical, social and emotional support is required immediately post discharge from tertiary hospitals for families. This could be provided by social workers, nursing professionals, special needs counsellors or by spina bifida family support workers
4. Equitable frequency of access to therapy professionals is required based on the child and family's needs.

Physiotherapists and Occupational Therapists are two professionals who are most frequently involved in reviewing children with spina bifida and are involved in various aspects of their care. Two of the researchers involved in gathering and analysing data for this research project came from a physiotherapy and occupational therapy background. While every attempt was made to contact all professionals working with children with spina bifida nationally (see methodology and samples chapter), many of the most frequently involved team members consisted of Physiotherapists and Occupational Therapists and these professionals may have been more interested in becoming involved given the professions of the researchers. The next section describes in more detail results relating to occupational therapy, physiotherapy and equipment, which all have a large impact on the lives of children with spina bifida.

## ***Occupational Therapy***

### **Background**

Occupational Therapists work with people facing physical, mental and social disabilities to help them do the things they want to do (COT, 2014). Occupational Therapists work with children with various physical and sensory disabilities including spina bifida (Rogers et al., 2001) however little empirical evidence exists in peer reviewed journals regarding the outcome of occupational therapy specifically with children with spina bifida. In a similar review of services regarding access to paediatric Occupational Therapy services in Northern Ireland for children with Cerebral Palsy 79% of children were reported to have access (Parkes et al., 2002). Occupational Therapists can play a vital role with children with spina bifida in relation to neurological, physical, social, psychological and environmental status and how this relates to activities of daily living in order to maximise functional independence (Watson, 1991). Ryan et al. (1991) noted that an Occupational Therapist should be included on the treatment team for children with Myelomeningocele and Watson (1991) and Delmarva Foundation (2006) highlighted the importance of Occupational Therapists presence at multidisciplinary Spina Bifida clinics for children with spina bifida.

Interventions in which Occupational Therapists become involved are complex and varied. Occupational therapy has been described as a necessary tool to allow children with spina bifida to develop skills necessary for everyday activities, including getting dressed and to provide practical solutions and equipment, as this can help to boost self-esteem and independence (NHS, 2012). The role of occupational therapy and guidelines for interventions with children and adolescents with spina bifida were detailed by a Canadian clinic (Watson, 1991). This study may not be transferable as it consisted of one case review and respected opinion however the article highlights ideas about timing for occupational therapy assessments within a Spina Bifida clinic. Various OT interventions throughout the lifespan were described. The role of the Occupational Therapist was defined as maintaining health by developing or

restoring activities of daily living as well as enabling children to engage in age-appropriate occupational roles (Watson, 1991). Four main stages in the lifespan of a child with spina bifida were seen as critical for OT interventions including infancy, preschool, school age and adolescence. The role described was very wide-ranging including input to developmental assessment, feeding, working with the upper limb, oral motor development, equipment, play, school assessment, visual perceptual and visual motor integration, sexuality, social skills, independent living, transportation and pre-vocational exploration.

Where Occupational Therapists are available in school (as in the USA) they should be involved in assessing for early changes in upper extremity functioning and they or the local Occupational Therapist should remain in contact with the child's spina bifida team (Porter et al., 2009)

Equipment assessment and provision is another area of practice with children with spina bifida in which Occupational Therapists often become involved. Occupational therapy has been described as a key profession involved in the assessment and prescription of wheelchairs and seating systems (Kenny and Gowran, 2014). Many children with spina bifida require wheelchairs or specialised seating to support their occupational performance and their importance cannot be underestimated (WHO., 2008). The Occupational Therapist is often involved with toileting equipment, when necessary for functional gain as a result of poor sitting balance and the need for catheterisation (SBHI 2009). Occupational Therapists may be involved in obtaining the optimal seating position, providing adaptive equipment and assisting with motor planning and sensory integration difficulties and also working with visual perceptual function. (Sandler 1997)

Early intervention in the area of occupational therapy should be encouraged to compensate for the motor skill deficits in spina bifida and to assist with achieving developmental milestones (SpinaBifidaHQ., 2014). Feldman et al. (2008) results suggested that a negative impact on psychosocial quality of life can occur for children with physical disabilities as a result of lengthy delays in accessing occupational therapy rehabilitation services. D'Eath (2005) highlighted long waiting time for assessment for occupational therapy in Ireland and availability of occupational therapy services in any capacity were seen to be very limited and inadequate to meet the needs of people with disabilities. Similarly Buran et al. (2002) reported that this worsens in adolescence with the majority of adolescents reporting not receiving occupational therapy at home or in school.

## **Results**

Seventy eight Occupational Therapists responded by questionnaire, 84% were female with an average of 8.1 years of experience. The mean numbers of children with spina bifida on their caseloads per Occupational Therapist was 2.8.

Table 8

Occupational Therapists - gender, experience and caseload

Occupational Therapists Gender, Experience & Caseload		
Males % / Females %	Years of experience	N caseload
16% / 84 %	8.1	2.8

The services where Occupational Therapists worked are presented in the figure below. The highest proportion of Occupational Therapists worked in Early Intervention Teams, followed by School aged Teams and primary continuous and community care teams. They also represented a national geographical spread across the 4 HSE regions with a small proportion providing a national service.

Figure 66

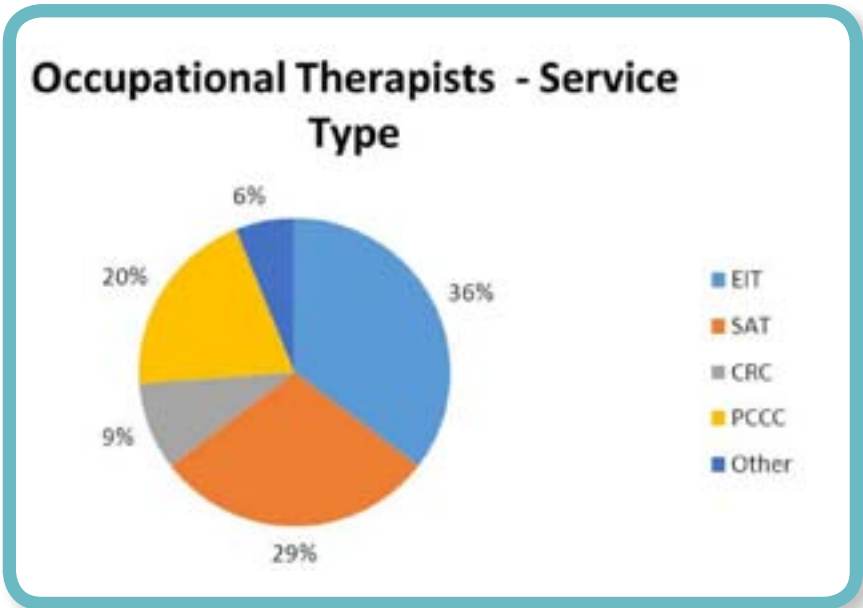


Figure 67



The majority (87%) of parents indicated that their child had access to Paediatric Occupational Therapy. Of the 13% (n=19) that indicated that their child did not have access, 10 parents reported that the service was not required for their child, 4 were awaiting review and an additional 4 reported that Occupational Therapy was not available to them. The highest proportion of parents reported seeing Occupational Therapist every 3 months, with a large spread from weekly all the way to less than once a year. 16% of parents indicated ‘other’ which is representative of blocks of therapy throughout the year rather than on-going intervention at regular intervals.

Figure 68



The services where the children reported accessing Occupational therapy are listed in the tables below according to service type. The highest proportion of children access Occupational Therapy through Enable Ireland services nationally, followed by the HSE and CRC.

Table 9

Service type occupational therapists work in

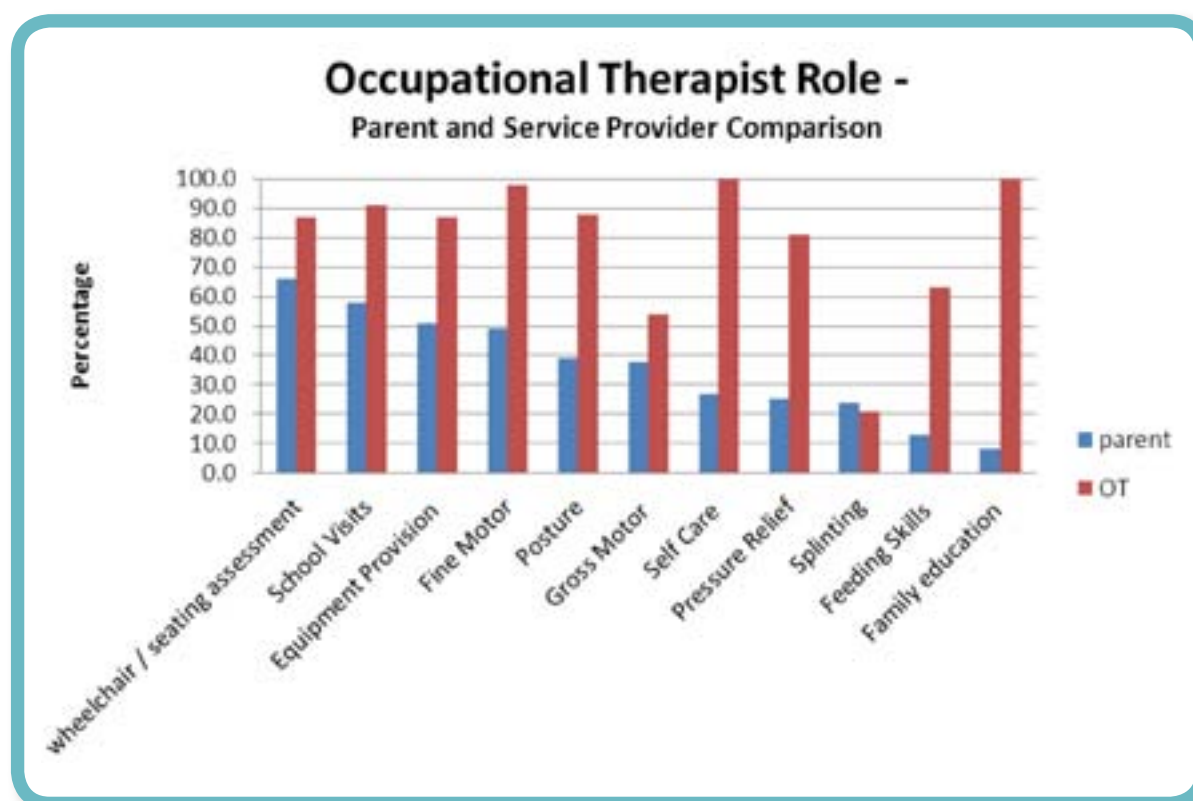
Service Type Occupational Therapists work in	Per cent
Enable Ireland	44.7
Central Remedial Clinic	19.5
Health Service Executive	27.6
Acute Hospital Service	4.9
Charitable Organisation	2.4
Other	0.8



The main interventions performed by Occupational Therapists are outlined below from the perspective of parents and Occupational Therapists themselves. Although some discrepancy existed between parents and Occupational Therapists perspectives, a large range of interventions were reported by both. The most commonly reported interventions by parents were wheelchair / seating assessments (reported by 66% of parents), school visits (reported by 58% of parents), equipment provision (reported by 51% of parents) and fine motor skills (reported by 49% of parents).

Occupational Therapists reported being most frequently involved in self-care (reported by 100% of Occupational Therapists), Education (reported by 100% of Occupational Therapists), Fine Motor (reported by 98% of Occupational Therapists), school visits (reported by 91% of Occupational Therapists) and posture (reported by 88% of Occupational Therapists). Areas of intervention that were mentioned by Occupational Therapists but not by parents were transfers, mobility, strengthening, learning and attainment and communication. The biggest areas of discrepancy were education, self-care, pressure relief and feeding skills. Further clarification of the role of Occupational Therapy should be explored with parents to understand the relevance of all interventions performed.

Figure 68



Over half of the Occupational Therapists indicated that there was no waiting list to access services. However in interview Occupational Therapists in some areas indicated that they have high caseloads and up to 3 years waiting lists.

*“In the early intervention I’m the only one person dealing with 180 case” (Service Provider)*

*“About 3 years, 3 and a half years [to access OT], so unfortunately anyone with spina bifida comes down and they are asked, they have to be waitlisted” (Service Provider)*

Home visits were performed more frequently than school visits, and only small numbers indicated that they would never review a child at home or school. Home / school visits are very important for this client group to ensure they have the appropriate equipment and set-up to access their environment.

Figure 70

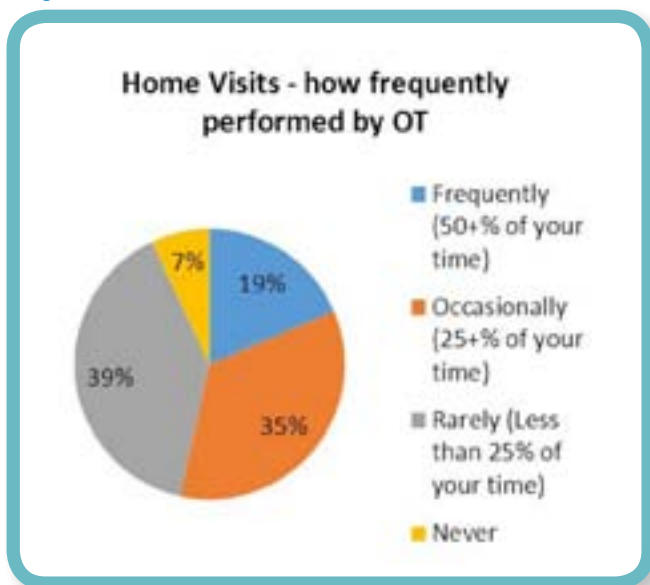
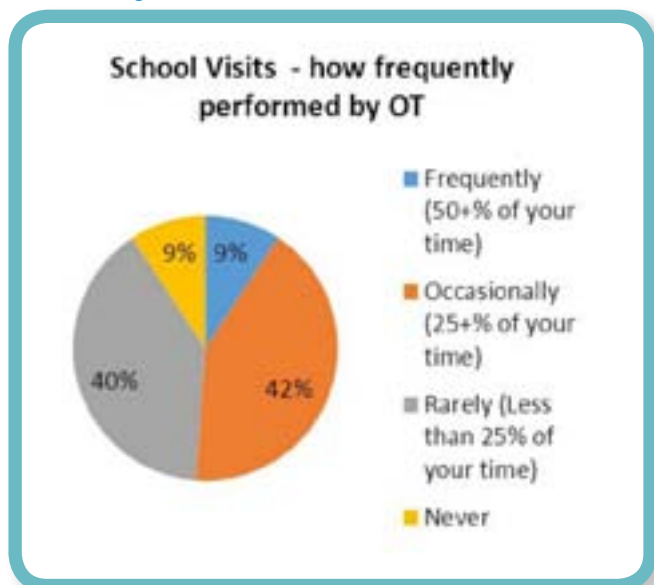


Figure 71



In relation to satisfaction with Occupational Therapy services received, parents reported greater satisfaction with quality than they did with availability.

Figure 72

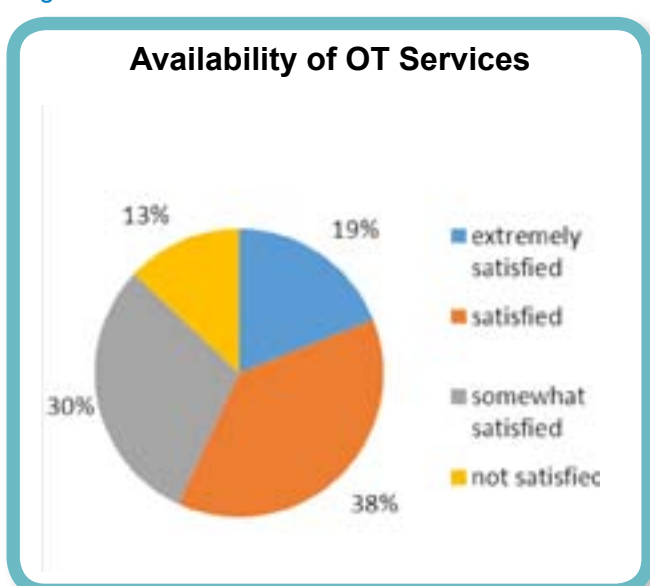
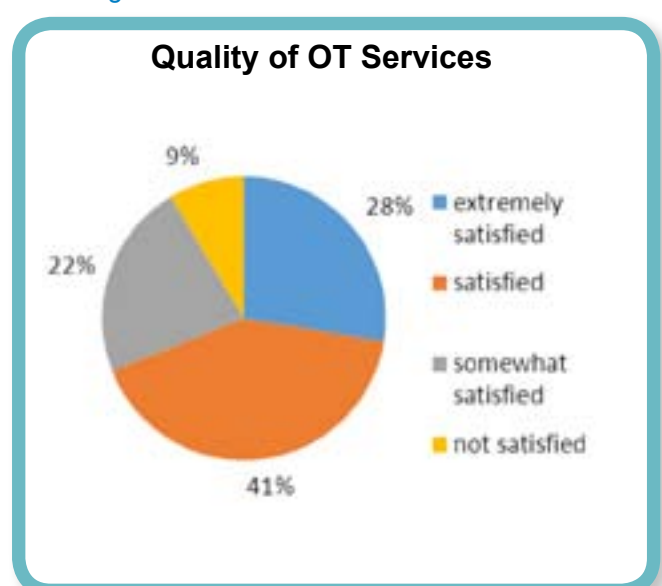


Figure 73



Reasons for dissatisfaction were discussed to often be in relation to the perception that sessions were used for equipment review, repair and provision rather than therapeutic intervention, and that sessions are scheduled in blocks of sessions with periods of time without direct occupational therapy input. Parents should be informed of the plan for service delivery once accepted to their local service to reduce frustration and dissatisfaction.

*“We get occupational therapy only if I ring and say there’s a problem with a wheelchair” (Parent / Guardian)*

*“Amazing, I have to say, I couldn’t speak more highly of them. They have been a great support” (Parent / Guardian)*

*“The OT, she’s fabulous.” (Parent / Guardian)*

*“We went through a period of not having OT, we had no OT for six months. And it was a period of time where I felt he could really do with it. We have had a diagnosis of sensory processing disorder as well, and he has an awful lot of food issues...and we were going through all this and had no OT. It was difficult.” (Parent / Guardian)*

### **Occupational Therapists frequency of review:**

When asked about frequency of review, the highest proportion of Occupational Therapists indicated that they would see infants monthly, whereas they would recommend they be reviewed every 2 weeks. Wheelchair-users are most frequently reviewed monthly which is consistent with their professional recommendation. Children who mobilise with splints are currently not seen by the highest proportion of Occupational Therapists, however they recommend that they be reviewed every 3 months. Similarly independent mobilisers are currently not seen by the highest proportion of Occupational Therapists, and again they would recommend that they should be reviewed every 3 months.

Overall, as indicated in table 10 currently Occupational Therapists see younger and wheelchair-users more frequently than others. They would like to see younger children and ambulant children more often.

Table 10

Frequency of review by Occupational Therapy current and recommended

	Currently	Recommended
Infants and Pre-mobilisers	Monthly	Every 2 weeks
Wheelchair users	Monthly	Monthly
Mobile with splints	Not seen	Every 3 months
Independently mobile	Not seen	Every 3 months

The overall priorities for Occupational Therapy are outlined below, with Equipment, Mobility, Pressure Relief, Psychosocial and self-care listed as the highest priorities.

Figure 74



## Discussion

While there is a high prevalence of spina bifida in Ireland, because these children are evenly distributed throughout country, most Occupational Therapists only have small numbers on caseload. It is very important therefore that local Occupational Therapists have access and links to specialist services (e.g. TSCUH, SBHI) to access advice and education regarding the management of these children. The results support previous literature highlighting the varied & complex role of Occupational Therapists who work with children with spina bifida (Watson, 1991). This requires initially a broad understanding of the condition & co-morbidities, the developmental profile and potential issues, and areas to monitor through OT interventions.

Most of the Occupational Therapists recommended that they would like to review children more frequently than is currently occurring. To ensure this, it is vital that sufficient staffing is in place and supported to provide increased service. It is important for Occupational Therapists to consider non-equipment needs (e.g. visual, self-care, psychosocial). Many Occupational Therapists are not resourced to provide a service for children who are mobile and those without splints and as children get older many studies find their needs increase while funding decreases (Johnson et al., 2007). Frequency of review should be based on Occupational Therapist's assessment and clinical recommendation of need rather than prescriptive blocks of therapy.

## Recommendation

1. Increased frequency of provision of occupational therapy services is recommended based on therapists' assessments and recommendations and also the child's needs rather than prescriptive blocks of therapy or pre-determined time frames throughout the year.

# ***Physiotherapy***

## **Background**

Paediatric physiotherapy is a key service in the management of children with developmental disabilities, aiming to improve motor function, prevent complications and deterioration in child's condition (Parkes et al., 2004). Mobility, posture and flexibility are important aspects of physical and orthopaedic care in children with spina bifida. Common complications include frequent fractures, joint contractures and deformities ((Brown, 2001, Dosa et al., 2007). Brown (2001) promotes early intervention of physiotherapy to promote range of motion, flexibility and ambulation as well as occupational therapy to develop motor skills, balance, body awareness and promotion of independence. Early physical evaluation and assessment are recommended (Cottalorda et al., 2012). There is a consensus that physiotherapy should remain a basic right for children with physical disabilities (Bax, 2001).

Many papers have reported on the physical and developmental delay experienced by children with spina bifida. Lomax-Bream et al. (2007) noted that children with spina bifida scored significantly lower in both the cognitive and motor scale of the Bayley development assessment than typically developing children, with significantly lower motor abilities in higher level lesions and those with a shunt in place to manage hydrocephalus. Children with spina bifida also scored lower on the Paediatric Evaluation of Disability Inventory (PEDI) as a functional measure of ability than did normally developing children, and the scores showed a significant correlation with their clinical neurological levels, walking ability, intelligence, and bowel and bladder functions. The PEDI was therefore identified as a valuable functional performance measure for children with spina bifida (Tsai et al., 2002). The presence of hydrocephalus is a significant factor in the prediction for functional independence in adults with spina bifida as demonstrated by the Functional Independence Measure (Verhoef et al., 2006)

Reduced strength and endurance even in sacral level high-functioning children with spina bifida suggest the need for on-going training and exercise programmes (Schoenmakers et al., 2009). A systematic review of the effects of muscle strengthening programmes in children with spina bifida by Dagenais et al. (2009) concluded that muscle strength can be increased by electrical stimulation, exercise training or motor skills training by physiotherapists. Further research is warranted to demonstrate any carry-over of strength gain into function.

This level of access is in line with a similar review of paediatric physiotherapy services in Northern Ireland who also reported that 96% of children with cerebral palsy had access to paediatric physiotherapy (Parkes et al., 2002).

Results

One hundred and one Physiotherapists completed the questionnaire detailing their service, experience and recommendations for children with spina bifida. 89% of Physiotherapists were female with a mean of 9.2 years’ experience. On average each Physiotherapist had 4.3 children with spina bifida on their active caseloads.

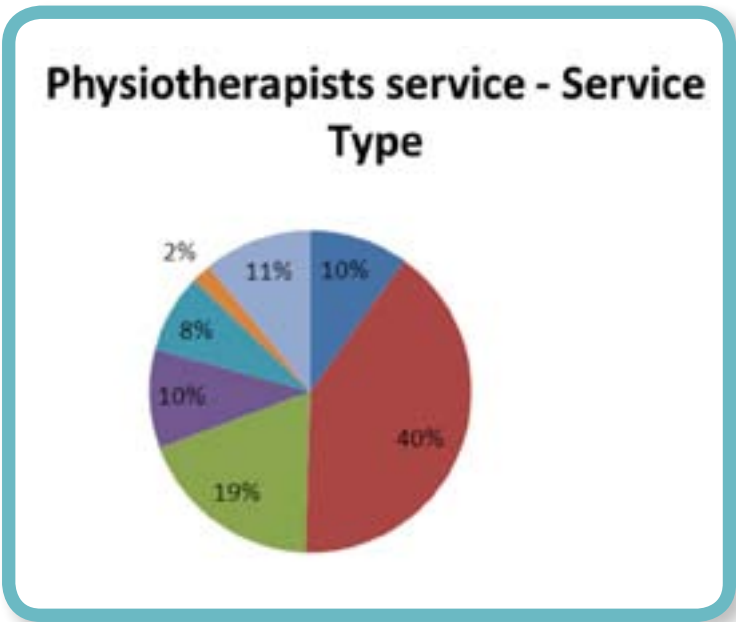
Table 11

Physiotherapists - gender, experience and caseload

Physiotherapists Gender, Experience & Caseload		
Male% / Female%	Years of experience	N caseload
11% / 89%	9.2	4.3

The services where Physiotherapist worked are presented in the chart below. The highest proportion of Physiotherapists worked in Early Intervention Teams (40%), followed by school-age teams (19%). They represented a national spread from all four HSE regions.

Figure 75



Overall, 96% of parents reported that their child had access to a paediatric Physiotherapist. Only 4% of the children did not have access (n=6), and of these 3 reported that this service was not required by their child, only 1 indicated that they were waiting for this and another 2 reported that the service was not available.

The highest proportion of parents reported seeing a physiotherapist on a monthly basis, with the greatest spread attending from every 2 weeks to every 6 months.

Figure 76

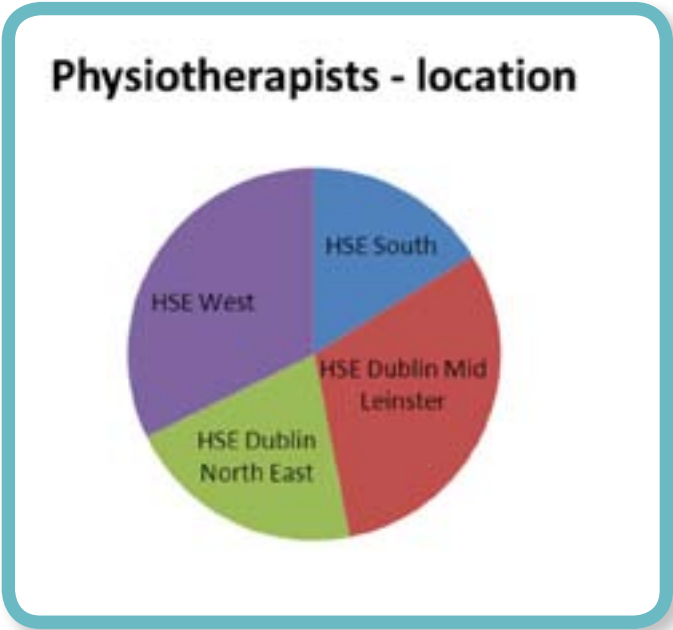
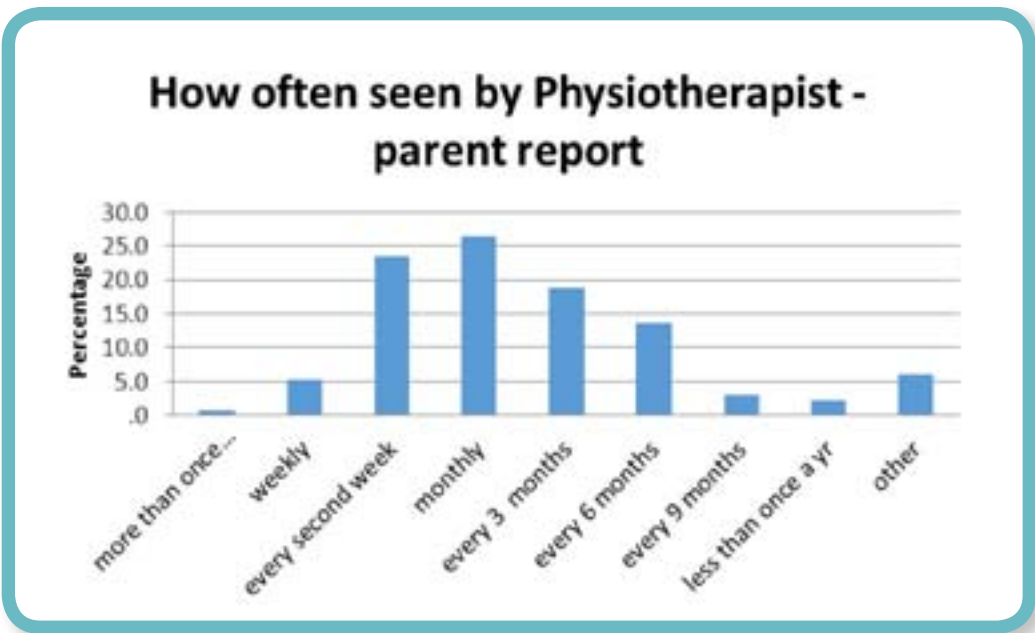


Figure 77



The services where the parents reported their children accessed physiotherapy services are listed below by service type. The highest proportion of children accesses their services through Enable Ireland services nationally, followed by the HSE and the CRC.



Table 12

Service type physiotherapists work in

Service Type Physiotherapists work in	Per cent
Enable Ireland	41.4
Central Remedial Clinic	17.3
Health Service Executive	28.6
Temple Street	1.5
Charitable Organisation	9.0
Other	2.3
Total	100%
Private	5.2

A few parents (5%) reported supplementing service provision by accessing private physiotherapy in the questionnaire, although more commented on this through interview. Pressure for seeking private therapy was due to infrequency of physiotherapy, the perception that increased therapy input will improve functional gain and to access interventions that were not available in their local service. Parents reported the significant financial cost of accessing private therapy with some having to fundraise privately. They also commented that they were unsure what exactly they were seeking to gain from private therapy. Goals were often not discussed and there was limited contact with local primary service due to fear of being discharged which added to anxiety. D'Eath et al. (2005) commented that private therapy can also be difficult to access due to the lack of private paediatric therapists. There is a lack of liaison between private therapists and local services which can lead to differing goals, confusion and at times unrealistic expectations.

*“I mean if you afford to pay for private services it’s out there and it does work, it definitely does but I mean it’s a privilege, that’s just the way it is”  
(Service Provider)*

*“When I could afford it [private physiotherapy], when I can get a few pound together I definitely would but like that it’s not sustainable but yeah, I have in the past” (Parent / Gaurdian)*

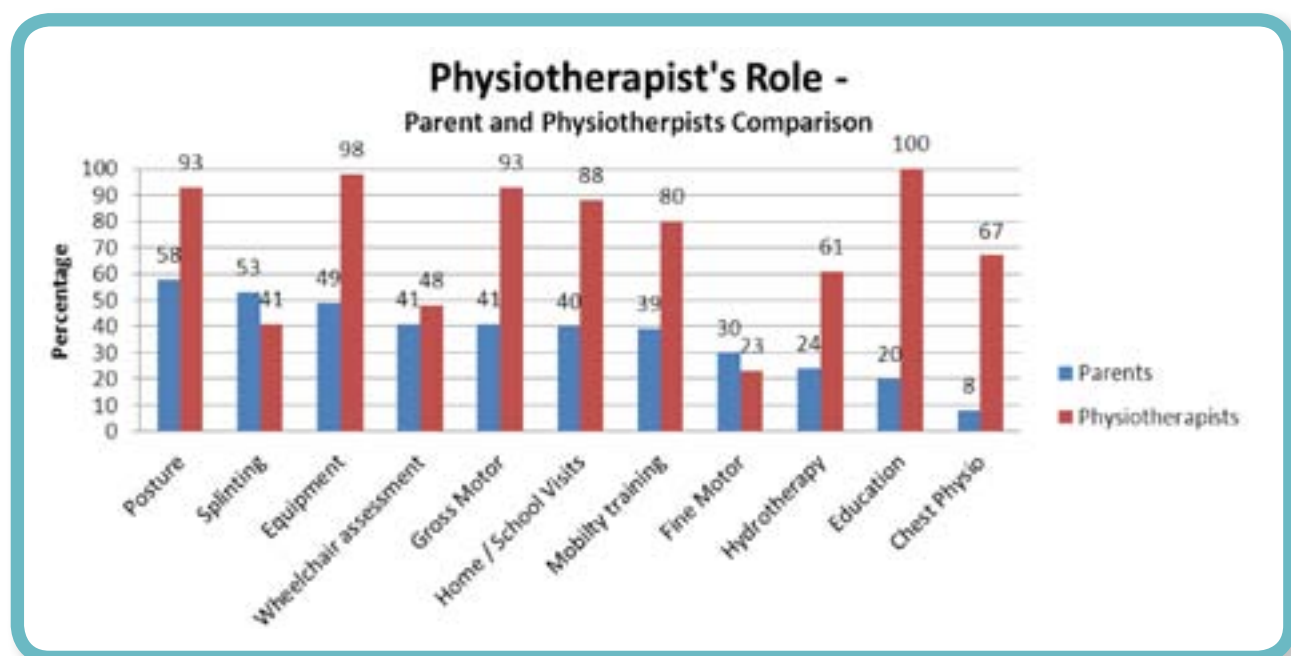
*“So next week we will be concentrating on standing and walking. So he’s doing spider cage therapy here. Which is something that they do in the States a lot, but they don’t here at all” (Service Provider)*

The main interventions performed by Physiotherapists are outlined below from the perspective of parents and physiotherapists themselves. Some disparity in relation to frequency of various roles, and a range of interventions were commonly mentioned. The most common interventions outlined by parents were posture / positioning (reported by 58% of parents), upper and lower limb splinting (reported by 53% of parents), assessment and prescription of mobility aids (reported by 49% of parents), wheelchair and seating (reported by 41% of parents) and gross motor skills (reported by 41% of parents). The top 4 interventions as described by parents relates to equipment needs to support posture and mobility.

Physiotherapists reported being most frequently involved in education (reported by 100% of physiotherapists), equipment provisions and assessment (reported by 98% of physiotherapists), gross motor (reported by 93% of physiotherapists), posture (reported by 93% of physiotherapists) and home and school visits (reported by 88% of physiotherapists).

Some areas of intervention were mentioned by physiotherapists but not by parents, specifically transfers, which may have been considered as part of gross motor skills, sexual function which was not commented on by parents, pressure relief, learning and attainment, and self-care (see figure below). This discrepancy in perception of interventions and role of physiotherapy requires further clarification of the role for parents.

Figure 78



Half of the physiotherapists reported that there was no waiting list for initial access to services, with an additional 46% indicating that there was a wait time of less than 3 months. Only 4% reported waiting more than 3 months for access to services. In interviews physiotherapists also acknowledged very short waiting times.

*“And we didn’t have a waiting list here, and if we did, it was very short. So they would have been seen more or less straight away kind of within a month” (Service Provider)*

*“No we don’t operate a waiting list... I mean what we constantly have to do is try and prioritise and reprioritise” (Service Provider)*

*“We don’t have a waiting list so if say the forum happens on the first Monday then the child will be seen within 2 weeks” (Service Provider)*

*“From a physiotherapy point of view there is no waiting list” (Service Provider)*

Home and school visits:

Physiotherapists reported that they would frequently (more than 50% of the time) complete home visits (13% of Physiotherapists) and school visits (7% Physiotherapists). Home visits were performed slightly more frequently by physiotherapists than school visits (see figures below).

Figure 79

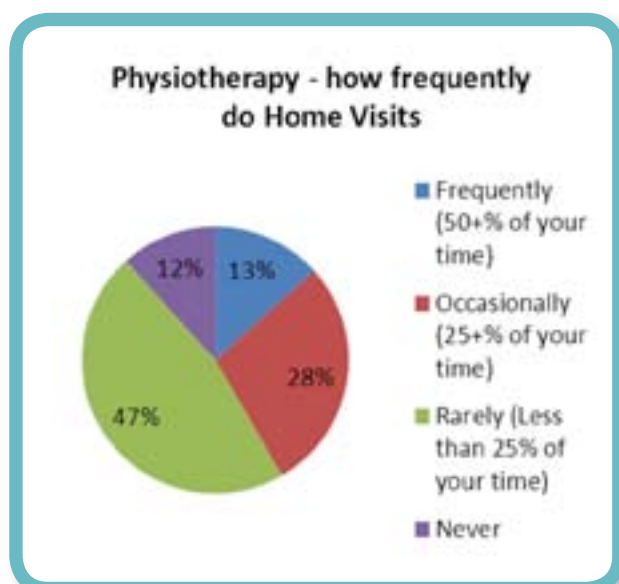
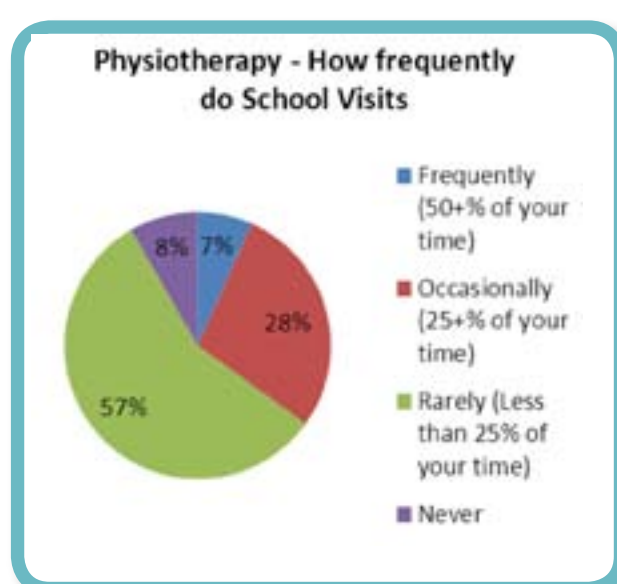


Figure 80

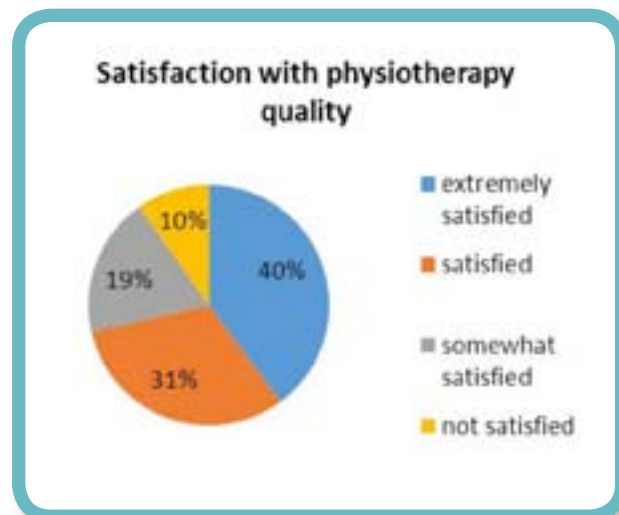


In relation to satisfaction with Physiotherapy Services, similarly to the Occupational Therapy services, parents were generally more satisfied with the quality than they were with the availability.

Figure 81



Figure 82



Comments received from parents were generally requesting more frequent physiotherapy sessions and intervention.

*"You'd be given a home programme to do which you will do at home but I mean these children deserve a better service which is to be seen weekly or fortnightly not just under review every so months to see how they're doing, It's not good enough" (Parent / Guardian)*

*"I have a friend who is a physio who will regularly see her. So I'm not in as horrible a position as what a lot of other people are" (Parent / Guardian)*

*"We're not one bit happy with the physio and OT"  
(Parent / Guardian)*

### Physiotherapist frequency of Review:

When asked about the frequency at which they review children with spina bifida, the highest proportion of Physiotherapists indicated that they see infants every 2 weeks, whereas they would recommend they be reviewed on a weekly basis. Wheelchair users and those that mobilise with splints are most frequently reviewed monthly which is consistent with their professional recommendation. Independent mobilisers are currently seen by the highest proportion of Physiotherapists every 3 months, and they would recommend that they be seen less frequently at every 6 months.

Overall, as indicated in the table below, currently Physiotherapists see younger children (pre-mobilisers) more frequently than others. Teulier et al. (2009) reported that as walking onset is delayed by an average of 2 years in spina bifida additional intervention is required within this timeframe. Physiotherapists agree with the frequency at which they review children who are wheelchair-dependent and those that mobilise with splints. They would recommend seeing children who mobilise independently less frequently than currently.

Table 13

Frequency of review by physiotherapists- current and recommended

Frequency of Review by Physiotherapy Current & Recommended		
Functional Category	Currently	Recommended
Infants and Pre-mobiliser	Every 2 weeks	Weekly
Wheelchair users	Monthly	Monthly
Mobile with splints	Monthly	Monthly
Independently mobile	Every 3 months	Every 6 months

Comments from physiotherapists often related to this frequency of review being based on individual needs and difficult to quantify by category apart from the infants and children whose mobility is not yet established. Social, family and school factors can also influence the frequency of need and review as well as surgical intervention or change of splints affecting the need for blocks of intervention.

*“Just because it’s spina bifida, doesn’t mean they should have physiotherapy for life, because it is for living. And we should be there and accessible when they need us and disappear when they don’t.” (Service Provider)*

*“Input is needs led and varies according to the needs of the child”  
(Service Provider)*

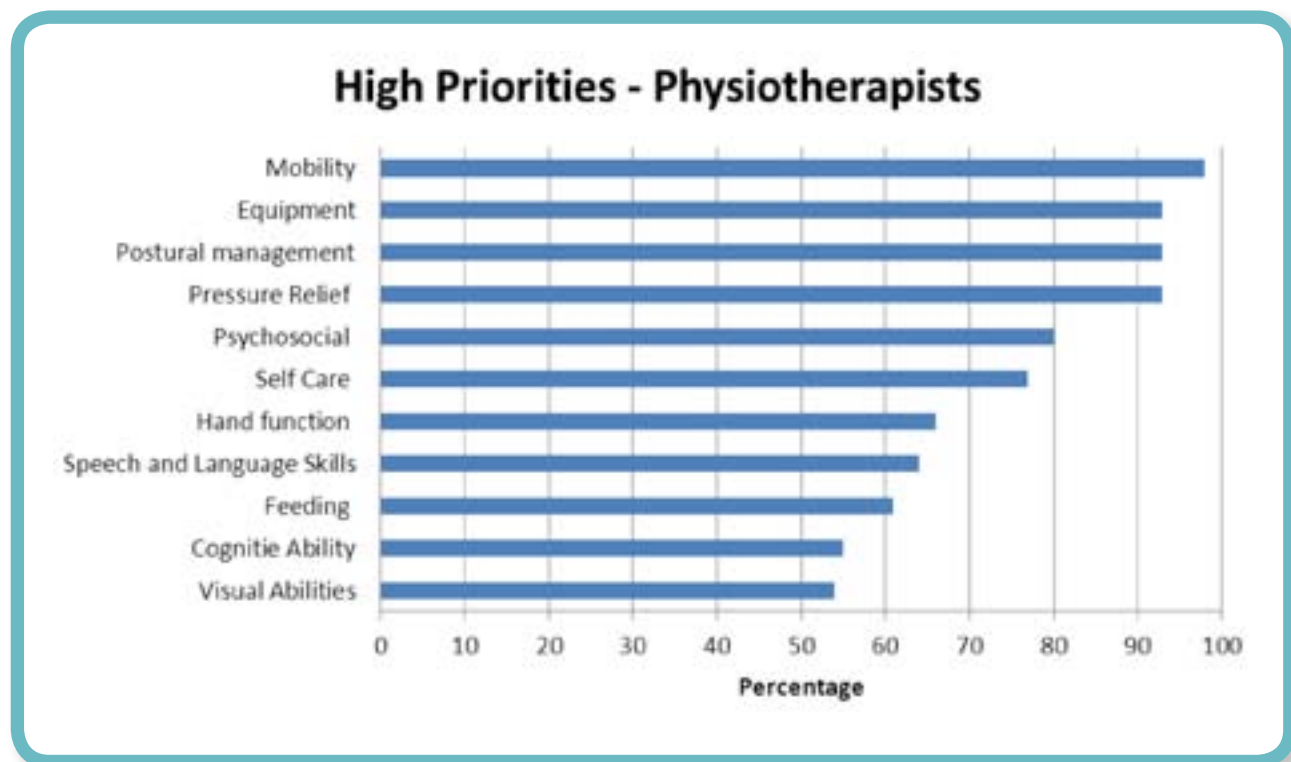
*“Generally after mobility is established and there is a gross motor plateau, children are seen as required. Frequency of treatment will depend on ability and potential also time constraints when attending local school” (Service Provider)*

*“This depends on their aims and level of need, more frequently if potential progress, less frequent if stable/static” (Service Provider)*

*“Intervention levels don’t just depend on the categories above, Surgery, problems with splints, growth spurt, and environmental / social needs all contribute”  
(Service Provider)*

The overall priorities for Physiotherapists are presented in the bar chart below. The top four priorities for Physiotherapists were mobility, followed by equipment provision, postural management and pressure relief.

Figure 83

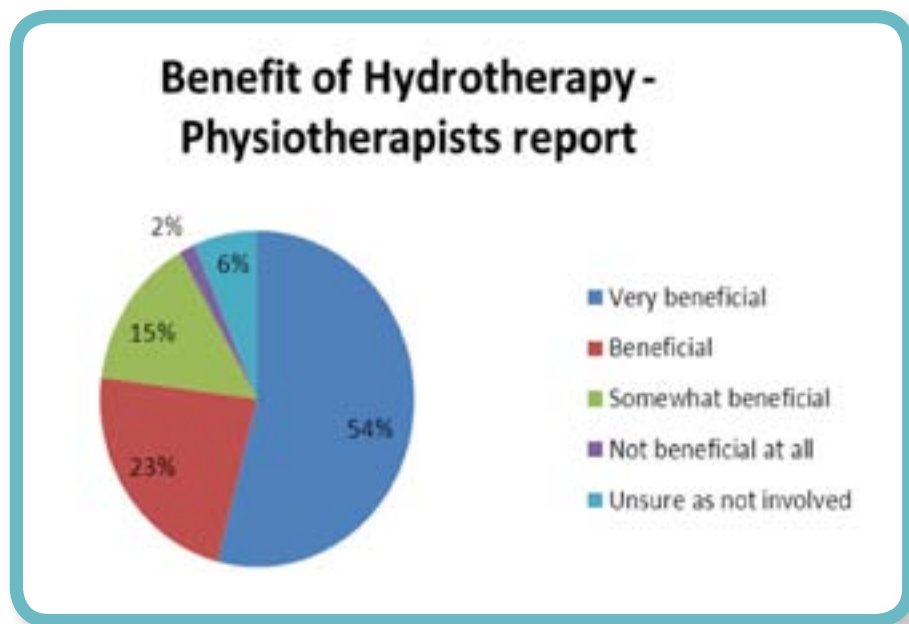


## Hydrotherapy

Hydrotherapy is a ‘therapy programme utilising the properties of water, specifically designed by a suitably qualified physiotherapist for an individual to maximise function which can be physical, physiological, or psychosocial.’ (Hydrotherapy Association of Chartered Physiotherapists 2007) Studies by Mulligan and Polkinghorne (2013), Boulter (1992) report the benefits of a hydrotherapy pool for people with physical disabilities as an invaluable facility which provides an opportunity to be physically active in a way that is ‘pain free, enjoyable and beneficial for mind and body’.

75% of Physiotherapists reported that hydrotherapy was beneficial for the children with spina bifida.

Figure 84



Comments received were in relation to the physical benefits of the exercise, pressure relief, and manual handling advantages compared to ground-based therapy:

*“In terms of exercise pool swimming therapeutic swimming would be great to work on their weight management, pressure relieving care and it’s a lot easier to work with patients in the pool especially with spina bifida clients, they are a lot more buoyant” (Service Provider)*

In relation to access, 66% of physiotherapists indicated that they had access to a hydrotherapy pool within their area; 61% of physiotherapists reported providing hydrotherapy as an intervention; however only 32% of parents indicated that their physiotherapist had provided hydrotherapy to their child. Given the evidence and perceived benefits by therapists, it would appear that hydrotherapy is a valuable resource that is not being accessed equitably by this patient group.

## Discussion

There is a high level of need and demand for physiotherapy for children with spina bifida. Similarly to Occupational Therapists, Physiotherapists in Ireland have small numbers of children with spina bifida on their caseloads (4.3 per Physiotherapist) and work in a variety of service types indicating that these children are spread throughout services and teams nationally. The changing services to geographical-based teams as opposed to dedicated physical disability teams will further spread the physiotherapy care of these children among therapists. As discussed in the Occupational Therapy section, the need for access to specialist services for support and education is recommended.



While parents indicated that they would like more frequent therapy physiotherapists suggest that infants and those children whose mobility is yet to be established would benefit from more frequent review. This is in line with support for early intervention both in the international literature as well as the national Health strategy (Shaer, 1997, Merkens, 2006). Physiotherapists also discussed the need for flexibility in frequency of review with certain stages of development, post operatively, and upon receipt of new splints being periods of more intensive input. Some parents reported supplementing with private physiotherapy due to perceived benefits from more intensive input at certain stages to achieve developmental gains or to access non-conventional therapies. Due to the potential conflict of developmental goals it is recommended that there is open communication and collaboration between primary and private therapists, setting clear expectations and goals

Despite reported benefits, hydrotherapy is underutilised. The increased use of hydrotherapy is recommended as a therapeutic intervention for children with spina bifida to assist with joint flexibility, aerobic exercise and pressure relief.

Although similarities exist in the interventions and priorities of physiotherapy and occupational therapy, they have unique roles and complementary services for children with spina bifida. All Physiotherapists and Occupational Therapists see education of the child and family as a key role. The most frequent interventions reported by Physiotherapists were equipment, gross motor skills and posture whereas the most frequent interventions reported by Occupational Therapists were self-care, fine motor skills, and wheelchair and seating assessments. The top priority for Physiotherapists was mobility and the top priority for Occupational Therapists was equipment.

## **Recommendations**

1. Increased frequency of provision of occupational therapy and physiotherapy services is recommended based on therapists' assessments and recommendations and also the child's needs rather than prescriptive blocks of therapy or pre-determined time frames throughout the year.
2. Any private therapy if sought or obtained, should work in collaboration and communication with the primary teams with parental consent in order to clarify expectations and goals.
3. The increased use of hydrotherapy is recommended as a therapeutic intervention for children with spina bifida to assist with joint flexibility, aerobic exercise and pressure relief.



## ***Equipment***

### **Background**

Equipment use is high among children with spina bifida. As reported in Chapter 4.1 in the equipment section of physical impact of spina bifida, 83% of all children had received equipment to support their needs. Results in chapter 4.1 indicate that the most frequently used pieces of equipment were orthoses, standing frames, manual wheelchairs and mobility aids (e.g. walkers, crutches).

Limited literature exists in relation to the timing and provision of equipment. Brown (2001) indicated that a stander is recommended from the age of 12 – 15 months if the child is able to pull to stand. Orthoses and adaptive equipment are vital for children with low-level lesions to achieve independent mobility (Johnson et al., 2007, Malas, 2011). The importance of appropriate wheelchair and seating cannot be underestimated, and must meet physical and social needs, suit the environment, be safe and durable, and correctly fit and control postural needs (WHO 2008). Merkens (2006) indicates that a wheelchair should be obtained for mobility and transport at the pre-school age if appropriate. In Ireland no pathway exists for seating assessment and provision (Kenny and Gowran, 2014), this would be very beneficial to ensure consistency and quality of care for children with spina bifida.

### **Results**

Who is involved in equipment assessments?

All service provider respondents were asked about their involvement in assessments of specific equipment. Only Physiotherapists and Occupational Therapists reported to be directly involved in the assessment and prescription of equipment. The table below indicates the proportion of Physiotherapists and Occupational Therapists involved in specific equipment. Eighty seven of Occupational Therapists were involved in wheelchair assessments, none in stander assessments and 46% in sleep system assessments. 98% of physiotherapists were involved in stander assessments, 48% in wheelchair assessments and 77% in sleep system assessments.

Table 14

## Physiotherapist &amp; Occupational Therapist involvement with equipment.

	Wheelchair Assessments	Stander Assessment	Sleep System Assessment	Splints / Orthotics Assessment
Occupational Therapists	87%	0%	46%	25%
Physiotherapists	48%	98%	77%	92%

The prescription of orthotics is a vital aspect of the care of a child with spina bifida to achieve upright standing and ambulation (Carroll, 1987). This is often supported in conjunction with Orthotist, Physiotherapist, Occupational Therapist and Orthopaedic Surgeon. Ninety two per cent of physiotherapists reported being involved in this process and 25% of Occupational Therapists. Unfortunately no Orthotists responded to the quantitative questionnaire, despite the fact this was sent to all services and gatekeepers were asked to forward it to all members of the MDT. Orthotists are often private practitioners and not directly employed by the service and/or team. This was disappointing as all children requiring splints and orthoses require access to a Orthotist and their feedback was not gathered via questionnaire data.

Fifty one per cent of all service providers indicated that they are not involved in splint and/or orthoses provision. The reasons for this included that this service was provided by someone else on the team (51.4%), or a specialist service (28%), or was completed by an externally contracted Orthotist (22%). Only 7% indicated that they had a lack of splinting facilities, and 11% indicated they had a lack of training or skills related to splinting and orthotics. Only one Orthotist volunteered to participate in an interview with the researchers. In interview, this Orthotist commented on the vital relationship between the Physiotherapist and Orthotist in assessing and prescribing orthotics and described this as a joint role:

*“We are still constantly talking to each other about the patients, how we do this and that. And I think that’s the way to go forward. Everybody has to put in their knowledge together and we get great results” (Service Provider)*

## Where is specialist equipment being accessed?

Specialist seating was required by 68% (n=106) of children according to their parents. This service was predominantly accessed through Enable Ireland, Health Service Executive or the Central Remedial Clinic. A greater spread of services were accessed for specialist splints and orthotics with 69% (n=107) of parents accessing these (see Figures 73 and 74 below). Four and five per cent of parents respectively reported accessing specialist seating services and splints and orthotic services privately.

Figure 85

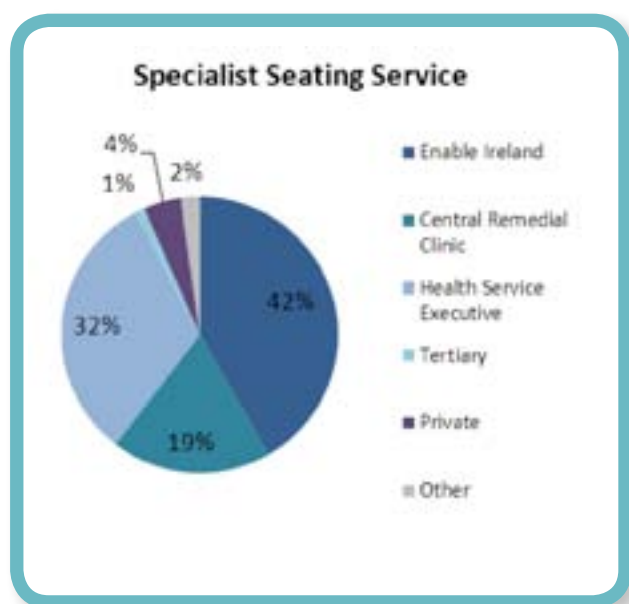
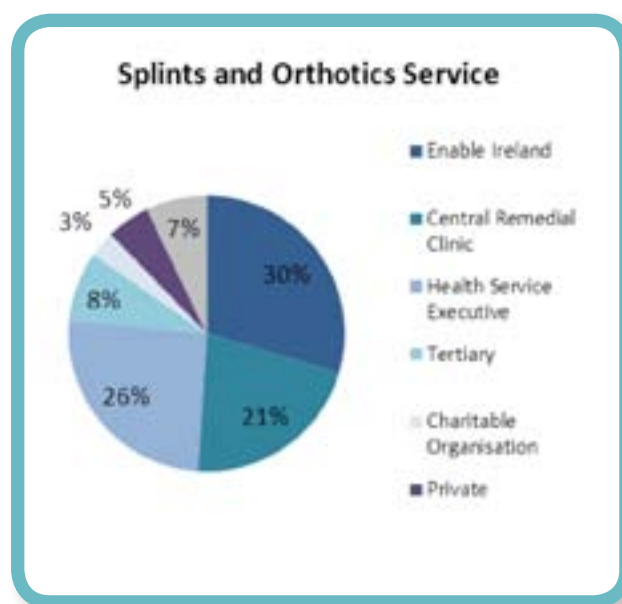


Figure 86

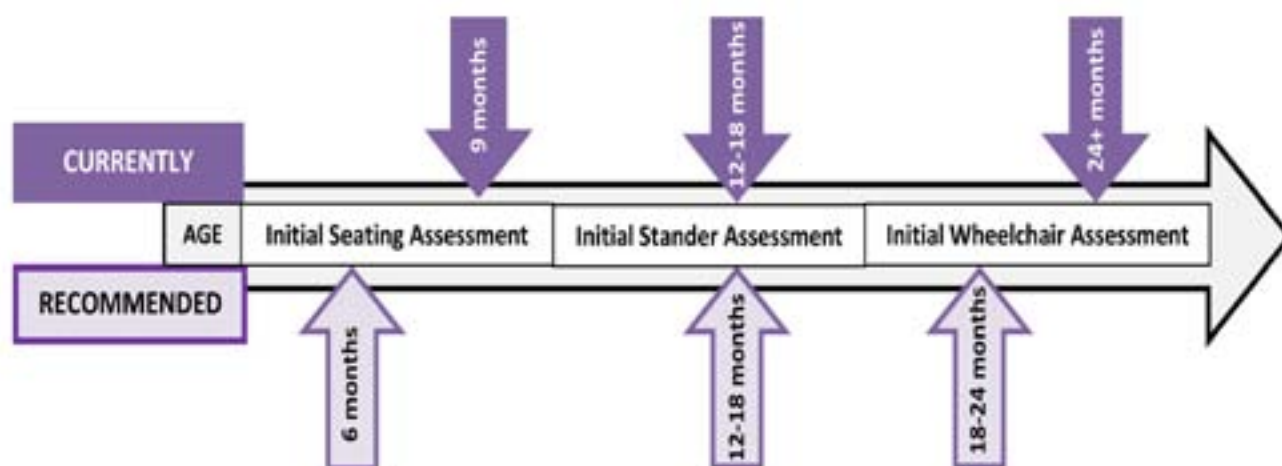


## When are equipment assessments occurring?

Those service providers that are involved in the equipment assessment process (Physiotherapists and Occupational Therapists) were asked when initial seating, wheelchair and stander assessments are currently occurring within their service and when they would recommend these assessments should be occurring for children with spina bifida.

Figure 87

## When equipment assessments occur and recommended



Initial seating assessments were reported to occur most frequently in the first 9 months of life. Service providers recommended that this assessment should occur earlier than is currently occurring, within the first 6 months of life.

The majority (60%) of service providers report that the first wheelchair assessment is not completed until after 24 months. Their recommendations suggest that this should occur earlier, with most suggesting between 18 and 24 months as the optimum time for an initial wheelchair assessment. Although no research was found regarding the recommended timing of seating and wheelchair assessments for children with spina bifida, Wright et al. (2010) reviewed the standards of practice of seating and wheelchair assessments for children in the UK and Ireland. He concluded that the research supports the need for specialist seating and wheelchairs in children with physical disability however the need for guidelines in assessment and provision was identified.

The highest number of service providers in the current study reported completing the first stander assessment when the child is aged between 12 and 18 months however more recommended that the assessments should occur between 6-12 months or 12-18 months than is currently happening. This agrees with Brown (2001) who recommended standers to be used between 12—15 months when children begin to pull to stand.

Overall, service providers recommendation is that seating assessments should occur earlier than currently at 6 months of age, stander assessments should occur at 12-18 months which is

in line with current practice and wheelchair assessments should occur earlier than is currently happening at 18-24 months. In interview, service providers commented that equipment assessment is difficult to define by age or timeframe and should be based on the individual needs of the child.

*“Orthotics are usually considered with the active client once they pull to stand so we have guidelines of when to introduce splinting and standing”  
(Service Provider)*

*“Every child I can think of with spina bifida would have had a different (seating) need at a different time”  
(Service Provider)*

### **Wait time for receipt of equipment:**

Both service providers and parents were asked the wait-time for receipt of equipment once assessed. Quantitatively, the highest proportion of both groups indicated that equipment such as standers sleep systems, splints and orthoses arrived within 3 months of ordering. There were greater delays reported in relation to delivery of specialist seating and wheelchairs with a delay of 6 months by the highest proportion of parents and service providers. Some discrepancies were observed between service providers and parents perception of how long equipment takes to arrive, reasons for this are unclear but parents were reporting specifically related to their child’s experience while service providers could were providing more general estimations of wait times for all children accessing their services.

Table 15

	How long equipment takes to arrive*	
	Service Provider	Parents
Seating	3-6 months (41%)	1-3 months (54%)
Wheelchair	3-6 months (38%)	6-12 months (38%)
Stander	0-3 months (43%)	1-3 months (28%)
Sleep System	0-3 months (32%) 3-6 months (32%)	1-3 months (37%)
Splints	0-3 months (50%)	1-3 months (45%)
Orthoses	0-3 months (42%)	1-3 months (41%)

\* Reported as the highest proportion of service providers / parents

In interview, however, there were further discussions in relation to the delay in receipt of equipment which included assessment and funding. The lack of funding for equipment, inefficiencies and inconsistencies between areas and organisations caused a huge stress and frustration for parents and service providers and some feel this is worsening and requires greater transparency.

*“What we are dealing with in our catchment is 2 areas of funding. Within our group we might have a group that is funded quicker than the other depending on where you live” (Service Provider)*

*“It depends on geographical area how long you’re going to wait” (Service Provider)*

*“We haven’t had an issue here with delays for equipment. And we have one child with very complex needs” (Service Provider)*

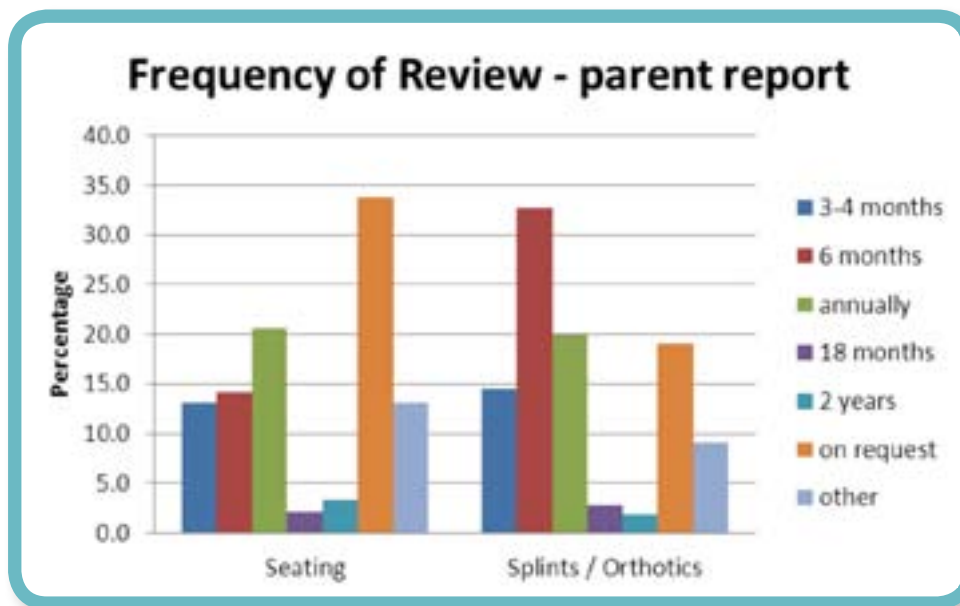
*“And the waiting times for things like his HKAFOS, he just got a new set there, two weeks ago. And we ended up paying for them ourselves because the wait time through the HSE is just so long” (Parent / Guardian)*

*“And lately I found a lot of parents are going out buying things themselves, because it’s easier. I mean the length of time they are waiting is ridiculous, you know they are nearly moving onto the next thing, by the time they get what was on list already” (Service Provider)*

### **Equipment Review and Repair:**

The frequencies at which specialist seating and orthoses were reviewed are listed below. The largest portion of parents indicated that seating is only reviewed on request (34%), with an additional 21% being reviewed annually. The highest proportion of parents reported that splints and orthoses were reviewed most frequently every 6 months (33%) followed by annually (20%).

Figure 88



In interview, both service providers and parents expressed concern and frustration in relation to the delay in receiving and repairing appropriate equipment thus impacting on functional or therapeutic goal attainment.

*“She’s without her splints more often than she’s with them because they’ve gone off for a repair or a change or she can’t wear them for whatever reason”*  
(Parent / Guardian)

*“I think the more complex the equipment often the longer the wait”*  
(Service Provider)

*“So a lot of his therapy has really being delayed by lack of equipment”*  
(Parent / Guardian)

*“He hasn’t had a chance yet to try standing or walking, because the HKAFOs, the pervious set just weren’t any good. So that is very frustrating as well. And I know it is for therapists too”* (Parent / Guardian)

### Potential solutions to Equipment Delays:

Service providers frequently commented on the potential benefits of equipment recycling as a method to deal with delays in funding. This would allow for general equipment to be accessed, but may not meet the specialist needs of children with spina bifida.

*“It would be really great to have the second best possibility until you get what you want in. I think that’s really needed”  
(Service Provider)*

*“There are delays in certain areas for sanctioning of equipment but we are kind of fortunate that we have a fair amount of stock of you know it mightn’t be the exact type that you ordered for the child but it will do until then”  
(Service Provider)*

*“We have a good stock of equipment here and we have a loan system here where we have a good stock of items that we regularly use, we have an agreement with the HSE that we can service and loan out other HSE equipment once its been fully serviced so we don’t usually have a delay” (Service Provider)*

*“You see fortunately, we have a store and we kind of recycle and upcycle all of these things...they might not have got the right one, or at the right time”  
(Service Provider)*

*“If you needed something very specific for an individual child that you wouldn’t have in a storeroom” (Service Provider)*

Flexibility in funding is required to meet complex needs (e.g. needing multiple pieces of equipment together such as AFOs and a stander) was an additional area of stress, whereby parents and service providers struggle to receive sanctioning for more than one piece of equipment at a time.

*“You are waiting a year for every piece of equipment which is entirely ridiculous and you can’t really send in two prescriptions at the same time, they don’t seem to understand that a child needs AFOs and HKAFOs and a wheelchair and a stander and this and that, and the other, you are allowed to have one prescription in at a time and that will take a year” (Parent / Guardian)*

*Further upset and frustration was expressed by parents having to re-apply for a medical card and being ineligible for a long term illness card. (Parent / Guardian)*



*“He was 14 when he qualified for the medical card and up till that we were paying for everything” (Parent / Guardian)*

*“More interested in saving pennies and they are losing the grasp with themselves, because in the end, they are actually losing pounds” (Parent / Guardian)*

## **Discussion**

There is a high demand for a range of specialist equipment for children with spina bifida to support independence and mobility. Physiotherapists and Occupational Therapists are the professionals most frequently involved in the assessment and prescription process for these pieces of equipment in the local teams along with support from specialist clinics and orthotists. Their recommendation is that seating and wheelchair assessments should be occurring at a younger age than is currently occurring, but should also be based on the individual needs of the child. A seating assessment should be considered when a child is approximately 6 months, and if applicable an initial wheelchair assessment between 18 months and 24 months of age. Waiting until school age for a child who requires a wheelchair may impair their ability to engage in day to day activities and participate in cognitive and psychosocial development (Wright, Casey, & Porter-Armstrong, 2010). It is important to note however that every child is individual and assessment and provision of equipment should be based on careful assessment and judgement by a trained professional (Batavia, Gianutsos, Vaccaro, & Gold, 2001).

The delay in receipt of equipment is a cause for concern and can impact the ability for a child to make functional gains, or receiving equipment that has already been outgrown, thus resulting in further wastage. Wheelchairs experienced the greatest delays with the highest proportion taking 3-6 months to arrive post assessment. This does not compare favourably with a review in the UK which indicates that wheelchairs are in place 9 weeks post assessment (Care Quality Commission, 2012)

Inequities in equipment provision were raised during interview with parents and services providers commenting on the delays and disparities in acquiring funding for equipment. Increased funding and more equitable access is required. The establishment of local equipment recycling facilities with appropriately trained staff where equipment can be cleaned, refurbished and redistributed, particularly in emergency situations may ease the burden for families and service providers for more generic equipment or while experiencing delays awaiting specialist equipment.

Equipment provision could be improved by appropriate training of local staff with regard specialist or generic pieces of equipment that might be of benefit for children with spina bifida. Increasing the funding for aids and appliances throughout the HSE regions could also occur or consider reallocation of funding from savings made by reduced wastage and increased recycling.

## Recommendations

1. It is recommended that initial seating and wheelchair assessments should occur earlier than therapists' report this is currently happening.
2. It is recommended that there should be a reduction in inefficiencies of funding equipment throughout the country to ensure essential equipment is provided in a timely manner.
3. Increasing the funding for aids and appliances throughout the HSE regions could also occur or consider reallocation of funding from savings made by reduced wastage and increased recycling.

## 5.2 SPECIALIST SERVICE

### Introduction

The care of people with spina bifida requires multiple specialists with various expertise (Brustrom et al., 2012). Children with spina bifida have complex needs which require specialist tertiary services that are integrated with local service systems. Liptak and El Samra (2010) described this nicely, highlighting the need for:

*“Specialists who can address issues like hydrocephalus, neurogenic bowel and bladder, mobility, and learning disabilities; they need generalists who can address health promotion, including nutrition and exercise; and they need an integrated system to deliver this complex care and to align and inform all the providers”.*

Children and adults with spina bifida require frequent access to specialist tertiary services with many reports highlighting increased hospital admission rates (Young et al. 2005). A comprehensive, integrated and appropriately staffed specialist service is required to meet the needs of children with spina bifida. This is required in the antenatal and neonatal period and also throughout childhood to prevent complications specific to spina bifida ranging from shunt complications, renal failure, scoliosis or postural deformities to obesity. Some of the subspecialties involved in monitoring these areas are neurosurgery, urology and orthopaedics and these professionals are required as part of the Multidisciplinary Team (MDT) to prevent secondary disabilities. Secondary disabilities are preventable conditions, directly or indirectly related to the disability which are reflective of the person within their environment and have been seen to be particularly challenging for people with spina bifida (Simeonsson et al., 2002). As a result, specific and specialised MDTs are required to coordinate and manage

the care of these children with efficient communication and appropriate transition planning at major transition stages. It is important to note that access to other professionals including imaging and Radiology are also essential to support scans and investigations required by Neurosurgery, Urology and Orthopaedics. Aspects of specialist care related to the antenatal and neonatal period, access to specialist professionals (neurosurgery, urology, orthopaedics, spina bifida nurse), the specialist multidisciplinary Spina Bifida clinic, communication and transition will now be discussed.

## ***Antenatal and Neonatal Period***

### **Background**

As highlighted in Chapter 4.3, family impact, parents and families have a difficult time coping with the diagnosis of spina bifida and the neonatal stage of adjusting to having a child with spina bifida. As a result it is essential that they are appropriately supported at these initial stages.

Neural Tube Defects (NTDs) including spina bifida can often be diagnosed antenatally which Hüsler et al. (2009) found to be crucial for appropriate counselling.

Ireland has no accepted national policy regarding prenatal screening and diagnosis, with no specific NTD screening policy and inconsistency in the availability of tests including maternal serum alphafetoprotein has been reported (Lynch and Malone, 2007, McDonnell et al., 2014). A recent review noted an antenatal diagnosis rate of NTDs for Europe at 88% (range 25-94%) with variation influenced by the presence or absence of ultrasound screening policies and attitudes to termination (Boyd et al., 2008). In Ireland, McDonnell et al. (2014) completed a study reviewing the incidence rates and epidemiological data of NTDs between 2009-11, this study identified that 74% of children with spina bifida were diagnosed antenatally with 62% of these being diagnosed before 24 weeks of gestation. Gestational age at diagnosis varies and appears to be improving with the advancement of ultrasound technology, some studies highlight the mean gestational age at diagnosis to be 21.2 weeks (16–35 weeks) (Hüsler et al., 2009) and 22 weeks median gestation (16-39) for antenatal diagnosis in an Irish context (Boyd et al., 2008).

As previously cited, the Spina Bifida Association of America (SBA) established “Guidelines for Spina Bifida Health Care Services throughout life” (Merkens 2006) following a conference on evidence-based practice (Liptak 2004). In these guidelines the antenatal priorities include ensuring families receive accurate information about spina bifida regarding a range of outcomes (Merkens, 2006). At present in Ireland, following receipt of a diagnosis of spina bifida antenatally, expectant mothers can access one of the Consultant Neurosurgeons, the Spina Bifida Nurse Specialist from TSCUH, a Foetal Medicine fellow and a consultant obstetrician at the national foetal neurosurgical clinic. This service has been in existence for

3 years to provide antenatal counselling service for those who have received an antenatal diagnosis of NTD. Not all parents who receive an antenatal diagnosis are referred to this service even though its existence has been highlighted to all obstetric units.

There is no clear evidence from a neurosurgical perspective to favour Caesarean Section in the absence of gross hydrocephalus, breech or other obstetric indications, and incidence rates of neonatal meningitis were not affected by delivery method (Thompson, 2009). Caesarean deliveries may be chosen to assist with the planning of delivery and proximity to tertiary care. Merkens (2006) recommends that following the delivery of a baby with spina bifida they should be referred to the tertiary Neonatal Intensive Care Unit (NICU) where multidisciplinary care is required for optimal care. At present in Ireland, tertiary neonatal care is provided for children with spina bifida in TSCUH.

## Results

Many of parents during interview revealed that their child had not received an antenatal diagnosis of spina bifida or that this diagnosis was very late in the pregnancy (after 32 weeks gestation). Where an antenatal diagnosis was obtained, parents who had a meeting with members of a specialist MDT (namely the Consultant Neurosurgeon and spina bifida nurse specialist) found this to be beneficial with positive comments including:

*“He was brilliant... he said, now I know these children, I see these children grow up ... I’m going to be much more upbeat about everything. Because I see these kids grow up, it was great so see him” (Parent / Guardian)*

*Parents appeared to place value on attending a tertiary neonatal centre, highlighting excellent care in comparison to local maternity hospital where some parents perceived there to be a lack of experience with these children:  
(Parent / Guardian)*

*“She was born in a local hospital, they’re not prepared for kids like this”  
(Parent / Guardian)*

Parents also reported some negative perceptions at the neonatal stage regarding the physical separation of mother and baby and the pressure and expectations placed on fathers while mothers remained in maternity hospitals, usually following a planned Caesarean Section:

*“I was in one place, and she was in another. I had to wait a few days before I was transferred over, so that was stressful in itself” (Parent / Guardian)*

*“I was 5 days without seeing her” (Parent / Guardian)*

*“My husband was a little bit bewildered, he stayed with her for the first couple of nights, because I had had a c-section... I think he found that a little bit overwhelming” (Parent / Guardian)*

The busy physical environment of the neonatal ward was sometimes reported to be stressful for parents, particularly if they weren't sure what to expect:

*“I was totally lost in the world of hospital” (Parent / Guardian)*

*“It was too scattered, far too scattered. it's tough being in a hospital and its tough having a new baby to find yourself landed in this pure turmoil, cos that's what it is,] you're coming to terms with your baby who is not what you expected and that's not addressed and then on top of that, you're being bombarded with information, medical information that's a little bit too much too soon. They all meant well, everybody meant well, but definitely it could be co-ordinated, absolutely yeah and I hope it has improved since then” (Parent / Guardian)*

Finally parents varied regarding the amount of information that they received from service providers during the neonatal period. Some parents reported receiving too much information from numerous professionals yet some felt more information would have been beneficial:

*“Mayhem, Piccadilly circus, it was an endless stream of people, medics, social workers, everybody and it was too much information too soon, and I couldn't take it in and although they kept telling me to rest they kept coming in to me with more information. It was far too much too soon” (Parent / Guardian)*

*“I can't remember any information or support” (Parent / Guardian)*

## **Discussion**

Although high rates of antenatal diagnosis of spina bifida early in the pregnancy are noted in recent literature, (McDonnell et al., 2014), many of the parents interviewed in the current sample received a late diagnosis or none at all. Reasons for this are unclear however interview respondents represented a national spread throughout Ireland with Pasquier et

al. (2007) suggesting the rate of antenatal diagnosis decreases with increasing distance between where parents/families live and their referring centre. Similarly the interview cohort represented children aged 0-18, whereas McDonnell et al. (2014) data may represent a more accurate representation of the current diagnosis rate. An antenatal diagnosis of NTD should occur in at least 88% of cases as this is the average diagnosis rate for Europe (Boyd et al., 2008). As discussed earlier Ireland currently has no national policy for prenatal screening and NTD diagnosis. Ways that the antenatal diagnosis rate might be improved may include more equitable access to ultrasound, for example in the form of a national ultrasound screening programme as is the case with many European countries (Boyd et al., 2008).

While many parents valued the presence of antenatal counselling when present this service was not available or accessed by a large cohort of parents who were interviewed. Dunleavy (2007) emphasised the role of the nurse coordinator as part of a multidisciplinary team necessary to counsel and educate parents and families after the receipt of an antenatal diagnosis of spina bifida. Referral to the national foetal neurosurgical clinic for all parents who receive an antenatal diagnosis of spina bifida is recommended.

Currently children born with spina bifida in Ireland since 2008 attend TSCUH as their tertiary neonatal centre where they receive specialist and multidisciplinary care, this appears valued by parents and professionals. The busy nature of this environment and information provided to parents should be coordinated so as not to overwhelm them as some parents reported stress with too much information being provided during the neonatal period. Multidisciplinary care of the neonate with spina bifida is required for optimal care (Merkens, 2006) therefore appropriate staffing of the neonatal ward is required. This should include the provision of a paediatrician, who is not presently available due to resource issues. Parents raised concerns regarding the physical separation from their baby following delivery due to the need to transfer to the paediatric neurosurgical centre.

## Recommendations

1. Services should strive to provide an early antenatal diagnosis for all NTDs in Ireland.
2. Antenatal counselling by members of the multidisciplinary spina bifida team which includes discussion of management options and potential outcomes should continue to be available to any parent who desires this.
3. The full specialist multidisciplinary team should be available at the initial neonatal stage to support children and families during this complex time. At present there are no resources to provide a paediatrician review for neonatal inpatients with spina bifida after their surgical repair. To promote continuity of care and specialist input, involvement of the specialist paediatrician at this stage is recommended.
4. Consideration should be given to parental anxiety and distress following the separation at birth for transfer to the neonatal service.

## ***Access to Specialist Service Professionals***

Within specialist services, various professionals are necessary to meet the needs of children with spina bifida. Some key health professionals are only available within the tertiary hospital setting for example the Neurosurgeon, Urologist, Orthopaedic Surgeon and the Spina Bifida Clinical Nurse Specialist. Access to these professionals, a review of complications, interventions and needs are discussed below in relation to results from the perspective of parents and service providers.

### **Access to Neurosurgery**

#### **Background:**

Merkens (2006) guidelines for spina bifida healthcare management throughout the lifespan recommend a thorough neurological examination of a neonate with spina bifida as well as access to a neurosurgeon throughout the lifespan to monitor hydrocephalus, Chiari II malformations, syringomyelia, and tethering of the spinal cord. For children with spina bifida, Northrup and Volcik (2000) discussed the need for various invasive surgeries soon after birth, particularly in the case of an open defect such as a Myelomeningocele where surgical closure of the lesion is required within 72 hours and the insertion of a cerebrospinal fluid shunt may also be necessary to manage hydrocephalus.

The recent introduction of foetal repair of MMC was reported by Houtrow & Dicannio (2014) to be one of the most important advancements in Myelomeningocele management in recent years. The Management of Myelomeningocele Study (MOMS) randomised control trial Adzick et al. (2011) completed foetal repair before 26 weeks' gestation. Results suggested a reduced need for shunting at 12 months, a less severe degree of hindbrain herniation and better than expected outcomes of motor functioning adjusted for lesion level. However pregnancy complications were more common with 13% having preterm delivery before 30 weeks and while potential benefits of antenatal surgery need to be balanced against maternal risk a follow-up study MOMS2 will be concluded in 2016 and will provide neuropsychological, functional, urological and radiographic outcomes at school age (Adzick et al., 2011). At present only selected medical centres throughout the world complete antenatal or foetal Myelomeningocele closure as it is still very new and evolving. In Ireland at present only post natal surgical repair/closure is completed. A recent review of literature regarding neurosurgical treatment strategies Bowman and McLone (2010) highlighted that the surgical repair or closure of the lesion should optimally occur within 72 hours following the child's birth.

Other necessary neurosurgical interventions include normalising the size, pressure or fluid dynamics of cerebrospinal fluid (CSF) in the lateral ventricles and preservation of cognitive function when treating hydrocephalus, maintaining brainstem cranial nerve function with a Chiari II malformation, preserving lower spinal cord neurological function and upper extremity function present at birth when a syringomyelia or tethered spinal cord is present (Merken, 2006). Bowman and McLone (2010) reported the neurosurgical goal when treating children with spina bifida to be the maintenance of stable neurological functioning throughout life. In order to do this frequent access and review by a Neurosurgeon is necessary. In relation to access to neurosurgeons, the NHS recommends that "children should expect to be treated by a paediatric-trained neurosurgeon, with access to care, advice and support 24 hours a day, 7 days a week" and children with lifelong conditions such as spina bifida require ongoing care and support as well (Steers and Stower, 2010).

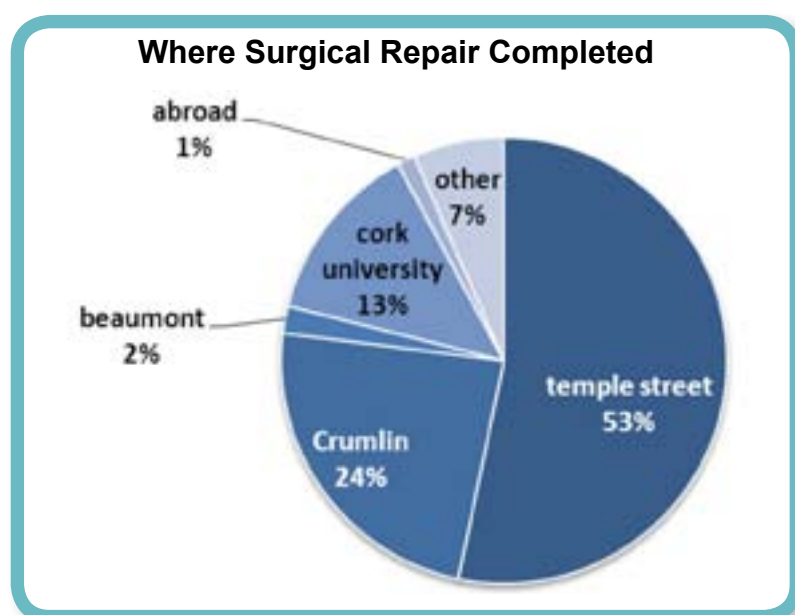
## **Results:**

As highlighted in chapter 4.1 impact on child in the physical impact section, 64% of parents in the current sample report their child has a Cerebrospinal Fluid Shunt (namely a VP shunt) in situ to manage hydrocephalus, with almost half of these which were inserted in the first week of life. Multiple surgeries are common with 20% of children who required more than 4 shunt related surgeries.

There was a lot of variation in where children had their surgical closure completed with the highest proportion (53%) reporting this was completed in TSCUH (see Figure 88 for distribution of location of closure) Those who selected 'other' were generally for children who had closed spina bifida not requiring surgical repair.



Figure 89



The sample presented in this research demonstrated children ranging from 2 month to 18 years of age, therefore frequencies were calculated to highlight differences in location of closure with age. As previously discussed, the reconfiguration of neurosurgical services occurred in 2008, and research data collection occurred in June 2013, therefore comparisons were made between children aged under 4.75 years and children over 4.75 years to demonstrate the effect

of this reconfiguration. Results demonstrate that 83.7% of the total sample aged under 4.75 years had their lesion repaired in TSCUH whereas the majority of children aged over 4.75 years (53.8%) had their lesion repaired in Our Lady's Hospital Crumlin (see Table 16) therefore demonstrating the change in service delivery in 2008 from Crumlin / Beaumont to Temple Street.

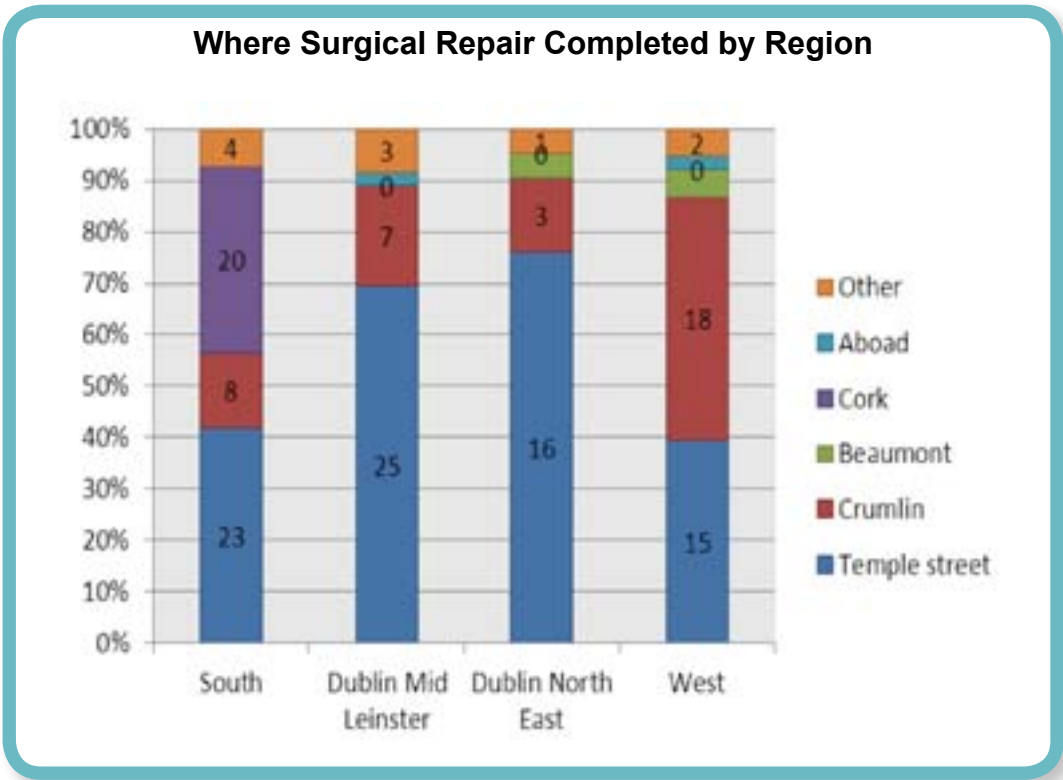
Table 16

Neurosurgery frequency of review

NEUROSURGERY	3 monthly or more	Twice a year	Once a year	Not this year	Never
Temple Street Children's University Hospital	22.9	21.7	42.0	11.6	2.9
Our Lady's Hospital Crumlin	0.0	3.0	48.5	33.3	15.2
Cork University Hospital	0.0	5.6	27.8	50.0	16.7
Beaumont	0.0	0.0	66.7	33.3	0.0

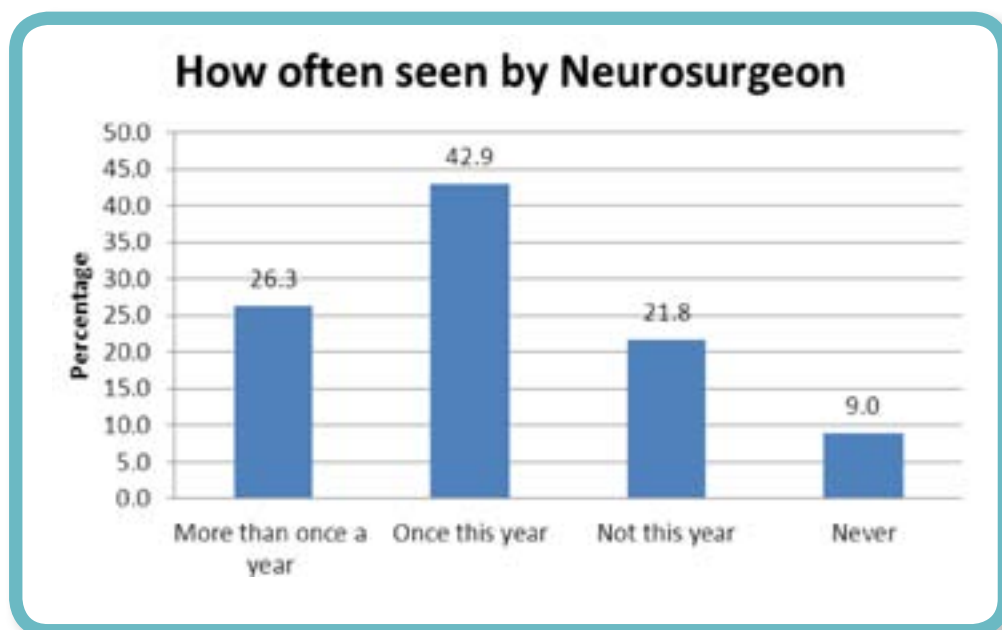
Interestingly even since the formation of TSCUH as the national neurosurgical centre for children aged under 6 years, 8.1% of children aged under 4.75 years had their lesion repaired in Cork University Hospital. These children were all over 3 years old demonstrating that in the past 3 years all children in the current sample have had their neonatal closures completed in TSCUH.

Figure 90



While the above table demonstrates that neurosurgical services for children under 6 years has moved to TSCUH, the sample of children in this research represents children up to 18 years and therefore access to neurosurgery is likely to still be varied dependent on where you live. For this reason comparisons were made based on where a child’s neonatal closure was completed and the HSE region where they live, results suggest that their address was a significant factor as to where their neonatal closure occurred ( $p=0.003$ ). Figure 90 demonstrates that only children living in HSE South had accessed neurosurgical services in this area but TSCUH was the most frequently accessed by respondents from all areas except for HSE West where respondents were more likely to have their neonatal closure completed in Our Ladies Children’s Hospital in Crumlin. Reasons for this are unclear and could have been influenced by many factors including personal preference, ease of access and transport.

Figure 91



In relation to having one neurosurgical centre service providers commented on the increase ease of access and communication with having one point of contact with comments including:

*"I think from a shunt point of view, pulling the neurosurgery into Temple Street has made a massive difference. The attitude of the appointed Neurosurgeons has changed a hundred and eighty degrees how, in terms of the medical supports"*  
(Service Provider)

*"The neurosurgeons I must say, they provide a fantastic service, just amazing. And they always follow them up very quickly, if they have concerns about shunt malfunction"* (Service Provider)

With regards frequency of access to neurosurgery professionals, overall 69% of parents report their children was seen at least once in the previous year by a neurosurgeon, compared to only 9% who have never accessed a neurosurgeon (Figure 91).

There was however a significant difference in frequency of access to Neurosurgery depending on where the child had undergone their lesion repair ( $p=0.008$ ) (see table 16). Children who had their lesion repaired in TSCUH have much greater access to regular neurosurgical review; 86.6% accessing neurosurgery at least once a year, compared to 51.5% in Crumlin, 33.4% in Cork and 66.7% in Beaumont. 15.2% of children who had their lesion closed in Crumlin, and 16.7% of children who had their lesion closed in Cork had never seen a Neurosurgeon, which reflects the former practice of general surgeons completing lesion repairs in these sites.

Table 17

Neurosurgery access depending on where **lesion closed**

NEUROSURGERY	3 monthly or more	Twice a year	Once a year	Not this year	Never
Temple Street Children's University Hospital	22.9	21.7	42.0	11.6	2.9
Our Lady's Hospital Crumlin	0.0	3.0	48.5	33.3	15.2
Cork University Hospital	0.0	5.6	27.8	50.0	16.7
Beaumont	0.0	0.0	66.7	33.3	0.0

## Discussion

Just over half of the current sample had their lesion surgically repaired in TSCUH (53%) with the next largest group having this completed in OLCHC (24%). The age spread was unsurprising with most of the younger children attending TSCUH and the older children attending OLCHC or other centres. This reflects the reconfiguration of neurosurgical services. Following the move, some resources were put in place for the specialist spina bifida service in TSCUH however many of the resources were subsumed by the broader needs of

Neurosurgery within the hospital resulting in very limited resources available to the children with spina bifida.

The current study highlights benefits reported by parents and service providers when access to one national neurosurgical centre was available. However disparities with access to neurosurgical services still exist. Similar variation in infrastructure and access to neurosurgical services was noted in England by Steers and Stower (2010). Children born before September 2008, who were not previously linked with TSCUH, are unable to access the national paediatric neurosurgical centre in TSCUH. These children may have access to neurosurgery through other hospitals however consistency and standardised access to neurosurgical care is recommended to support children with spina bifida.

## Recommendations

1. All children with spina bifida should have access to a paediatric trained Neurosurgeon for emergency advice and support.
2. A paediatric Neurosurgeon should be available to review children with spina bifida throughout the lifespan annually as part of the multidisciplinary team clinic review.

## Access to Urology

### Background

As discussed in more detail in chapter 4.1 Physical Impact on the Child (Bladder and Bowel Impact), the majority of children with spina bifida present with bladder and bowel dysfunction and require various investigations, evaluations and interventions to support these needs. Although 90% of children with spina bifida are born with normal renal function, 50% may suffer from upper urinary tract damage due to lower urinary tract hostility (Bauer and D, 1990) therefore highlighting the need to access urology professionals.

In the Irish context one study highlighted increased pressure on the urological speciality in both paediatric and adult care due to the fact Ireland has the “lowest ratio of consultant urologist to population of all the European Board of Urology affiliated countries” (Cahill and Kiely, 2003 p. 182). Efforts to recruit a paediatric urologist to CUH Temple St. have not been successful. There is limited access to paediatric Urology in OLH Crumlin. There is a Urologist available in Cork. In order to investigate this further, parents and service providers were asked who was involved in supporting children’s bladder and bowel needs, their access to Urologists and to Spina Bifida Nurse Specialists. Results are presented in relation to overall access to urology, how this differs in each HSE region and how this differs based on which hospital the child’s spina bifida lesion was repaired as this impacts access to hospital services for children born before 2008.

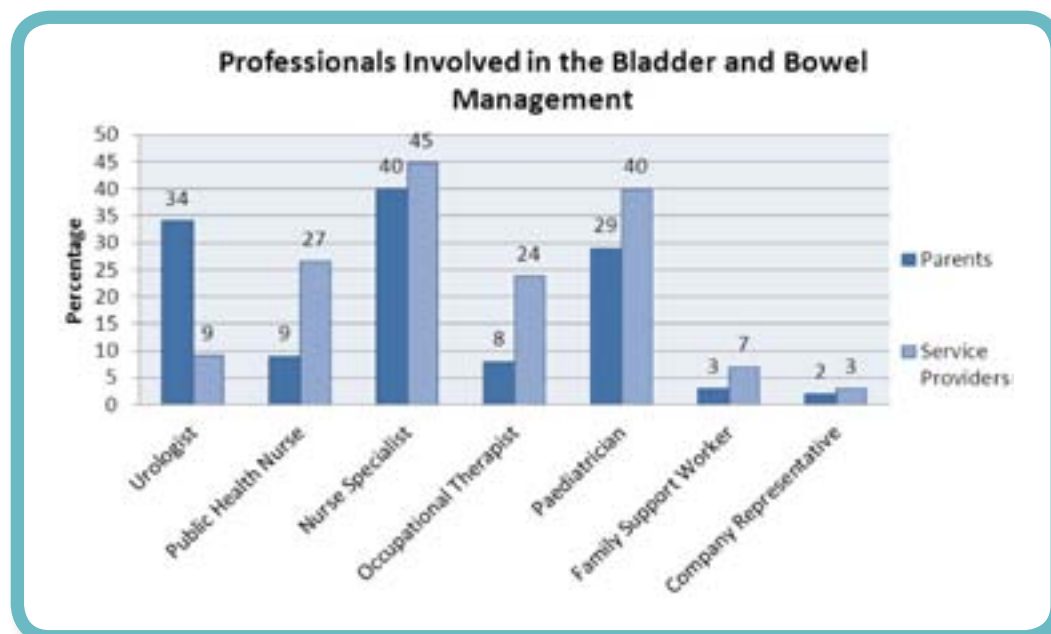
## Results

### Professionals Involved

Parents and service providers were asked which professionals they felt were involved in addressing the bladder and bowel continence needs of children with spina bifida (see Figure 80). The majority of parents (74%) report their child had at least one professional involved in addressing their bladder and bowel continence needs. Parents identified the Nurse Specialist (40%), Urologist (34%) and Paediatrician (29%) as the professionals most involved in addressing the bladder and bowel continence needs of children with spina bifida. Service

providers highlighted the most involved professionals to include the Nurse Specialist (45%), the Paediatrician (40%) and the Public Health Nurse (27%). However not as many service providers highlighted that a Urologist was involved in this management. This reflects the situation at the time of the study where there was no surgeon on the multi-disciplinary team at Temple Street addressing urological issues.

Figure 92



## Urologist Access

In the current sample there were large numbers of children (38%) who had never accessed a Urologist (Figure 92). Also despite the young age of the overall sample an additional 25% of the sample report their child had not seen a Urologist in the previous year.

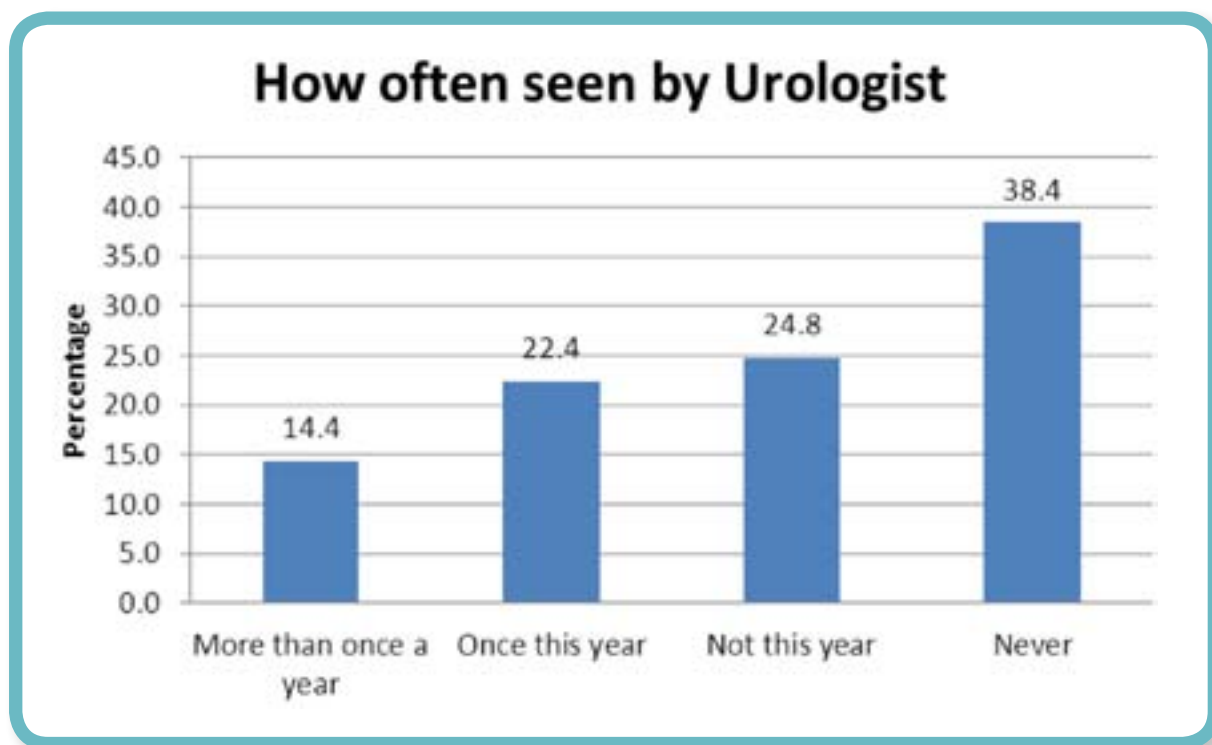
Access to professionals also varied nationally with better access to Urologists reported in the south of the country (HSE South) with 28% who had never accessed a Urologist compared to 50% in the Dublin Mid Leinster Region. See figure 93 for more information. Figure 93

Figure 18 Frequency of urology access based on where lesion closed

UROLOGY	3 monthly or more	Twice a year	Once a year	Not this year	Never	TOTAL
Temple Street Children's University Hospital	6.3	7.9	12.7	23.8	49.2	100
Our Lady's Hospital Crumlin	0.0	15.6	28.1	28.1	28.1	100
Cork University Hospital	5.9	11.8	29.4	35.3	17.6	100

A trend was evident suggesting that a greater proportion of children who had their lesions closed in TSCUH or Beaumont had never accessed a Urologist (49.2% and 66.7%) compared to those who had their lesion closed in OLCHC or Cork (28.1% and 17.6%). Only 3 children had their lesions surgically repaired in Beaumont hospital none of whom had accessed a Urologist in the previous year therefore their figures are excluded from the table below.

Figure 93



The lack of Urology services available was one of the main areas of concern and frustration raised by parents and service providers in interview, both in terms of investigations and interventions. This resulted in anxiety about the child's future kidney function. In interviews comments included:

*"Urology is a big deficiency" (Parent / Guardian)*

*"What they miss is their urology bladder, bowel and urology" (Service Provider)*

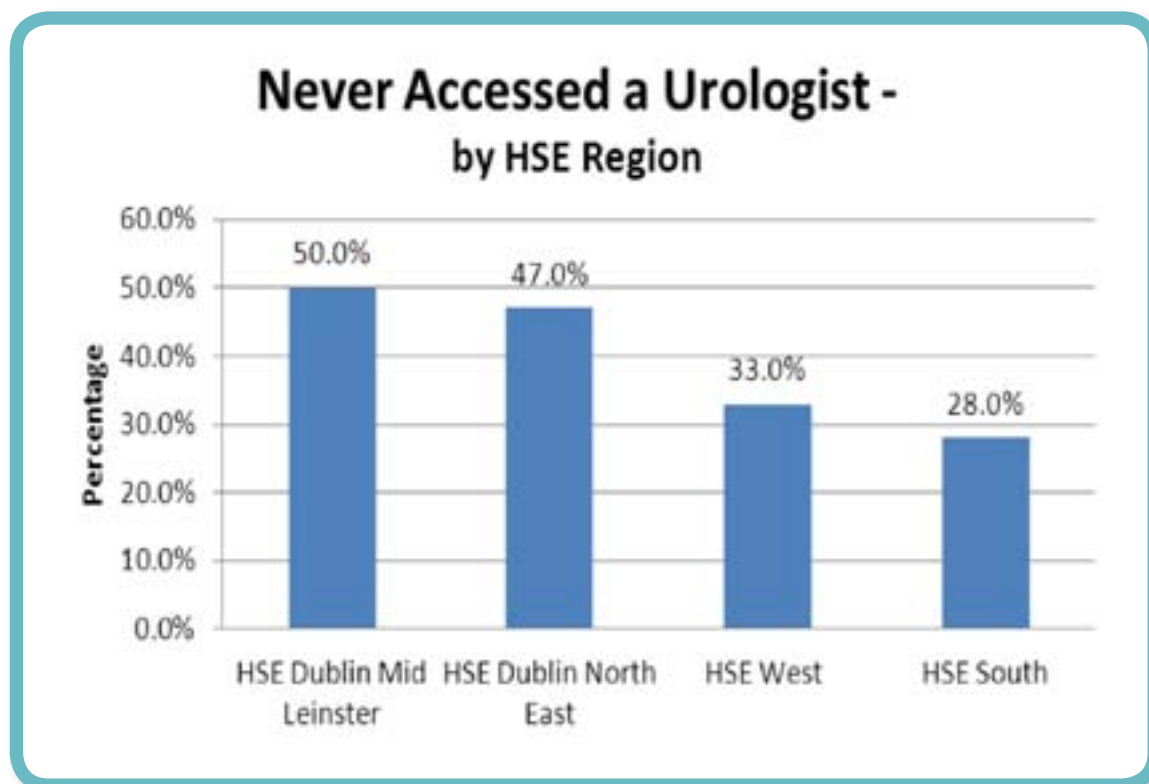
*"Access to Urology, I think that's the really critical one, there are two things that kill kids with spina bifida and when shunts are well looked after, that just leaves their kidneys, and we have to look after their kidneys. And I think it's neglect that's going on, and an ultrasound once a year, doesn't even touch the bare minimum"*  
(Parent / Guardian)



*“The Urology point of view is disjointed and so we just wing that here locally to be quite honest with you” (Service Provider)*

*“To be honest with you it’s the parents manage it” (Service Provider)*

Figure 95

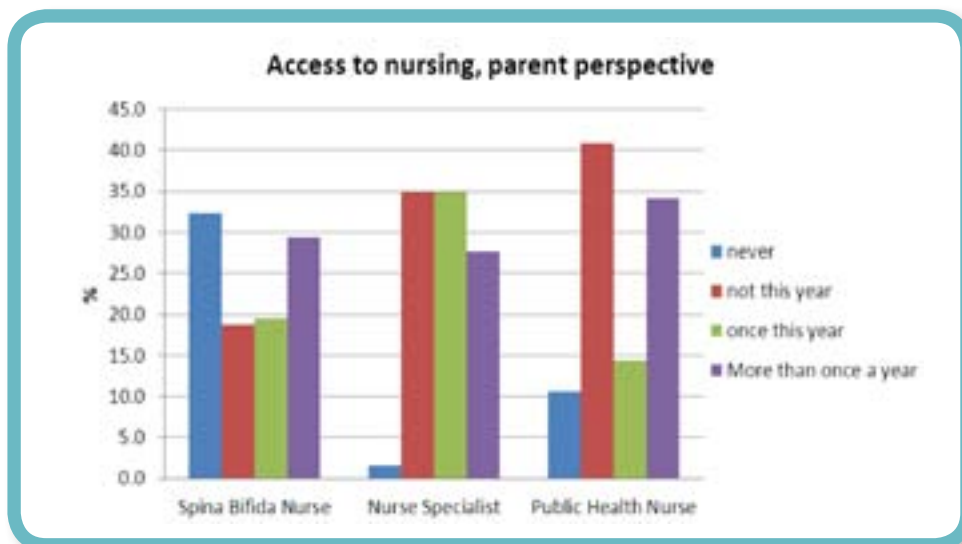


### **Nurse Specialist for Urology**

In relation to accessing Nurse Specialists, 32% of parents reported that they had never seen a Spina Bifida Nurse Specialist (Figure 94). Parents were not specifically asked about access to other Nurse Specialists who may be involved including Urology Nurse Specialists, further research is required to investigate access to other Nurse Specialists not specific to spina bifida. Access to a Spina Bifida Nurse Specialist also varied depending on where the child’s spina bifida lesion was repaired with only children attending TSCUH having access to this service. These results are discussed in more detail in the later section detailing access to Spina Bifida Nurse Specialist not solely in relation to urology.



Figure 95



Both parents and professionals recommended the need for more specialist nurses locally to provide education, training and support in relation to continence needs. They commented that where this was available, it was positively received. As well as the lack of specialist service, local service providers were not confident in meeting the bladder and bowel needs of these children and requested information/support from specialist services. While some continence nurses are available throughout the country, these services are in place to cover the whole lifespan, and at present are not sufficiently specialised or resourced to assist with training for CIC and other procedures for children with spina bifida. Parents similarly found local services lacking in knowledge about bladder/bowel needs.

*“The parents would love to meet the incontinence nurse, sometime and they don’t really have access now that’s difficult, they are coming to me and they are asking questions and I don’t really have an answer - I find this difficult.”*  
(Service Provider)

*“We formerly had a specialist nurse who provided advice particularly around bladder and bowel issues, but that nurse hasn’t been replaced and I think that’s been a great loss”* (Parent / Guardian)

*“There are continence nurses in the community but they’re overwhelmed because they’re birth to grave for all people”* (Service Provider)

*“I suppose having a continence nurse more locally...There isn’t someone that is really specialised”* (Service Provider)

## Discussion

Results suggest that the lack of availability of urology services in Ireland is a concern for both service providers and parents. Access to both Urologists and Nurse Specialists who assist with the management of bladder and bowel function was seen to be poor. Complex bladder and bowel difficulties associated with neurogenic bladder and bowel are often present in children with spina bifida and difficulties such as incontinence can have vast physical and psychological impacts on the child and the family (Verhoef et al 2005). This was echoed by parents and families in this study with comments made on the social impact of incontinence for the child and expressed as a concern for psychosocial wellbeing.

Therefore early and frequent access to the appropriate professionals is required to ensure bowel and urine continence and the best quality of life possible for the children. Local service providers lack the expertise of management of the complex bladder and bowel needs of these children, requiring advice from specialist professionals.

The access to urology professionals, while poor generally, was dependent on where the child had the spina bifida lesion repaired, suggesting a geographical inequity. Earlier evaluation and more vigilant investigations are required to ensure the necessary monitoring of the complex bladder and bowel needs of these children.

## Recommendations

- 1 All children should have access to a Urologist with a specialist interest in spina bifida to ensure early surveillance and management to support their bladder and bowel needs.
- 2 Trained nursing professionals should be available outside the national centre to assist with support and training of families and children with regards to achieving social continence. This support could be provided through up-skilling of existing continence nurses, or training public health nurses or early intervention nurses to assist with these interventions.

## Access to Orthopaedics

### Background

The aims of orthopaedic management of a child with spina bifida have been described by a number of authors:

‘preventing joint contracture, correcting deformity, preventing skin sores, and obtaining the best possible locomotor function’ (Mazur and Menelaus, 1991)

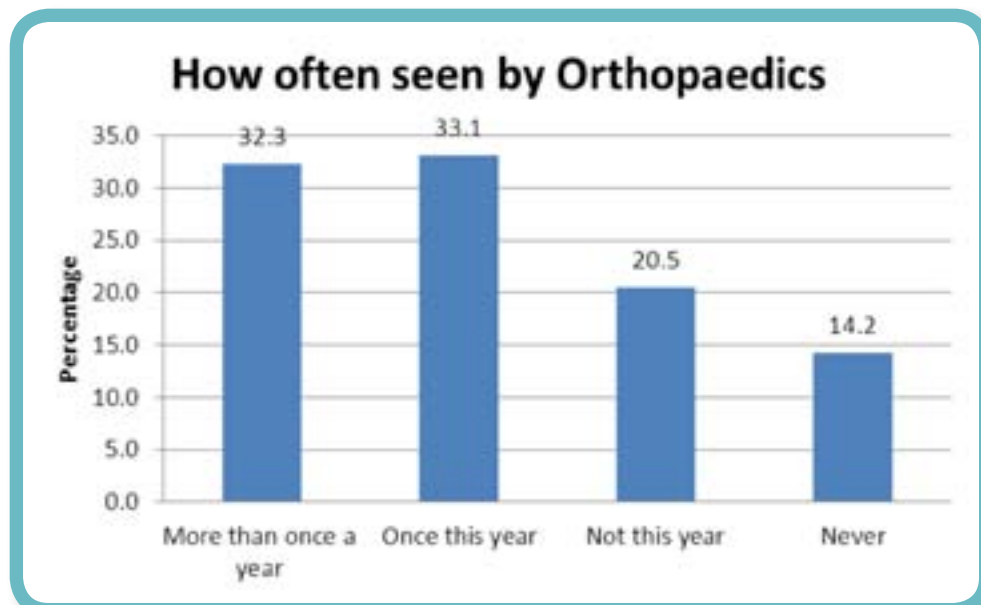
‘maintaining, restoring, or preventing anticipated future decline in physical function’ (Wright, 2011)

‘preventing or correcting deformities to maximize mobility and independence while staying within the realistic expectations of the patient’s functional neuro-segmental level’(Thomson and Segal, 2010)

The common factor as described above is on maintaining and promoting function in order for the child to achieve physical well-being. The impact of independent mobility on quality of life for children with spina bifida has been reported (Schoenmakers et al., 2005). As discussed in Chapter 4.1 the physical impact of spina bifida on the child, children can present with various orthopaedic complications including hip dislocations, (Broughton et al., 1993), scoliosis (Trivedi et al., 2002) foot deformities (Brown, 2001) and fractures (Dosa et al., 2007).

Although the focus for orthopaedic surgeons is on physical function, the positive effects that improved physical function can have on mental health, social function, and overall physical fitness in children and young adolescents cannot be underestimated (Wright, 2011). Thomson and Segal, (2010) discussed the importance of ensuring that orthopaedic surgery relies on patient function rather than simply radiographic imaging along in order to be able to promote independence.

Figure 96



In a US review of Spina Bifida clinics (Delmarva Foundation, 2006) found that 87% of Spina Bifida clinics reported having an Orthopedist available at clinic; and various studies suggest that a child should be reviewed by an orthopaedic surgeon at least annually unless more frequently indicated (Merkens, 2006, Delmarva Foundation, 2006).

Table 19

Frequency of orthopaedics access based on where lesion was closed

ORTHOPAEDICS	N=	3 monthly or more	Twice a year	Once a year	Not this year	Never
Temple Street Children's University Hospital	81	9.0	29.9	40.3	9.0	11.9
Our Lady's Hospital Crumlin	36	10.0	23.3	26.7	33.3	6.7
Cork University Hospital	20	5.6	11.1	16.7	33.3	33.3

Merkens (2006) guidelines for children with spina bifida across the lifespan recommends review from as early as infancy to ensure that any indicated orthopaedic surgeries are performed as well as regular and individualised orthopaedic evaluations throughout the lifespan.

## Results

Parental questionnaire responses suggest that in the previous year 65% of children had seen an orthopaedic surgeon.

Access to Orthopaedic services was not affected by where the child had the lesion surgically repaired ( $p=0.345$ ). Most children in this sample had their lesion closed in TSCUH, Our Ladies Children's Hospital Crumlin or Cork University Hospital with only 3 children who had their lesion repaired in Beaumont, due to their small numbers Beaumont numbers are excluded from the table below with regards orthopaedic access. Results suggest that 79.2% of children who had their lesion closed in TSCUH who had accessed Orthopaedics in the last year, 60% who had their lesion closed in Crumlin, 33.4% who had their lesion closed in Cork.

During qualitative interviews parents raised concerns regarding the waiting time to access orthopaedic surgery and appointments highlighting the impact of this on their child and family:

*“There’s a long waiting list for surgeries as well so this child I’m seeing was scheduled for surgery the summer and then was cancelled until next summer”  
(Service Provider)*

*“I think the orthopaedic service is under enormous pressure because of the numbers of patients that are referred and because of the paucity really of orthopaedic surgeons” (Service Provider)*

*“I actually had to go private because it was in relation to her scoliosis and it was rapid and needed it seen to, so there was a 22 month wait. So I went private and I was seen within a week” (Parent / Guardian)*

Parents and service providers both suggested the need for a specialist orthopaedic surgeon who will review all orthopaedic aspects of a child with spina bifida rather than differing appointments for different joints. There was also request for a level of specialism in the area of disability and spina bifida.

*“When you go to just the orthopaedic clinic, you’ve got every child of every description, so they’re not as specific I feel... they’re not just totally zoned in on spina bifida children they’re dealing with everything on the day.”  
(Parent / Guardian)*

*“One orthopaedic person that could get really involved because it’s very hard to know who you send them to, you know for backs it’s here, for feet it’s here.”  
(Service Provider)*

*“We went up to (orthopaedic surgeon), because he was an orthopaedic surgeon, but he said, he doesn’t concentrate on the top half of the body.”  
(Parent / Guardian)*

With respect to orthopaedics, mixed feedback was received from parents regarding the ease of access to orthopaedic surgeons and communication with families and teams.

*“Communication needs to be improved” (Parent / Guardian)*

*“It was like someone opened the door and the sun was shining. We went in and got her x-ray, he was helpful, he was able to talk to her, and he was able to talk to us. He explained everything”  
(Parent / Guardian)*

## **Discussion**

Orthopaedic care of the child with spina bifida is vital to prevent and correct contractures and deformities and to help achieve independent mobility (Mazur and Menelaus, 1991, Thomson and Segal, 2010) . Children with spina bifida present with multiple orthopaedic complications which increase with frequency and complexity as they age (Broughton et al., 1993, Trivedi et al., 2002). Therefore regular review by an orthopaedic surgeon is necessary to advise and intervene when appropriate (Merkens, 2006, Delmarva Foundation, 2006).

Debate continues in the literature in relation to timing and benefits of orthopaedic intervention, but suggests a functional and participation outcome measure be used. Access to orthopaedics was high in the current study (67% accessing within the last year), although the wait time for surgical intervention was a cause for concern. Both service providers and parents recommend the need for a single orthopaedic surgeon to attend to all needs of the child with spina bifida and that this professional be present at the spina bifida MDT clinic and have dedicated theatre space.

## **Recommendations**

1. A dedicated paediatric disability orthopaedic surgeon to review the whole child and be part of the MDT Spina Bifida clinic is required.
2. Increased access to theatre space and resources are also required to reduce lengthy waiting times to access surgery.

## Access to Spina Bifida Nurse Specialist

### Background

As discussed in the access to Urology section, Spina Bifida nurses are often involved in assisting with the management of bladder and bowel difficulties. The role of the Spina Bifida Nurse Specialist includes various other aspects of care which should not be overlooked. The nurse coordinator for a multi-disciplinary spina bifida service has been seen to have the most knowledge and information about the family situation and plays an important role in co-ordinating care, educating and triaging the child when difficulties arise (Dunleavy, 2007). Brei (2007) commented on the increasing role of nurse practitioners, which is a US equivalent of a nurse specialist, in the care of children with spina bifida. At the time of the questionnaires there was only one spina bifida nurse in Ireland - working within the national spina bifida MDT service in TSCUH. Therefore this service was only accessible to a portion the children aged 0-18 with spina bifida in Ireland.

### Results

Access to a spina bifida nurse specialist was poor, with 32% of parents reported that they had never been seen by a spina bifida nurse (Figure 87). It is important to note however that a few nurse specialists (e.g. Intellectual Disability, Palliative Care) not specific to spina bifida, are present in some regions, and support families with different aspects of their physical disability. However access to this is not consistent, and parental report about accessing a nurse specialist may refer to other clinical specialists related to orthopaedics or urology where available.

Access to a Spina Bifida Nurse was significantly different based on where the child had their lesion surgically repaired ( $p=0.014$ ). There was significantly more access to a Spina Bifida Nurse for those children who had their lesion closed in TSCUH with 85.9% seeing a spina bifida nurse once a year or more compared with 6.2% of children who had their lesion closed in Our Lady's Children's Hospital Crumlin, 5.6% of children who had their lesion closed in Cork, and none of children who had their lesion closed in Beaumont

Figure 97

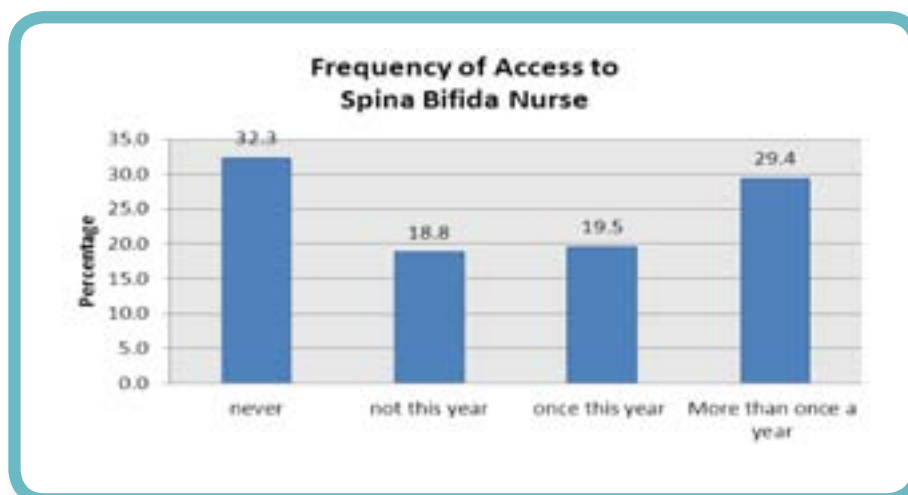


Table 20

Frequency of Spina Bifida Nurse access based on where lesion was closed

SPINA BIFIDA NURSE	3 monthly or more	Twice a year	Once a year	Not this year	Never
Temple Street Children's University Hospital	33.8	18.3	33.8	8.5	5.6
Our Lady's Hospital Crumlin	0.0	3.1	3.1	34.4	59.4
Cork University Hospital	0.0	0.0	5.6	22.2	72.2
Beaumont	0.0	0.0	0.0	66.7	33.3

## Discussion

Despite the growing recognition for the role of a spina bifida nurse specialist/coordinator or nurse practitioner in the case of the US (Dunleavy, 2007, Brei, 2007) access to this specialist clinician is very poor in the Irish context. This is dependent on where the child had the neonatal surgical repair, resulting in only children who attend TSCUH having better access to a spina bifida nurse.

## Recommendations

1. A spina bifida nurse specialist should be available for all children and families to access to assist with coordination of care, specialist education and interventions.
2. Specialist multidisciplinary spina bifida care and clinic



# The Multidisciplinary Spina Bifida Clinics

## Background

There are multiple benefits of multidisciplinary spina bifida care and specific clinics in order to provide on-going comprehensive and co-ordinated quality care (Brei, 2007, Brustrom et al., 2012, Delmarva Foundation, 2006, Aldana et al., 2010). The Children's Neurosurgical Specification Standards (NHS., 2011) recommend that centres ensure as a minimum on-site multi-disciplinary team including data collection manager, Neuropsychologist, Dietitian, Play Therapist, Physiotherapists, Occupational Therapist, Speech and Language Therapist, Paediatric Pharmacist (access to advice at all times), Hospital School Teacher (access to service) all who have specialised training in managing patients within paediatric neurosurgery and Paediatric Neuro-Rehabilitation. The MDT must also have access to an orthotist and Child and Adolescent Mental Health Services at the relevant stages in the child's care (NHS., 2011). NHS (2011) also recommends that children with NTDs have access to a specialist MDT for review, on-going rehabilitation and support, which should also include good communication and coordination.

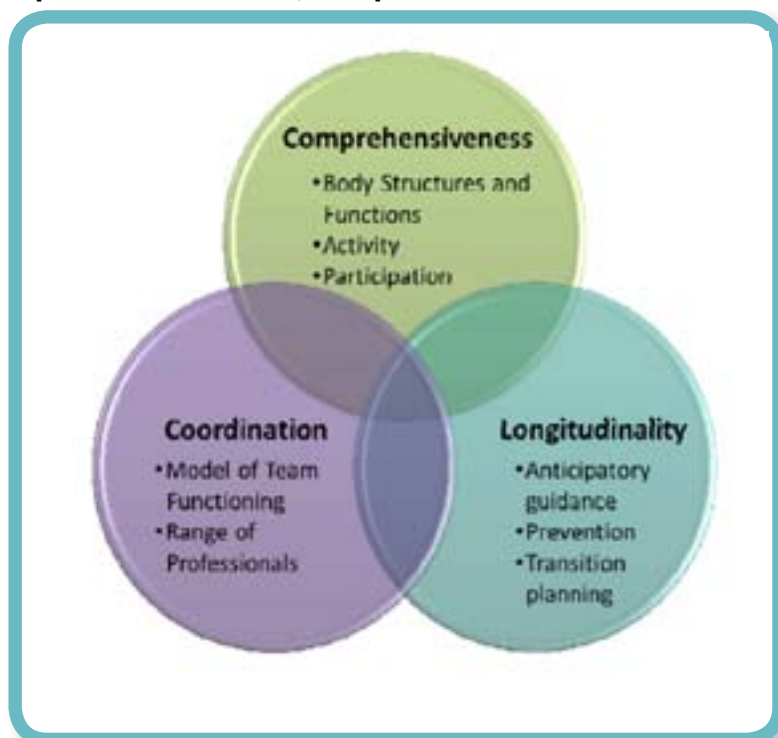
Merkens (2006) guidelines for the healthcare of individuals with spina bifida assumes the receipt of ongoing, comprehensive, coordinated care throughout the lifespan, which includes referral to a Spina Bifida multidisciplinary clinical team from as early as possible after receipt of an antenatal diagnosis. The guidelines state that there is "sufficient data to recommend that care or patients with spina bifida should be delivered in the context of a multidisciplinary clinic" (Merkens, 2006).

Having a multidisciplinary perspective including multiple disciplines was seen as vital in spina bifida due to the complexity of these children and the need to be responsive to their variable outcomes (Fletcher and Brei, 2010). The negative outcomes of disbanding of a multidisciplinary clinic for children with spina bifida have been reported in the United States when Kaufman et al. (1994) surveyed 87 parents three years after their clinic closed. Results suggested poor medical follow-up and an increase in serious morbidity and recommended coordination of care to ensure that care is effective.

To ensure a functional perspective in Spina Bifida clinics, Kinsman et al. (2000) recommends a new model of spina bifida integrating both medical and rehabilitation services. The model consists of three dimensions (See figure 96): firstly comprehensiveness, highlighting the need to meet the multiple health and functional issues as identified in the ICF (formerly ICIHD); next is coordination which is required to establish the model of team functioning which requires a range of professionals; and finally the third dimension of longitudinality is introduced which involves anticipatory guidance over time, prevention through education and planning for major transitions (Kinsman et al., 2000).

Figure 98.

### Spina bifida Model, Adapted from Kinsman et al 2000



Multidisciplinary Spina Bifida clinics can increase collaboration, care coordination, can reduce family burden and result in greater convenience for families (Brei, 2007, Brustrom et al., 2012). More research is required regarding the coordination, and structure of multidisciplinary Spina Bifida clinics and how information is shared as well as their cost-effectiveness to ensure sufficient personnel and funding (Brei, 2007, Delmarva Foundation, 2006).

While there is growing body of literature to support the importance of running multidisciplinary Spina Bifida clinics, gaps in the literature still exist in relation to the logistics of running such a clinic. Most international studies or reviews recommend regular follow-up in a dedicated MDT Spina Bifida clinic, with at least an annual review with a variety of professionals (Liptak and El Samra, 2010, Delmarva Foundation, 2006). A national review of Spina Bifida clinics in the United States (Delmarva Foundation, 2006) identified that an average of 8 patients are reviewed per clinic with a 4 hour average clinic duration with considerable variance in the coordination of care, type of care provided and staff available to provide these services. The review found that the following professionals were available in over 60% of Spina Bifida clinics: Urologist, Orthopedist (Orthopaedic Surgeon), Physical Therapist (Physiotherapist), Social Worker, Neurosurgeon, Clinical Nurse Specialist, Occupational Therapist, Paediatrician and Dietitian/Nutritionalist with 87% of clinics having a dedicated care coordinator as well (Delmarva Foundation, 2006). During the time of data collection, the Irish National Multidisciplinary Spina Bifida clinic which is based in TSCUH was only staffed with 4 out of the 10 aforementioned professionals, namely: Physiotherapist, Neurosurgeon, Clinical Nurse Specialist and Paediatrician. It is important to note that the Physiotherapist and Paediatrician were not adequately resourced resulting in them working to full capacity (despite a growing caseload). Also of note, since October 2013 there has been a visiting surgeon with an interest in bladder and bowel difficulties in attendance at the clinic and during the completion of this research project an Occupational Therapist was also in attendance thanks to the assistance of the TSCUH fundraising department while completing this research project.

Other literature would support the Delmarva (2006) findings and the need for multiple professional with others listed to include psychiatry, clinical psychology, speech and language

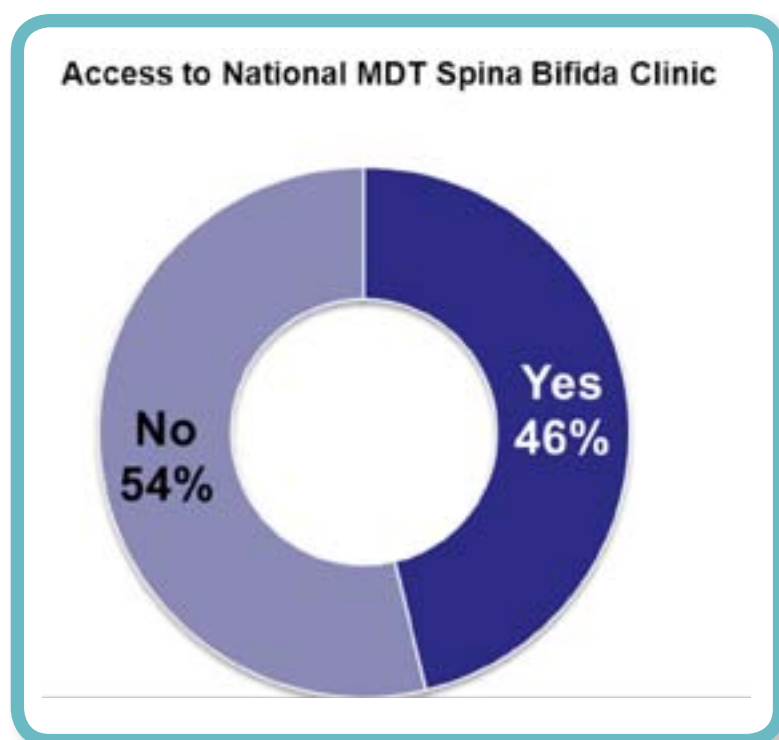
therapy/pathology, gastroenterology nurse and administration (Aldana et al., 2010, Dunleavy, 2007).

When no multidisciplinary Spina Bifida clinic is in place, Kaufman et al. (1994) highlighted the possibility of significant difficulties with coordination or care, as neither parents nor clinicians coordinate the child's care thus resulting in adverse effects. However when good communication exists between clinics and families this can facilitate and assist with the development of trust, (Brustrom et al 2012) and is therefore a vital component for the functioning of any Spina Bifida clinic. It is also vital that as well as ensuring care for children that funding, resources and programmes are available for adults with spina bifida also as this is seen as greatly lacking in the literature (Brei 2007). The TSCUH service established a multidisciplinary Spina Bifida clinic in 2011. This clinic currently has approximately 220 children on its caseload. However the population is increasing with an average of 35.4 new children born with spina bifida per year.

At present the clinic usually runs for approximately 5 hours on a monthly basis and will see on average between 12 and 14 children per clinic. A previous MDT clinic was available in Our Lady's Children's Hospital in Crumlin. However once the service moved to TSCUH this service was disbanded. While some older children with spina bifida still attend services in Our Lady's Children's Hospital in Crumlin, a coordinated MDT Spina Bifida clinic is no longer available.

## Results

Figure 99



Parents were asked whether they had access to a MDT Spina Bifida clinic. In interview both parents and service providers were also asked for feedback regarding the functioning of the MDT Spina Bifida clinic.

Only 63% out of the 134 parents questioned, reported their child had access to a Spina Bifida clinic. Of those 46% (62 children) attended the National MDT Spina Bifida Clinic which is located in TSCUH as detailed above and is known to be the only MDT Spina Bifida clinic

in Ireland consisting of both medical professionals (consultants) and health and social care professionals (therapists and nursing). The only other spina bifida specific clinic which parents report attending is located in Cork University Hospital which consists of medical and nursing professionals with no therapy input. Only 46% of parents report having access to the national MDT Spina Bifida Clinic in TSCUH.

Positive comments from parents and service providers relating to access to the MDT Spina Bifida clinic included:

*“The Spina Bifida clinic is good, the children get an overall view” (Parent / Guardian)*

Parents who had previously attended the Spina Bifida MDT Clinic in Our Lady’s Childrens Hospital in Crumlin expressed disappointment and frustration that this service had disbanded and that they could not access the national MDT Spina Bifida clinic in TSCUH. This has resulted in very dispersed care, when they would prefer more centralised care.

*“And we were left sitting with no service, we fell through the holes” (Parent / Guardian)*

*“It would be lovely if there was a team, that urology, orthopaedics, physio, OT and that they all knew her and her situation” (Parent / Guardian)*

*“That’s the difficulty here, because those kids who are attending Spina Bifida clinics they are looked after, but those who are older, it’s very difficult at the moment ... I don’t feel they are looked after in that perspective well enough, and I’m talking about older kids” (Service Provider)*

*“If there was a one stop clinic or I suppose a one stop centre”(Parent / Guardian)*

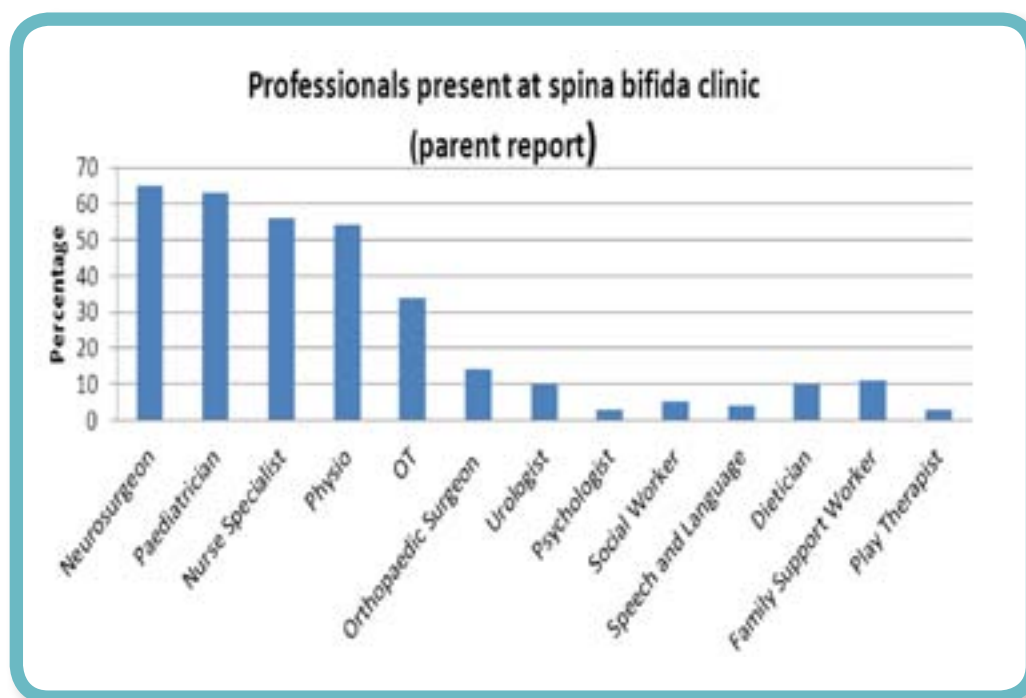
Where available, both parents and service providers commented on the benefits of seeing multiple professionals in a ‘one-stop shop’ and both felt reassured by that specialist review. However the role of this clinic was not always clear, and parents would like the clinic to occur more frequently.

*“That would definitely help, defining the role (of clinic) from your point of view, what the clinic aim is and what services are available to them there. Is something that is already being done locally if it doesn’t need to be looked at or is there definitely a gap in the child’s local service that needs to be resolved, so I guess we don’t really have a very clear understanding of what service you are providing and I don’t know if you have a very clear understanding of what service we are.”*

*(Service Provider)*

Of those that do attend a Spina Bifida clinic, parents were asked to comment on whether certain professionals were or were not present. The results are outlined below (Figure 90). The majority of parents reported that a neurosurgeon, paediatrician, nurse specialist and physiotherapist was present at their clinic. Some parents also commented on the presence of an Occupational Therapist, while other professionals were reported to be present less frequently.

Figure 100



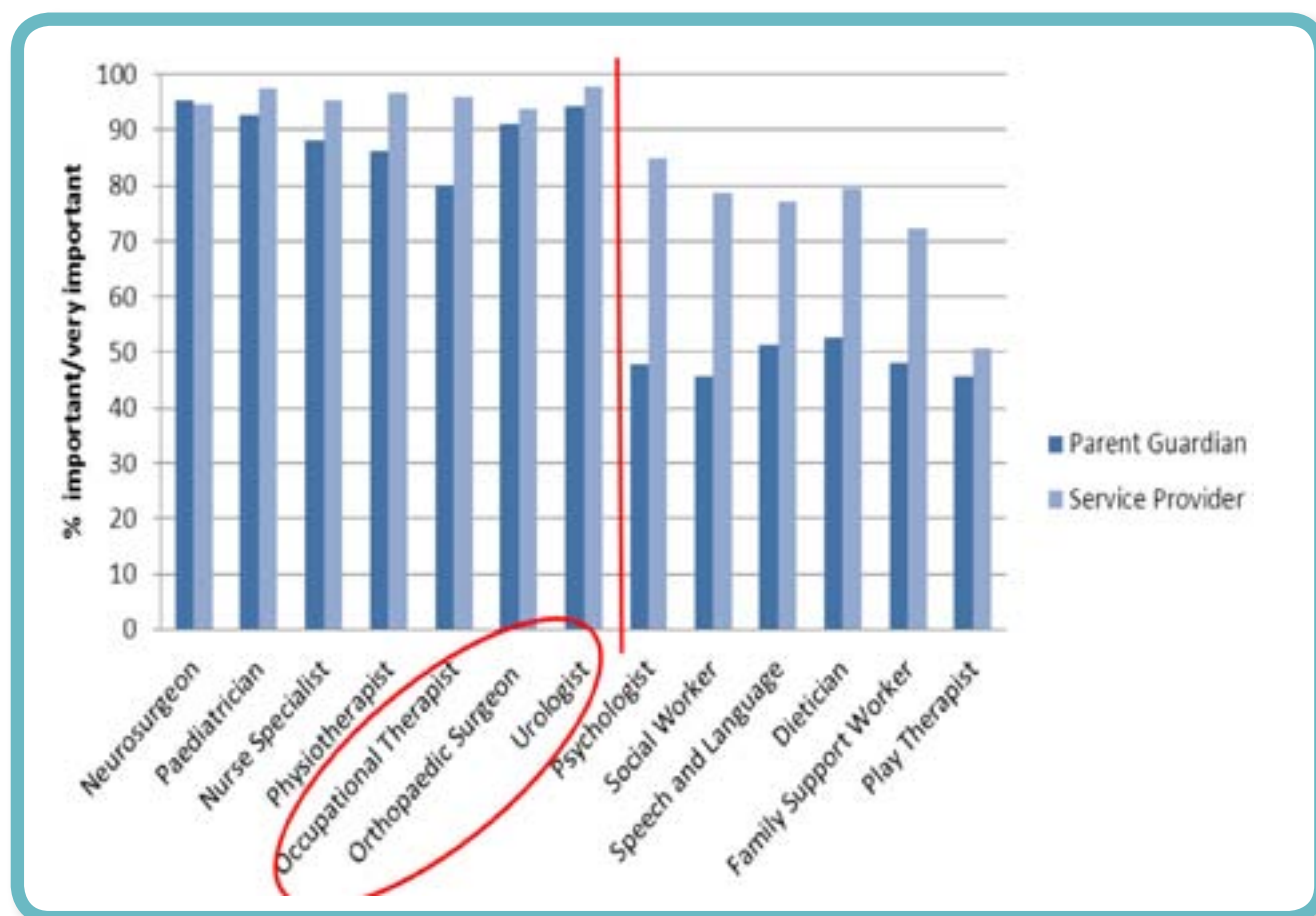
Parents and service providers were both asked their opinion on how important they felt the presence of professionals was for the effective running of a specialised Spina Bifida clinic. They were asked to rate the importance placed on professionals on a 5 point Likert scale including very important, important, moderately important, of little importance or unimportant.

Over 80% of both parents and service providers agreed that the presence of a Neurosurgeon, Paediatrician, Nurse Specialist, Physiotherapist, Occupational Therapist, Orthopaedic Surgeon and Urologist at a specialised Spina Bifida clinic was either important or extremely important. This is represented in figure 99 with all professionals seen as essential to the left side of the

vertical red line. The three professionals circled, as previously mentioned, are currently not available at the MDT Clinic in TSCUH.

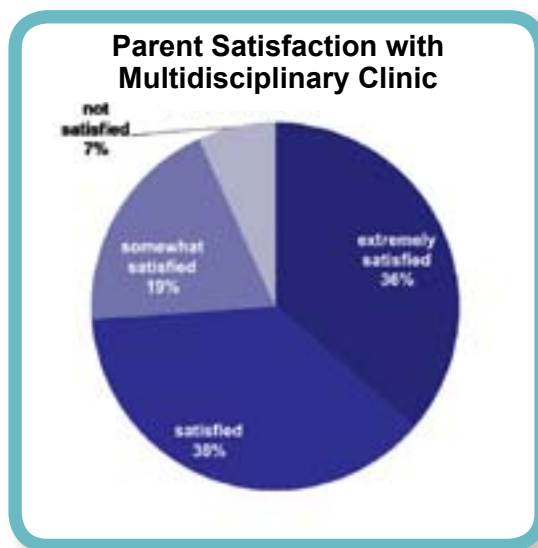
Figure 101

### Professionals whose presence at clinic was seen as very important



Parents attending a Spina Bifida clinic were asked their level of satisfaction on a 4 point Likert scale (Figure 100). Parental satisfaction with the clinic was very high with 36% being extremely satisfied and 28% satisfied with this service. 19% were somewhat satisfied and only 7% who reported being not satisfied. Comments included infrequency of review and the lack of a full MDT present.

Figure 102



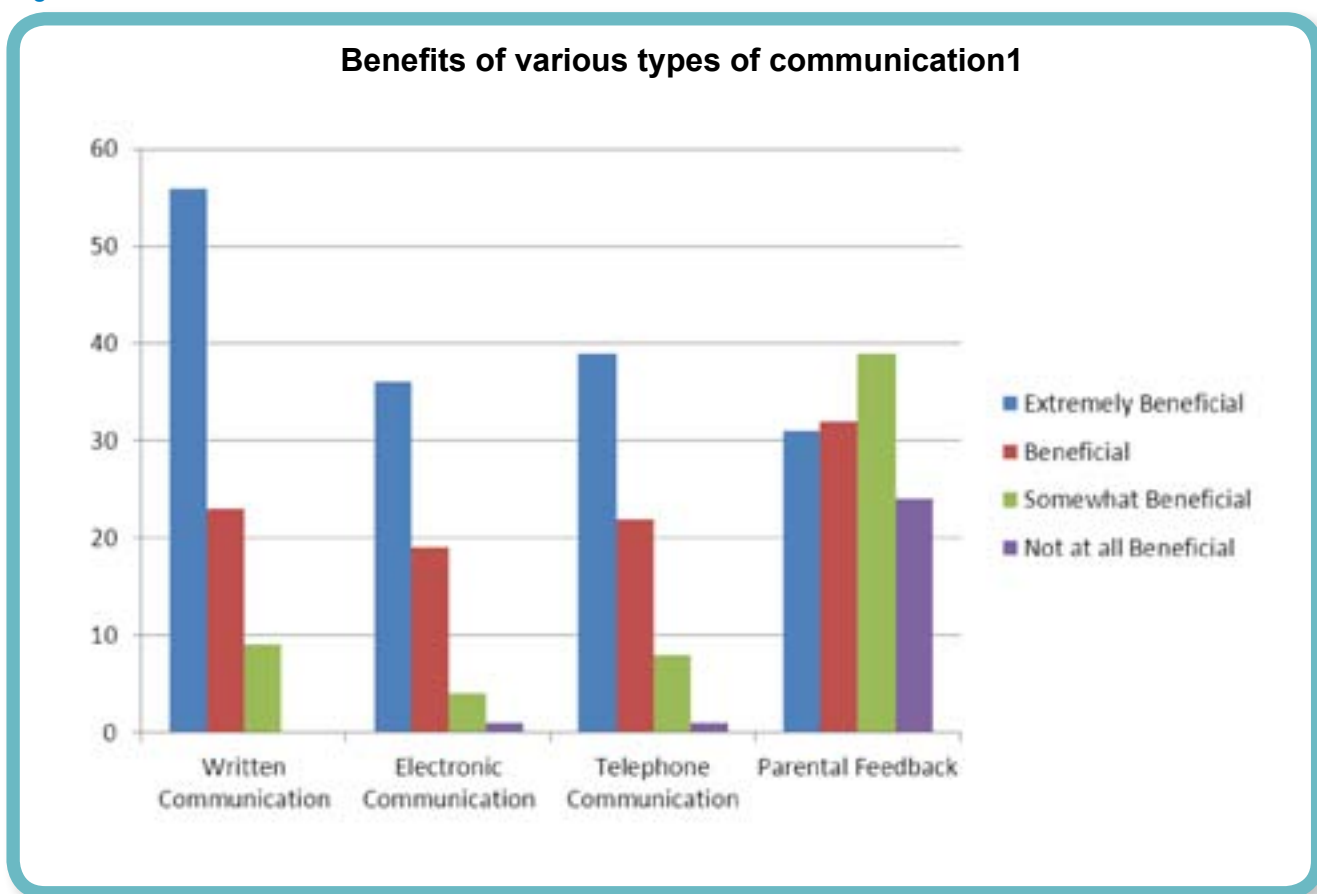
Service providers were asked whether any of their clients had attended a specialist Spina Bifida clinic and how beneficial overall to their work they found the MDT Spina Bifida clinic. 53% of service providers reported this was either beneficial or extremely beneficial. While the clinic was seen a beneficial to their role, service providers would like further education and training available to both parents and local services to ensure clarity of roles and increase understanding about how to optimise care.

*“I would love for the team to come down and give us a nice good session, educate us that is something that would be very welcome” (Service Provider)*

Service providers, although recommending a specialist service for a thorough overview, expressed concerns that local services remain involved and not be overlooked by parents. Perceptions were gathered regarding modes of feedback received from the MDT Spina Bifida clinic in relation to satisfaction with types of communication including written communication (letters), electronic communication (emails), telephone communication and parental feedback. The most beneficial form of communication appeared to be written communication (letters) and the least was parental feedback (see Figure 102). It appears that feedback to service providers from professionals present at clinic was more valued than relying on parents to relay that information between different services.



Figure 103



## Discussion

Results demonstrate poor access to the national MDT Spina Bifida clinic, with more than half of the national sample not having access, despite the fact that MDT care is recommended internationally (Merkens, 2006, Aldana et al., 2010). For those who did have access to the national MDT Spina Bifida clinic, various benefits were reported, with parents being mainly satisfied with the clinic and service providers finding it beneficial to their role.

Seven key professionals were identified by parents and service providers as being important or extremely important to be present at a MDT Spina Bifida clinic. However at the time of data collection the clinic was not staffed adequately with no Urologist, Orthopaedic Surgeon and Occupational Therapist employed to attend the clinic. Access to other professionals is also required, even if not directly present at clinic, including social work, clinical psychology, speech and language therapy, family support workers, and access to imaging services (Aldana et al., 2010, Dunleavy, 2007).

Issues related to resources available to the existing MDT Spina Bifida clinic were raised, with both the need for increased frequency of clinics to meet the needs of the increasing population, and the need for improved coordination and communication between professionals at the specialist and local level.



## Recommendations

1. A fully-staffed multidisciplinary spina bifida service should be available for inpatient and outpatient care with access to Neurosurgeon, Urologist, Orthopaedic Surgeon, Spina Bifida Nurse Specialist, Paediatrician, Physiotherapist, Occupational Therapist, Social Worker, Neuropsychologist, Nurse Specialists, Speech and Language Therapist, Dietitian, Neuro-ophthalmologist, Orthoptist, Play Therapist as well as access to imaging services, Radiology and administration support.
2. Children should be reviewed at the MDT Spina Bifida clinic on at least an annual basis.
3. A fully-staffed multidisciplinary Spina Bifida clinic is required which should consist of at least the following seven key professionals: Neurosurgeon, Urologist, Orthopaedic Surgeon, Spina Bifida Nurse Specialist, Paediatrician, Physiotherapist and Occupational Therapist.
4. The specialist spina bifida team is responsible for education and support of local teams.

## Communication

### Background:

Communication between specialist services and local services is essential to ensure consistency and a high quality of care. Pathways of care should be established between specialist and local services in a shared-care approach to try and provide as much care close to the child's home as possible (NHS., 2011). As part of coordinating care from a MDT Spina Bifida clinic, the Delmarva Foundation (2006) review found that almost all clinics were involved with coordination with community services and with coordination with schools and teachers. The NHS (2011) similarly highlights that good communication, coordination and clarity of team roles are essential within neurosurgical teams working with children with NTDs.

### Results:

Community service providers highly value communication from specialist centres where available and would like increased two way communication.

*“The communication has been excellent” (Service Provider)*

*“Communication is just so important. And I suppose having named people is helpful from that point of view” (Service Provider)*

*The need for improved communication was highlighted by parents and service providers also. Specific examples where improvements could be made were in speciality clinics including orthopaedics, urology and ophthalmology, which would aid the provision of appropriate resources locally. (Service Provider)*

*“I wouldn’t get that feedback. Maybe I think it comes down here and goes to their medical file, but it’s not sent around the disciplines here”  
(Service Provider)*

*“So you are depending on the parent to fill you in, which I feel is very unfair on the parents and a lot of pressure. And you know, some parents are very good at being on top of this information, and other parents find it just very overwhelming to try to explain to another consultant or other professionals, what a professional has said, so that causes a lot of stress” (Service Provider)*

*“We have never heard nothing from the hospital...but I’ve never looked either, it takes 2 for communication” (Service Provider)*

Specific comments regarding concerns with communication from the MDT Spina Bifida clinic included:

*“I’d love to know who I should contact” (Service Provider)*

*“We get all the reports... they might be a bit late, but we do get them” (Service Provider)*

*“There doesn’t seem to be consistent communication” (Service Provider)*

Methods suggested to assist with improved communication included electronic or paper patient file, video linking or outreach clinics to encourage sharing of expertise. One example might include the development of a patient passport (either paper or electronic) which children and families can be educated to utilise and transport to each appointment ensuring an up to date record of their medical and therapy needs.

*“I suppose... some passport, either a physical book or should we be looking at something electronically that the parent could carry that every time they're seen at a multidisciplinary meeting that the minutes or the recommendations of that are uploaded to this and you could have folders; a urology folder or whatever and you would be accessible by everybody, would be a way forward I think. And the parent could carry that information because we're already depending on them to carry it, and they're carrying it in their head - and you know sometimes in a clinic the dad might come or the parent who wasn't with them at Dublin appointment will come with them to the local review”*  
(Service Provider)

*“I mean the ideal would be, but it's national problem is, I suppose the shared files, where Temple St could have a shared folder on the children with spina bifida and the consultants then, the therapists in the area, could link into that”*  
(Service Provider)

Having a dedicated point of contact and coordination would be of benefit for local services to make contact with regards specialist concerns. This role could be completed by the spina bifida nurse coordinator if the post was sufficiently resourced to cater for all the children's needs.

*“It would be nice to have a link person that you could even call or email or something. You could say, this child has an issue, it's to do with x, y or z. And then even if they could say, you need to be talking to this person or that person. So I suppose a link person would be good, that one person to I suppose the idea of a key worker type of that you could contact them and they'd maybe tell you who to refer onto”*  
(Service Provider)

*“A phone call I find works very well”* (Service Provider)

## **Discussion**

Results suggest that communication between specialist services and local teams requires development. While positive comments were received in relation to coordinated care, particularly following MDT reviews, many parents and service providers provided suggestions as to how communication could be further improved. Having either a paper or electronic passport for each child which would store relevant information would be of benefit and would help reduce anxiety and confusion with parents having to transport information between professionals.

Communication from the spina bifida team before the MDT clinic with the local team would help to increase awareness of concerns and expectations and highlight any specialist assessments or interventions required.

Other ways to improve communication and appropriate referrals and liaison would be for the spina bifida team to provide regular education sessions about spina bifida specific complications and the services available within TSCUH to increase awareness.

Communication could be vastly improved if there were some form of central resource or shared platform/library of information for both children and families and for professionals locally. This information could be provided electronically, through a website or shared drive or physically within hospital or HSE library services. Similarly having a shared resource or platform for sharing information or resources would be of great benefit to service providers and families. A fully-staffed MDT spina bifida team within the national centre is required in order to advance many of the aforementioned suggestions.

## **Recommendation**

1. Communication between specialist services and local teams requires development with methods such as pre and post clinic liaison, development of patient passport, identification of a point of contact and co-ordination, educational sessions and a shared library of resources.

## ***Transition***

### **Background**

Turner (2007) described transition as “a psychological response to change” which is a significant life event that requires restructuring your view of yourself and the world. Children with spina bifida experience various transitions throughout their childhood including the transition from pre-school into primary school, primary school into secondary school and the transition from adolescence into adulthood and adult services.

When discussing the Life Needs Model of Paediatric Service Delivery, King et al. (2002) identified the importance of planning for significant life events such as all of these transitions as a process rather than a one-off event (King et al., 2006). In order to do this, families need to be supported throughout the process of each transition including the transition from one service to another during their childhood (e.g. transition from Early Intervention Team to School Age Team).

Children with spina bifida, are now surviving longer and living well into adulthood despite some health care systems still focusing on spina bifida as a childhood condition (Viner, 2001). Appropriate transition management has been seen as an essential part of best practice in any paediatric clinic (Viner, 2001). However Sawyer and Macnee (2010) have discussed a wealth of empirical evidence regarding barriers to the transition to adult services, and how services such as those for spina bifida have lagged behind with regards transition planning and service developments for adults.

Many children with spina bifida are managed in specialist multidisciplinary Spina Bifida clinics. However once they become adults they lose contact with these specialist treatment centres. Therefore the need for adult specialist Spina Bifida clinics has been highlighted and should be identified by the paediatric services (Crosthwaite et al., 2001).

The transition to adult services should involve a clear programme which is guided by education to help children become responsible for their own health care rather than the often haphazard and idiosyncratic way that the transfer to adult services can occur (Viner, 2001). Results from a study investigating mother–child discrepancies over perceptions of who is responsible for spina bifida medical tasks highlighted the need for parental involvement for optimal adherence during preadolescence and early adolescence, Psihogios and Holmbeck (2013) suggesting parents need to be involved in the education process to assist their children take control of their own care.

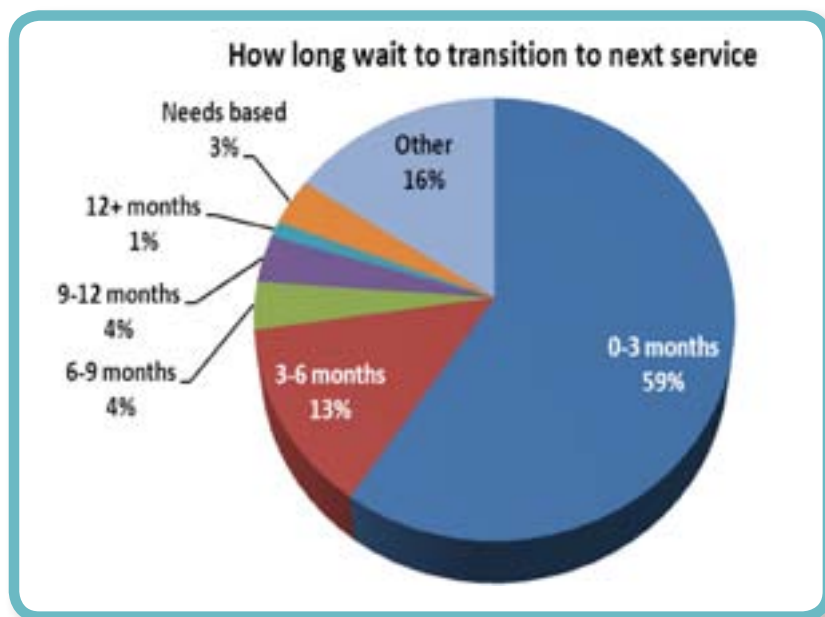
In Northern Ireland results of a survey of patients with spina bifida and hydrocephalus transitioning to adult services indicated a changing perception of need with more of a focus on employment (McGonnell, 2009). Therefore adult specialists who are knowledgeable about the specific spina bifida issues that might present in adulthood are necessary. Good communication between paediatric and adult services is essential, as when this is not evident it can result in a barrier to transition (Wright et al., 2009). There is much debate in the literature about when the process of transition into adulthood should commence. This could be as early as 12 or 13 years; however no right time has been identified - instead the need for flexibility, preparation and developmental readiness have been highlighted as key to a successful transition (Viner, 2001). Merkens (2006) guidelines recommend that adolescents with spina bifida who attend special education should commence transition planning at age 14 in order to plan for vocational and occupational domains.

Adult spina bifida services require organisation, management and support to ensure they are specialist enough to meet the complex needs of this population and also to ensure appropriate access to professionals within the multidisciplinary team (Sawyer and Macnee, 2010). Overall Turner (2007) found there is still a lot of work to be done in studying how adolescents' transition between life circumstances suggesting that more research is required to measure the transition practices and programmes in the area of spina bifida.

## Results

In relation to transitioning between services in childhood, service providers were asked how long children have to wait to transition to the next service, for example from Early Intervention Team to School Age Team. 60% of service providers (Figure 95) report that children transition from their service to the next within 0-3 months with some service providers commenting that in their service the transition was an “automatically transfer” or a “seamless transition”.

Figure 104



However 13% (figure 122) of service providers who answered this question reported that it can take between 3 and 6 months to transition to the next service which was reported to be mainly dependent on the length of waiting list in the next service or staff availability. So if the School Age Team had a lengthy waiting list or a therapist out on sick leave this would impact the child's transition from EIT into the SAT:

*“E.I.T transitions child to school and transfers to school age team after the first term. Due to staffing levels and large caseloads on school age team there may be a wait of 6 months plus for any intervention” (Service Provider)*

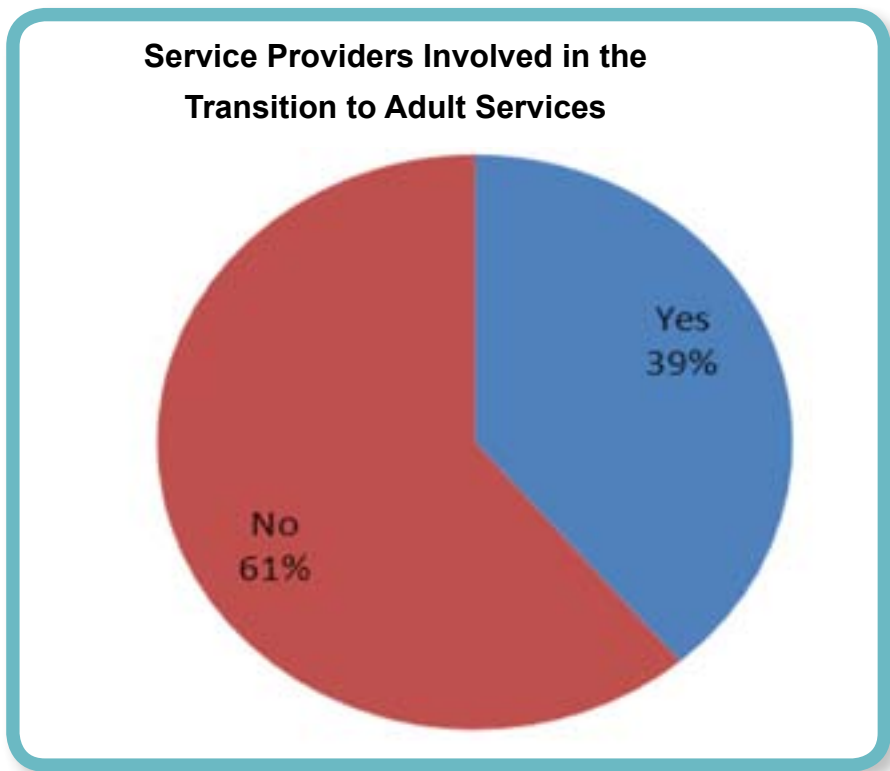
*“Transition speed dependant on the staff and service available to discharge to (if any!)” (Service Provider)*

Worryingly, many service providers reported that they were “unsure” or the wait times for transition into the next chronological service was “not known”.

Only 39% of service providers reported that they are involved in the transition of children into adult services (Figure 96). Reasons this number is so low is unclear but may be representative of the fact the majority of the sample who responded work in Early Intervention Teams meaning children transfer first to School Age Teams before adult services.

For service providers who reported that they are involved in the transition of children from their service into adulthood, qualitative comments received suggest that while a variation of interventions are in place locally no one system or method is used to support children with spina bifida transition from adolescence into adulthood. Most comments related to the logistics and administrative tasks related to transferring a child to adult services, rather than preparing the child themselves for the transition to adult services for example:

Figure 105



*“I currently complete discharge reports and liaise with new team members”  
(Service Provider)*

*“Most effort goes into accurate assessment at the time of transition”  
(Service Provider)*

*“Liaise with the appropriate therapists in adult services to provide a  
smooth transition” (Service Provider)*

*“Handover report completed and on occasion an introduction with the family  
to the new therapist” (Service Provider)*

However some service providers appeared to be completing transition planning and education with families with examples including:

*“An information evening for young people and parents on options and services in the area. A transition planning meeting with family and young person. These could be developed further by starting earlier with families e.g. at age 15/16 and preparing and assisting in looking at options”*  
(Service Provider)

*“Input begins as the child completes Junior Cert/Transition year. Liaison with parents and child to establish plan for transition from school to college and /or employment. Emphasis on domestic ADLs to establish ability for independent living and supports required. Liaison with OT in adult primary care/community services and scheduling of most appropriate time for handover of case management”*  
(Service Provider)

*“A transition plan is implemented and gone through with parents and aids goals in the transition process. Our aim in transitioning is to contribute to a better understanding of the challenges of transition to achieve self-management and social development for young adults with spina bifida.”*  
(Service Provider)

Parents were asked about their concerns for the future and how they felt about their child becoming an adult. They understandably highlighted various concerns about their child's needs and outcomes in the areas of mobility, continence, education, employment, accessing adult services, funding of equipment, gaining weight and appropriate housing. However they appeared to have difficulty with planning for these future needs with quotes including:

*“I don't look too far into the future and that's been honest with you, because I'm inclined to take things day by day”* (Parent / Guardian)

*“I kind of haven't thought about adult services, it's too far down the road”*  
(Parent / Guardian)

*“I don't look that far ahead to be honest with you. I don't know if I'd be able to ... you know it's a kind of coping mechanism for me to deal with things as I have them, and not to look too far ahead”* (Parent / Guardian)



Interestingly however during the child focus groups while some positive views were provided about getting older and becoming an adult due to increased independence anxieties were also raised regarding coping as independent adults:

*“Ok, I am so nervous about getting married, I’m like ok if I don’t know how to like plan kind of my future then I will probably get stressed out and I’ll probably end up calling my Mum over to help me out”(Focus Group)*

From the parents’ perspective limited information regarding transition to adult services is available to families and professionals, resulting in feelings of dread due to the perceived loss of expertise and increased waitlists for review. Parents and service providers both commented on anxieties and confusion and the need for further transition planning development.

*“I am kind of dreading the day that he is completely finished in (Children’s Hospital)” (Parent / Guardian)*

*“I am worried there is little available for 18 plus” (Parent / Guardian)*

*We are now in the transition period of transferring to adult services and do hope that the service is adequate and reassuring. (Parent / Guardian)*

*“In our area there is no adult disability service for children with spina bifida. They will be seen in the Primary Care setting. There is no transition process, only to make sure that equipment is currently in place” (Service Provider)*

*“There is no clear procedure re transition to adult services (e.g. where to refer 16 plus for an orthopaedic consultation?)” (Service Provider)*

*“I think 18 plus should have a better access to routine checks with a Neurosurgeon, Urologist, Orthopaedic Consultant” (Service Provider)*

*“When the child reaches age of 16/18 and is discharged...I have difficulties, I don’t know where to refer them. But there is no procedure in place” (Service Provider)*

Some suggestions were offered by service providers about how transition services might be improved in the future including:

*“Could be improved by improving communication/linking between child & adult services. Having a way to inform adult/community service of pending referrals and then following up after discharge regarding therapy/equipment/etc” (Service Provider)*

*“Transition to adulthood should be a flowing process from the start with functional goals set around independence, social skills confidence and self-esteem. It requires parents being educated in that mindset and encouraged to let their child take risks (age appropriate) and experience normality as much as possible. Over protection is very common and parents admit all the time to falling into that trap” (Service Provider)*

## **Discussion**

Children with spina bifida experience various transitions throughout their life, and they require adequate support for both local and specialist services to manage these challenging and stressful times. Results suggest that transition planning between services and from paediatric to adult services at both the tertiary (specialist) level and primary (local) level are inadequate and do not sufficiently support the child and families.

While some service providers report a seamless transition from one service to the next most commented on confusion or uncertainty about where children in their service go to after they leave their service. Some service providers are involved in transition planning although this appears to be mainly administrative, and only a few report being involved in family and child preparation and education for the transition to adulthood and adult services. Viner (2001) recommends that transition programmes should be an educational and therapeutic process rather than solely administrative in nature. Access to transition planning also appears to be inequitable and based on geographic location and the type of service attended based on the location of responses received.

Anxiety, confusion and apprehension were reported by parents and service providers who were unsure about what to expect from adult services and unsure of what would be available to meet the needs of children with spina bifida. Scal (2002) similarly found that “fear and ambivalence” can characterise health care transitions for people with chronic conditions. It is therefore vital to ensure that healthcare transitions particularly the transition from paediatric to adult services is appropriately supported.

## Recommendations

1. Transition planning for children as they negotiate through major life transition (into school, into adulthood) is urgently required.
2. Specific and directed transition planning should commence early in adolescence, (between 12 and 14 years) and should include education and preparation provided to the child/adolescent regarding practical skills and self management required to manage health and therapy needs in adulthood. The multidisciplinary spina bifida service should be involved in commencing the transition planning process and should liaise with local services about appropriate transition planning and programmes which could support adolescents locally.
3. It is essential that appropriate adult services are identified at an early stage and that the coordination of care within adult services is improved for example through the formation of an adult spina bifida service.

# **CHAPTER SIX**

## **RECOMMENDATIONS FOR THE FUTURE**

## RECOMMENDATIONS FOR THE FUTURE

PHYSICAL IMPACT	
Neurosurgical	
<ol style="list-style-type: none"> <li>1. Directed education regarding future pregnancy risk to extended families of children born with neural tube defects should be implemented by the national spina bifida service in Temple Street</li> <li>2. Parents should receive education about their child's level of lesion to help with understanding the potential impact of this.</li> </ol>	
Mobility	
<ol style="list-style-type: none"> <li>1. It is recommended that annual muscle charting be performed by an adequately trained Physiotherapist to monitor muscle function and document change. Communication between local and specialist Physiotherapists is essential to determine functional ability and results should be discussed with parents.</li> </ol>	
Orthopaedic	
<ol style="list-style-type: none"> <li>1. Frequent orthopaedic review is required for children with spina bifida throughout their lifespan.</li> <li>2. Orthopaedic surgery should only be completed when warranted and for functional gain</li> </ol>	
Equipment	
<ol style="list-style-type: none"> <li>1. Timely availability of a variety of generic and specialised equipment is required through local services to meet the needs of these children.</li> <li>2. Experienced staff who are most involved in providing equipment, namely Occupational Therapists, Physiotherapists, Orthotists and Nurses require adequate training.</li> </ol>	
Bladder and Bowel	
<ol style="list-style-type: none"> <li>1. Appropriate interventions, implemented earlier in the child's life, need to be in place to support difficulties, protect renal function and to assist with achieving social continence.</li> </ol>	

## **Growth & Nutrition**

1. It is recommended that height, weight and BMI (Centiles and Z score) of these children are measured at a minimum every 6 months by local professionals most frequently involved in the care of the child.
2. A multidisciplinary approach to prevention and management of increased weight is required specifically for children with spina bifida due to their increased risk. Parents, families and professionals should be educated about specific nutritional difficulties of children with spina bifida, be given advice on healthy eating, active living and promoting a positive self image. This should be discussed at annual review at a spina bifida MDT Clinic.
3. Children with spina bifida who are overweight/obese should have equitable access nationally to a dietitian for assessment and intervention

## **Visual Difficulties**

1. A comprehensive functional visual assessment should be available to all children with spina bifida who present with visual difficulties.
2. This could occur in conjunction with the TSCUH MDT spina bifida service but would require extra resources and staff in the area of ophthalmology, occupational therapy and neuropsychology.

## **Speech and Language Difficulties**

1. Access to a Speech and Language Therapist should be should be available to all children with spina bifida for assessment of language skills and appropriate interventions as required.

## **Tissue Viability**

1. It is the responsibility of all health care professionals to be vigilant of pressure areas and to act on concerns when raised.
2. Children and parents require education regarding the risks of pressure sores and appropriate equipment needs to be in place to prevent the development of pressure areas.
3. Those at high risk of pressure sores, particularly those who are immobile, should have access to pressure mapping assessments regionally to determine appropriate preventative measures which need to be provided (i.e. pressure cushions or supportive equipment).

## PSYCHOSOCIAL IMPACT

### Cognition and Schooling

1. Mainstream education, along with necessary supports, should be encouraged for children with spina bifida, when this is appropriate to their learning needs. Access to mainstreams school should not be influenced by physical or continence needs.
2. Supports are required to assist with physical or continence needs might include SNAs, classroom assistants, resource teaching or school nursing.
3. Improvements in physical accessibility of schools and communities are necessary to increase independence and active participation in daily life.

### Psychosocial Presentation

1. All team members should be aware of and responsive to the child's psychosocial needs and there is a need for on going psychological interventions when cognitive or psychosocial issues present.
2. Exposure of children to peer support activities for example extra curricular activities, sports or social events, is essential.
3. Children with spina bifida should have frequent monitoring of their cognitive and psychosocial needs with access to a cognitive assessment and interventions to support their abilities when necessary.

## FAMILY IMPACT

1. There is a need for increased public awareness about spina bifida, pre-conceptional care, folic acid fortification and supplementation in Ireland to reduce anxiety on receiving a diagnosis of spina bifida and to support the families and individuals to be seen as part of their community.
2. Parents and families require earlier support, practical and emotional assistance following the antenatal diagnosis of spina bifida.
3. On-going family, peer and sibling support should be available to all families wishing to avail of this
4. Improved coordination of care and communication is required to reduce parental burden.

## LOCAL SERVICES

### Team Configuration

1. Local multidisciplinary teams (MDTs) should be available for children with spina bifida across the lifespan and they should consist of at least physiotherapy, occupational therapy, speech and language therapy, psychology, social work, and paediatrics. Improved access to speech and language therapy, social work, family support workers, nursing, orthotists, psychology and dietitian is required.
2. Education and support for local MDT professionals should be readily accessible from the national specialist centre when required.

### Access and Frequency of Review

1. Improved practical, social and emotional support is required immediately post discharge from tertiary hospitals for families. This could be provided by social workers, nursing professionals, special needs counsellors or by spina bifida family support workers
2. Equitable frequency of access to therapy professionals is required based on the child and family's needs.

### Occupational Therapy and Physiotherapy

1. Increased frequency of provision of occupational therapy and physiotherapy services is recommended based on therapists' assessments and recommendations and also the child's needs rather than prescriptive blocks of therapy or pre-determined time frames throughout the year.
2. Any private therapy if sought or obtained, should work in collaboration and communication with the primary teams with parental consent in order to clarify expectations and goals.
3. The increased use of hydrotherapy is recommended as a therapeutic intervention for children with spina bifida to assist with joint flexibility, aerobic exercise and pressure relief



## Equipment

1. It is recommended that initial seating and wheelchair assessments should occur earlier than therapists' report this is currently happening.
2. It is recommended that there should be a reduction in inefficiencies of funding equipment throughout the country to ensure essential equipment is provided in a timely manner.
3. Increasing the funding for aids and appliances throughout the HSE regions could also occur or consider reallocation of funding from savings made by reduced wastage and increased recycling.

## SPECIALIST SERVICE

### Antenatal and Neonatal

1. Services should strive to provide an early antenatal diagnosis for all NTDs in Ireland.
2. Antenatal counselling by members of the multidisciplinary spina bifida team which includes discussion of management options and potential outcomes should continue to be available to any parent who desires this.
3. The full specialist multidisciplinary team should be available at the initial neonatal stage to support children and families during this complex time. At present there are no resources to provide a paediatrician review for neonatal inpatients with spina bifida after their surgical repair. To promote continuity of care and specialist input, involvement of the specialist paediatrician at this stage is recommended.
4. Consideration should be given to parental anxiety and distress following the separation at birth for transfer to the neonatal service.

### Neurosurgery

1. All children with spina bifida should have access to a paediatric trained Neurosurgeon for emergency advice and support.
2. A paediatric Neurosurgeon should be available to review children with spina bifida throughout the lifespan annually as part of the multidisciplinary team clinic review.

## **Urology**

1. All children should have access to a Urologist with a specialist interest in spina bifida to ensure early surveillance and management to support their bladder and bowel needs.
2. Trained nursing professionals should be available outside the national centre to assist with support and training of families and children with regards to achieving social continence. This support could be provided through up-skilling of existing continence nurses, or training public health nurses or early intervention nurses to assist with these interventions.

## **Orthopaedics**

1. A dedicated paediatric disability orthopaedic surgeon to review the whole child and be part of the MDT Spina Bifida clinic is required.
2. Increased access to theatre space and resources are also required to reduce lengthy waiting times to access surgery.

## **Spina Bifida Nurse Specialist**

1. A Spina Bifida Nurse Specialist should be available for all children and families to access to assist with coordination of care, specialist education and interventions.

## **Multidisciplinary Spina Bifida Service**

1. A fully-staffed multidisciplinary spina bifida service should be available for inpatient and outpatient care with access to Neurosurgeon, Urologist, Orthopaedic Surgeon, Spina Bifida Nurse Specialist, Paediatrician, Physiotherapist, Occupational Therapist, Social Worker, Neuropsychologist, Nurse Specialists, Speech and Language Therapist, Dietitian, Neuro-ophthalmologist, Orthoptist, Play Therapist as well as access to imaging services, Radiology and administration support.
2. Children should be reviewed at the MDT Spina Bifida Clinic on at least an annual basis.
3. A fully-staffed multidisciplinary Spina Bifida clinic is required which should consist of at least the following seven key professionals: Neurosurgeon, Urologist, Orthopaedic Surgeon, spina bifida Nurse Specialist, Paediatrician, Physiotherapist and Occupational Therapist.
4. The specialist spina bifida team is responsible for education and support of local teams.

## **Communication**

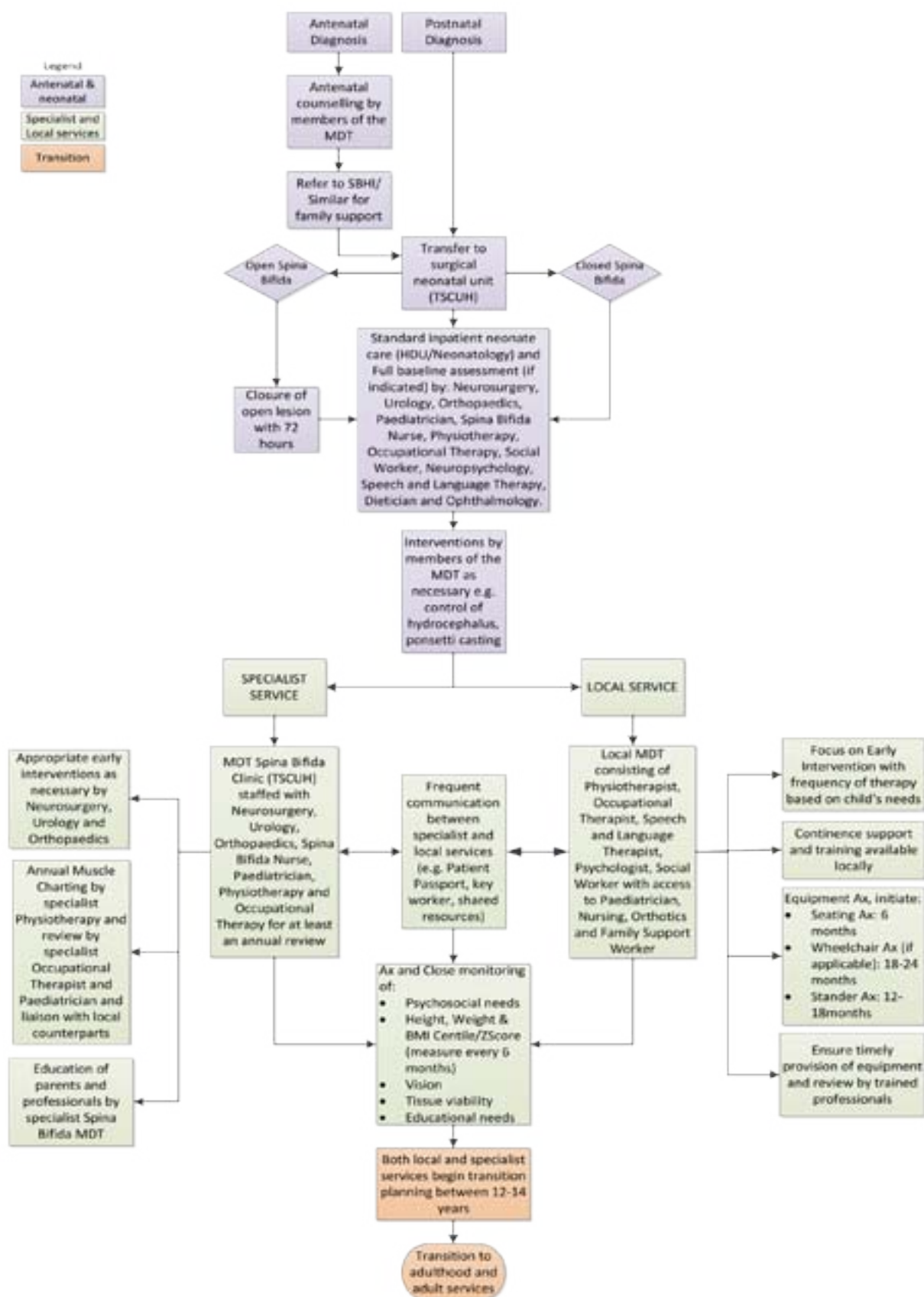
1. Communication between specialist services and local teams requires development with methods such as pre and post clinic liaison, development of patient passport, identification of a point of contact and co-ordination, educational sessions and a shared library of resources.

## **Transition**

1. Transition planning for children as they negotiate through major life transition (into school, into adulthood) is urgently required.
2. Specific and directed transition planning should commence early in adolescence, (between 12 and 14 years) and should include education and preparation provided to the child/adolescent regarding practical skills and self-management required to manage health and therapy needs in adulthood. The multidisciplinary spina bifida service should be involved in commencing the transition planning process and should liaise with local services about appropriate transition planning and programmes which could support adolescents locally.
3. It is essential that appropriate adult services are identified at an early stage and that the coordination of care within adult services is improved for example through the formation of an adult spina bifida service.

# APPENDIX 1

## National Care Pathway Based on Research Recommendations



# APPENDIX 2

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

### 4. What is your child's race/ethnicity or cultural background?

Tick all that apply

☐ White☐ Asian☐ Black / African or African American☐ Eastern European☐ Hispanic/Latino☐ Travelling Community☐ Other

Other (please specify)

### 5. please indicate below the number of children in your family, and where in the birth order your child with Spina Bifida lies.

Number of children in your family

Where in the birth order does your child with Spina Bifida lie (eg. 2nd child = 2)

### 6. How many parents/guardians live in the house with your child

☐ 1☐ 2

Other (please specify)

### 7. What is your families' employment status?

Please tick one per parent / guardian

	Parent / Guardian 1	Parent / Guardian 2
Full time employment	<input type="radio"/>	<input type="radio"/>
Part time employment	<input type="radio"/>	<input type="radio"/>
Not employed, looking for work	<input type="radio"/>	<input type="radio"/>
Not employed, not looking for work	<input type="radio"/>	<input type="radio"/>
Stay at home parent / guardian	<input type="radio"/>	<input type="radio"/>
Unable to work	<input type="radio"/>	<input type="radio"/>
Retired	<input type="radio"/>	<input type="radio"/>
Not applicable	<input type="radio"/>	<input type="radio"/>

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

### 8. Has your employment status changed as a result of your child with Spina Bifida?

- ☐ Yes  
☐ No

If Yes, please detail

### \*9. Which of the following areas do you currently live in?

Tick one

- |  |                                       |
|--|---------------------------------------|
| <input type="radio"/> Carlow               | <input type="radio"/> Laois           |
| <input type="radio"/> Cavan                | <input type="radio"/> Leitrim         |
| <input type="radio"/> Clare                | <input type="radio"/> Limerick        |
| <input type="radio"/> Cork South Lee       | <input type="radio"/> East Limerick   |
| <input type="radio"/> Cork North Lee       | <input type="radio"/> Longford        |
| <input type="radio"/> West Cork            | <input type="radio"/> Louth           |
| <input type="radio"/> North Cork           | <input type="radio"/> Mayo            |
| <input type="radio"/> Donegal              | <input type="radio"/> Meath           |
| <input type="radio"/> Dublin South         | <input type="radio"/> Monaghan        |
| <input type="radio"/> Dublin South East    | <input type="radio"/> Offaly          |
| <input type="radio"/> Dublin South City    | <input type="radio"/> Roscommon       |
| <input type="radio"/> Dublin South West    | <input type="radio"/> Sligo           |
| <input type="radio"/> Dublin West          | <input type="radio"/> North Tipperary |
| <input type="radio"/> North West Dublin    | <input type="radio"/> South Tipperary |
| <input type="radio"/> Dublin North Central | <input type="radio"/> Waterford       |
| <input type="radio"/> North Dublin         | <input type="radio"/> Westmeath       |
| <input type="radio"/> Galway               | <input type="radio"/> Wexford         |
| <input type="radio"/> Kerry                | <input type="radio"/> Wicklow         |
| <input type="radio"/> Kildare              | <input type="radio"/> West Wicklow    |
| <input type="radio"/> Kilkenny             | <input type="radio"/> Other           |

Other (please specify)



## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

### 10. Which type of educational setting does your child attend?

Tick one answer

- |  |  |
|--|--|
| <input type="radio"/> Too young as yet                 | <input type="radio"/> Mainstream secondary school        |
| <input type="radio"/> Mainstream Preschool             | <input type="radio"/> Special education secondary school |
| <input type="radio"/> Special education preschool      | <input type="radio"/> Home School                        |
| <input type="radio"/> Mainstream primary School        | <input type="radio"/> Other                              |
| <input type="radio"/> Special education primary school |  |

Other (please specify)

## 2. Medical Information

This section gathers information about family history of Neural Tube Defects, the level of lesion and details of surgeries.

### 11. Is there a family history of Neural Tube Defects or other Disabilities in your child's family?

- ☐ No
- ☐ Yes (detail below)

If Yes, please detail

### \*12. What is the level of lesion of your child/adolescents Spina Bifida (if known)?

- ☐ Cervical Region (C1-C7)
- ☐ Thoracic Region (T1-T12)
- ☐ Thoraco-Lumbar
- ☐ Lumbar Region (L1-L5)
- ☐ Lumbo-Sacral
- ☐ Sacral Region (S1-S5)
- ☐ Unsure
- ☐ Other

Other (please specify)

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

### 13. Where was your child's surgical closure (lesion repair) completed?

Tick one

☐ Temple Street Children's University Hospital

☐ Cork University Hospital

☐ Our Lady's Children's Hospital Crumlin

☐ Abroad (please detail where in the box below)

☐ Beaumont Hospital

☐ Other (please detail)

Abroad or Other (please detail)

### \*14. Does your child have a history of Hydrocephalus?

☐ Yes

☐ No

☐ Unsure

Comment

### \*15. If your child/adolescent has a shunt in place, when was this first inserted?

Tick one

☐ In the first week of life

☐ Shunt was removed and not replaced

☐ In the first 6 months of life

☐ No shunt in place

☐ In the first 2 years of life

☐ Other

☐ After 2 years

Other (please specify)

### \*16. How many shunt related surgeries has your child/adolescent had?

Tick one that applies

☐ None

☐ 3

☐ 1

☐ 4+

☐ 2

☐ other

Other (please specify)

## 2. Medical Information - Physical Implications

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

This section details the physical implication of Spina Bifida including functional ability, weight, height, pressure sores, bladder/bowel needs, orthopaedic issues, learning and self-esteem.

### 17. Which of the following statements best describes your child's level of mobility?

Tick one

- |  |   |
|--|---|
| <input type="radio"/> Too young to tell mobility level yet                         | <input type="radio"/> Walks without difficulty (with splints/aids)    |
| <input type="radio"/> Uses a wheelchair most of the time                           | <input type="radio"/> Walks without difficulty (without splints/aids) |
| <input type="radio"/> Uses splints and walking aid sometimes, wheelchair sometimes | <input type="radio"/> Other   |
| <input type="radio"/> Uses splints and sometimes needs help                        |   |

Other (please explain)

### \*18. What supportive equipment has your child received, or required to date?

Select one option for each piece of equipment

	Received	Awaiting	Required	Not Required at present
Walker / Crutches / Sticks	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthoses (AFO's, RGO's)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Resting Splints (eg. Boots and Bars)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hand or arm splints	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Stander	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Activity chair	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Transfer board	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Shower/Bath chair	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Sleep system	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Specialised bed / mattress	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hoist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Manual Wheelchair	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Powered Wheelchair	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please name)

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

**19. How long did it take for the following equipment to arrive once it was recommended by a therapist/staff member?**

**Select wait time for each piece of equipment, or N/A if equipment is not in place**

	1-4 weeks	1-3 months	3-6 months	6 months to 1 year	1 year +	N/A
Walker / Crutches / Sticks	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthoses (AFO's, RGO's)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Resting Splints (eg. Boots and Bars)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hand or arm splints	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Stander	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Activity chair	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Transfer board	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Shower/Bath chair	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Sleep system	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Specialised bed / mattress	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hoist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Manual Wheelchair	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Powered Wheelchair	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please name)

## 2. Medical Information - Height and Weight

**20. Please detail your child's weight (in pounds and ounces or kg) and height (in feet and inches or cm) below.**

Child's Weight

Child's Height

**21. How frequently are the following measurements taken for your child?**

	More than once per month	Every 1 - 3 months	Every 3 - 6 months	Every 6 months - 1 year	Annually	Less than once a year	Other
Height	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Weight	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Head Circumference	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please specify)

## 2. Medical Information - Pressure Sores

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

### 22. Does your child have a history of pressure sores

- ☐ Yes GO TO QUESTION 23
- ☐ No GO TO QUESTION 24

## 2. Medical Information - Pressure Sores

### 23. Have any of the following measures been put in place to reduce the risk of pressure sores?

- |   |  |
|---|--|
| <input type="checkbox"/> Pressure relieving wheelchair cushion        | <input type="checkbox"/> Diet advice             |
| <input type="checkbox"/> Positioning aids                             | <input type="checkbox"/> Regular skin inspection |
| <input type="checkbox"/> Air alternating mattress                     | <input type="checkbox"/> Other                   |
| <input type="checkbox"/> Frequent turning/re-positioning in chair/bed |  |

Other (please specify)

## 2. Medical Information - Visual Difficulties

### 24. Does your child have any of the following visual difficulties?

- |  |  |
|--|--|
| <input type="checkbox"/> Difficulty copying off the whiteboard                   | <input type="checkbox"/> Difficulties with jigsaws and/or staying inside the lines |
| <input type="checkbox"/> Difficulty with visual tracking (e.g. following a ball) | <input type="checkbox"/> Difficulty noticing details (e.g. in pictures)            |
| <input type="checkbox"/> Gazing at lights  | <input type="checkbox"/> Fatiguing easily from visual tasks                        |
| <input type="checkbox"/> Avoiding light (Photophobia)                            | <input type="checkbox"/> Other   |
| <input type="checkbox"/> Loss of visual range (visual field loss)                | <input type="checkbox"/> None of the above   |
| <input type="checkbox"/> Difficulties with hand eye-coordination                 |  |

Whom have you spoken to about these problems?

## 2. Medical information - Bladder and Bowel



## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

### 25. Which of the following health care professional are involved in addressing bladder and bowel continence needs of your child?

Tick all that apply

- |   |   |
|---|---|
| <input type="checkbox"/> Urologist              | <input type="checkbox"/> Public Health Nurse    |
| <input type="checkbox"/> Nurse Specialist       | <input type="checkbox"/> Company Representative |
| <input type="checkbox"/> Occupational Therapist | <input type="checkbox"/> Other                  |
| <input type="checkbox"/> Family Support Worker  | <input type="checkbox"/> Not required           |
| <input type="checkbox"/> Paediatrician          |   |

Other (please specify)

### 26. Which of the following measures have been put in place to support your child's urinary and/or bowel continence needs?

Tick all that apply

- |  |  |
|--|--|
| <input type="checkbox"/> Catheterisation                         | <input type="checkbox"/> Surgical intervention     |
| <input type="checkbox"/> Prophylactic antibiotics (preventative) | <input type="checkbox"/> Anal plugs                |
| <input type="checkbox"/> Nappies or Continence Pads              | <input type="checkbox"/> Botulinum toxin A (botox) |
| <input type="checkbox"/> Medication (laxative or enemas)         | <input type="checkbox"/> Other                     |

Other (please detail)

## 2. Medical Information - Orthopaedic complications

### 27. Has your child had any of the following orthopaedic complications?

Tick all that apply and add details below

- |  |  |
|--|--|
| <input type="checkbox"/> Fractures                 | <input type="checkbox"/> Scoliosis               |
| <input type="checkbox"/> Hip dislocation(s)        | <input type="checkbox"/> Kyphosis                |
| <input type="checkbox"/> Foot or ankle Deformities | <input type="checkbox"/> Lordosis/ Hyperlordosis |
| <input type="checkbox"/> Contractures              | <input type="checkbox"/> Other                   |

Other (please specify)

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

**28. Has your child had orthopaedic surgery in any of the following areas?**

**Tick all that apply and add details below**

☐

Arm

☐

Foot/ankle

☐

Hip

☐

Spine

☐

Leg

☐

Other

please specify surgery

### 2. Medical Information - Cognitive Assessment

**29. Has your child had his/her learning (cognitive ability) formally assessed?**

☐

Yes GO TO QUESTION 30

☐

No GO TO QUESTION 31

### 2. Medical Information - Cognitive assessment

**30. Which range did the most recent cognitive assessment place your child in?**

**Tick one and GO TO QUESTION 32**

☐

Average Intellectual ability

☐

Moderate Intellectual Disability

☐

Low Average Intellectual Ability

☐

Severe Intellectual Disability

☐

Borderline Intellectual ability

☐

Above average Intelligence

☐

Mild Intellectual Disability

☐

Other

Other (please specify)

### 2. Medical Information - Cognitive assessment

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

### 31. Why has your child not had his / her learning (cognitive ability) assessed?

- ☐ Not required / requested to date
- ☐ On waiting list
- ☐ Not available
- ☐ Other

Comments

## 2. Medical Information - Self-esteem

### 32. Do you feel that Spina Bifida has negatively impacted on your child's self-esteem?

- ☐ Yes
- ☐ No

If yes, please detail how or why

## 3. Therapy Information

This section gathers information about the type of therapeutic interventions that your child/adolescent receives with a particular focus of physiotherapy and occupational therapy interventions. It looks at therapy frequency, duration, quality and outcomes



## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

**\*33. In the last year, how frequently has your child seen the following professionals?**

**Tick one option per professional**

	Never seen	More than once per week	Weekly	Every second week	Monthly	Every 3 months	Every 6 months	Once this year	Not seen this year	Other
Community Paediatrician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hospital Paediatrician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Neurosurgeon	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Urologist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthopaedic Surgeon	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Spina Bifida Nurse	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Nurse Specialist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Public Health Nurse	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthotist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Psychologist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Speech and Language Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Dietician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
HSE Family Support Worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
SBHI family support worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Social Worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please specify)

**\*34. Does your child/adolescent have access to a paediatric occupational therapist?**

- ☐ Yes GO TO QUESTION 35
- ☐ No GO TO QUESTION 40

### 3. Therapy Information - Paediatric Occupational Therapy

## 35. Which service does your occupational therapist(s) (OT) work in?

If you have more than one OT, tick all that apply

- |   |  |
|---|--|
| <input type="checkbox"/> Enable Ireland                               | <input type="checkbox"/> Our Lady's Children's Hospital Crumlin        |
| <input type="checkbox"/> Central Remedial Clinic                      | <input type="checkbox"/> Other charitable service (e.g. SAOIRSE, COPE) |
| <input type="checkbox"/> Health Service Executive                     | <input type="checkbox"/> Private                                       |
| <input type="checkbox"/> Temple Street Children's University Hospital | <input type="checkbox"/> Other   |

Other (please specify)

## 36. How often (on average) do you see your occupational therapist(s)?

Tick one

- |   |   |
|---|---|
| <input type="radio"/> More than once per week | <input type="radio"/> Every 6 months        |
| <input type="radio"/> Weekly                  | <input type="radio"/> Every 9 months        |
| <input type="radio"/> Every second week       | <input type="radio"/> Annually              |
| <input type="radio"/> Monthly                 | <input type="radio"/> Less than once a year |
| <input type="radio"/> Every 3 months          | <input type="radio"/> Other                 |

Other (please specify)

## \*37. Has your occupational therapist(s) completed any of the following interventions with your child?

Tick all that apply

- |  |   |
|--|---|
| <input type="checkbox"/> Wheelchair and/or seating assessment                  | <input type="checkbox"/> Gross motor development                        |
| <input type="checkbox"/> Splinting (upper or lower limb)                       | <input type="checkbox"/> Self care (eg. washing / dressing / toileting) |
| <input type="checkbox"/> Pressure relief advice (positioning and/or equipment) | <input type="checkbox"/> Feeding skills                                 |
| <input type="checkbox"/> Position and/or posture advice                        | <input type="checkbox"/> Family Education                               |
| <input type="checkbox"/> Home and/or school visits                             | <input type="checkbox"/> Provision of equipment                         |
| <input type="checkbox"/> Fine Motor development                                | <input type="checkbox"/> Other  |

Other (please specify)

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

**38. How satisfied are you with the AVAILABILITY of your current occupational therapy service(s)?**

**Tick one**

- ☐ Extremely satisfied
- ☐ Satisfied
- ☐ Somewhat satisfied
- ☐ Not Satisfied

Comment

**39. How satisfied are you with the QUALITY of your current occupational therapy service(s)?**

**Tick one and GO TO QUESTION 41**

- ☐ Extremely satisfied
- ☐ Satisfied
- ☐ Somewhat satisfied
- ☐ Not satisfied

Comment

### 3. Therapy Information - Paediatric Occupational Therapy

**\*40. Why has your child not attended a paediatric Occupational Therapist?**

**Tick one**

- ☐ Not required / requested to date
- ☐ On waiting list
- ☐ Not available

Comment

### 3. Therapy Information - Paediatric Physiotherapy

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

### 41. Does your child/adolescent have access to a paediatric physiotherapist?

- ☐ Yes GO TO QUESTION 42
- ☐ No GO TO QUESTION 47

## 3. Therapy Information - Paediatric Physiotherapy

### 42. Which type of service does your physiotherapist(s) (PT) work in?

If more than one PT, tick all that apply

- |   |  |
|---|--|
| <input type="checkbox"/> Enable Ireland                               | <input type="checkbox"/> Our Lady's Children's Hospital Crumlin        |
| <input type="checkbox"/> Central Remedial Clinic                      | <input type="checkbox"/> Other charitable service (e.g. SAOIRSE, COPE) |
| <input type="checkbox"/> Health Service Executive                     | <input type="checkbox"/> Private                                       |
| <input type="checkbox"/> Temple Street Children's University Hospital | <input type="checkbox"/> Other   |

Other (please specify)

### 43. How often (on average) do you see your physiotherapist(s)?

Tick one

- |   |  |
|---|--|
| <input type="radio"/> More than once per week | <input type="radio"/> Every 6 months     |
| <input type="radio"/> Weekly                  | <input type="radio"/> Every 9 months     |
| <input type="radio"/> Every second week       | <input type="radio"/> Annually           |
| <input type="radio"/> Monthly                 | <input type="radio"/> Less than annually |
| <input type="radio"/> Every 3 months          | <input type="radio"/> Other              |

Other (please specify)

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

**\*44. Has your physiotherapist(s) completed any of the following with your child?**

**Tick all that apply**

- |  |  |
|--|--|
| <input type="checkbox"/> Mobility aid assessment or prescription | <input type="checkbox"/> Fine Motor development  |
| <input type="checkbox"/> Mobility training (walking)             | <input type="checkbox"/> Gross motor development |
| <input type="checkbox"/> Splinting (upper or lower limb)         | <input type="checkbox"/> Family Education        |
| <input type="checkbox"/> Hydrotherapy                            | <input type="checkbox"/> Provision of equipment  |
| <input type="checkbox"/> Home and/or school visits               | <input type="checkbox"/> Chest Physiotherapy     |
| <input type="checkbox"/> Wheelchair and/or seating assessment    | <input type="checkbox"/> Other                   |
| <input type="checkbox"/> Position and/or posture advice          |  |

Other (please specify)

**45. How satisfied are you with the AVAILABILITY of your current physiotherapy service (s)?**

- ☐ Extremely satisfied
- ☐ Satisfied
- ☐ Somewhat satisfied
- ☐ Not satisfied

Comment

**46. How satisfied are you with the QUALITY of your current physiotherapy service(s)?**

**Tick one and GO TO QUESTION 48**

- ☐ Extremely satisfied
- ☐ Satisfied
- ☐ Somewhat satisfied
- ☐ Not satisfied

Comment

### 3. Therapy Information - Paediatric Physiotherapy



## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

### \*47. Why has your child not attended a paediatric Physiotherapist?

- ☐ Not required / requested to date
- ☐ On waiting list
- ☐ Not available

## 3. Therapy Information - Specialised Equipment

### \*48. Which service provided the following equipment for your child?

Choose one option per equipment

	Specialist Seating (Wheelchair / Activity Chair etc)	Splints / Orthotics (AFO's, RGO's etc)
Enable Ireland	<input type="radio"/>	<input type="radio"/>
Central Remedial Clinic	<input type="radio"/>	<input type="radio"/>
Health Service Executive	<input type="radio"/>	<input type="radio"/>
Temple Street Children's University Hospital	<input type="radio"/>	<input type="radio"/>
Our Lady's Children's Hospital Crumlin	<input type="radio"/>	<input type="radio"/>
Other charitable service (e.g. SAOIRSE, COPE)	<input type="radio"/>	<input type="radio"/>
Private	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>
Not required to date	<input type="radio"/>	<input type="radio"/>

Comments

### 49. How often does your child's service review the following pieces of equipment?

Choose one option per equipment

	Specialist Seating	Splints / orthotics
Every 3-4 months	<input type="radio"/>	<input type="radio"/>
Every 6 months	<input type="radio"/>	<input type="radio"/>
Annually	<input type="radio"/>	<input type="radio"/>
Every 18 months	<input type="radio"/>	<input type="radio"/>
More than every 2 years	<input type="radio"/>	<input type="radio"/>
Only reviewed on request	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>

Other (please specify)

## 3. Therapy Information - Spina Bifida Clinic

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

### \*50. Does your child attend a specialised Spina Bifida Clinic?

☐ Yes GO TO QUESTION 51

☐ No GO TO QUESTION 54

If Yes, please name location

### 3. Therapy Information - Spina Bifida Multidisciplinary Team Clinic

#### 51. In your experience of attending a multidisciplinary Spina Bifida clinic, which of the following professionals have been available for consultation?

Tick one answer per row

	Available	Not available	Unsure
Neurosurgeon	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Paediatrician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Nurse Specialist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Physiotherapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Occupational Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthopaedic Surgeon	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Urologist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Psychology	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Social Worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Speech and Language Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Dietician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Family Support Worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Play Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please specify)

## PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire

**52. How important do you feel the presence of the following professionals is at the spina bifida clinic?**

**Tick one answer from each row**

	Very Important	Important	Moderately Important	Of Little Importance	Unimportant
Neurosurgeon	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Paediatrician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Nurse Specialist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Physiotherapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Occupational Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthopaedic Surgeon	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Urologist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Psychology	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Social Worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Speech and Language Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Dietician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Family Support Worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Play Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please specify)

**53. How satisfied are you with the service provided at the Spina Bifida Clinic?**

**Tick one**

- ☐ Extremely satisfied
- ☐ Satisfied
- ☐ Somewhat satisfied
- ☐ Not satisfied

**54. Can you think of any ways that the Spina Bifida Clinic could be further developed and improved?**

**\*Add additional answer sheet if required**

### 3. Therapy Information - Concerns and Suggestions



**PRINTED VERSION Spina Bifida Parent/Guardian Questionnaire**

**55. What are your main concerns (unmet needs) about your child at present?**

**\*Add additional answer sheet if required**

**56. Do you have any suggestions about how services could be improved for your child with Spina Bifida?**

**\*Add additional answer sheet if required**

# APPENDIX 3

# PRINTED VERSION Spina Bifida Service Provider Questionnaire

## Introduction

Dear Colleague,

Thank you for taking the time to complete this questionnaire. This is an important step to gathering information for a report on the services for children with Spina Bifida in Ireland. This research has been funded by the fundraising department of Temple Street Children's University Hospital and the researchers are Sarah Govermey, Senior Occupational therapist (BSc, MSc) and Eimear Culligan, Senior Physiotherapist (BSc, MSc, MISC) along with Dr Jane Leonard, Consultant Paediatrician (MB BCH BAO).

As a provider of services to children with Spina Bifida, this gives you the opportunity to highlight their needs and make recommendations to help improve services. The final results will be made available to families and professionals alike.

This questionnaire contains 3 sections regarding your services and recommendation, they include:

1. Role and Caseload
2. Assessment and Interventions
3. Transition, Shared Care and Service Development

If you see a \* next to a question, this is a compulsory question required for analysis purposes.

Kind Regards,  
Sarah & Eimear

Tel: 01-8784542 OR 01-8784563

## Role and Caseload

This section gathers details of your profession, demographic information, geographic location and your level of involvement with children (0-18 years) with Spina Bifida.

### \*1. What is your profession?

Tick one

- |   |   |                                    |
|---|---|------------------------------------|
| <input type="radio"/> Physiotherapist               | <input type="radio"/> Family Support Worker     | <input type="radio"/> Psychologist |
| <input type="radio"/> Occupational Therapist        | <input type="radio"/> Clinical Nurse Specialist | <input type="radio"/> Other        |
| <input type="radio"/> Paediatrician                 | <input type="radio"/> Liaison Nurse             |                                    |
| <input type="radio"/> Speech and Language Therapist | <input type="radio"/> Social Worker             |                                    |

Other (please specify)

### 2. What is your gender?

Tick one

- ☐ Male
- ☐ Female

**\*3. Which of the following services do you work in?**

**Tick one**

- ☐ Enable Ireland Early Intervention Team
- ☐ Enable Ireland School Age Team
- ☐ HSE Early Intervention Team
- ☐ HSE School Age Team
- ☐ Joint Initiative Early Intervention Team (detail agencies below)
- ☐ Joint Initiative School Age Team (detail agencies below)
- ☐ Central Remedial Clinic (CRC) (detail service type below e.g. specialist seating/feeding)
- ☐ Pre School Team (detail location below)
- ☐ Primary Care Team/Community Care
- ☐ Our Lady's Children's Hospital Crumlin
- ☐ Spina Bifida Hydrocephalus Ireland (SBHI)
- ☐ Other Voluntary Sector/Charity (please detail below)
- ☐ Temple Street Children's University Hospital
- ☐ Other

Please detail

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

### \*4. Geographically, which regions of Ireland does your specific service cover?

Tick all that apply

- |   |  |
|---|--|
| <input type="checkbox"/> Carlow               | <input type="checkbox"/> Laois           |
| <input type="checkbox"/> Cavan                | <input type="checkbox"/> Leitrim         |
| <input type="checkbox"/> Clare                | <input type="checkbox"/> Limerick        |
| <input type="checkbox"/> Cork South Lee       | <input type="checkbox"/> East Limerick   |
| <input type="checkbox"/> Cork North Lee       | <input type="checkbox"/> Longford        |
| <input type="checkbox"/> West Cork            | <input type="checkbox"/> Louth           |
| <input type="checkbox"/> North Cork           | <input type="checkbox"/> Mayo            |
| <input type="checkbox"/> Donegal              | <input type="checkbox"/> Meath           |
| <input type="checkbox"/> Dublin South         | <input type="checkbox"/> Monaghan        |
| <input type="checkbox"/> Dublin South East    | <input type="checkbox"/> Offaly          |
| <input type="checkbox"/> Dublin South City    | <input type="checkbox"/> Roscommon       |
| <input type="checkbox"/> Dublin South West    | <input type="checkbox"/> Sligo           |
| <input type="checkbox"/> Dublin West          | <input type="checkbox"/> North Tipperary |
| <input type="checkbox"/> North West Dublin    | <input type="checkbox"/> South Tipperary |
| <input type="checkbox"/> Dublin North Central | <input type="checkbox"/> Waterford       |
| <input type="checkbox"/> North Dublin         | <input type="checkbox"/> Westmeath       |
| <input type="checkbox"/> Galway               | <input type="checkbox"/> Wexford         |
| <input type="checkbox"/> Kerry                | <input type="checkbox"/> Wicklow         |
| <input type="checkbox"/> Kildare              | <input type="checkbox"/> West Wicklow    |
| <input type="checkbox"/> Kilkenny             | <input type="checkbox"/> Other           |

Specific area not included in the list above

### \*5. Which of the following are you currently involved with for children (0-18 years) with Spina Bifida?

Tick one

- ☐ Provide clinical inputs and/or services GO TO QUESTION 7
- ☐ Only involved in prescriptions/sanctioning of equipment GO TO QUESTION 6
- ☐ Not involved (services provided elsewhere) YOU ARE NOW FINISHED THIS QUESTIONNAIRE

Further Comments

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

If you are NOT INVOLVED then you are now finished this questionnaire - thank you very much for taking the time to complete this questionnaire.

For any queries please contact Sarah Governey (01-8784542) or Eimear Culligan (01-8784562)

### Role and Caseload

This section gives more information about your involvement in the prescription and/or sanctioning of equipment for children with Spina Bifida

**\*6. If you are involved in the prescriptions/sanctioning of the following equipment, how long does a child usually wait for it to arrive?**

**Tick one from each row**

	Not involved	0-3 months	3-6 months	6-12 months	12 + months
Wheelchairs and/or seating systems	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Sleeping systems	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Specialists beds/mattresses	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Walking aids	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Splint and/or orthoses	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Bath and/or toilet aids	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Ramps	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hoists	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Standers	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please specify)

### Role and Caseload

This section continues to gather information on your role and caseload highlighting the length of time you've been involved, the team you work with, the numbers of children you see with Spina Bifida and the waiting times.

**\*7. How many years have you been working with children with Spina Bifida?**

**Please indicate the figure in years (e.g. 5)**

# PRINTED VERSION Spina Bifida Service Provider Questionnaire

## 8. What is the availability of the following professionals within your service?

Please tick one answer per row

	Available	Required	Provided by another service	Not Provided
Paediatrician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Neurology/Neurosurgery	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthopaedic Surgeon	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Occupational Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Physiotherapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Clinical Nurse Specialist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Public Health Nurse	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Psychologist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthotist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Speech and Language Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Dietician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Social Worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Family Support Worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other, or if provided in another service please expand

## \*9. Please indicate below, how many children (under 18) with Spina Bifida you currently have on your various caseloads\*?

\*Please write 0 if you do not have any children in the listed area

Open or Active Caseload	<input type="text"/>
Waiting List	<input type="text"/>
Closed while Awaiting Equipment	<input type="text"/>

## \*10. How long, on average does a child with Spina Bifida have to wait for an appointment with your discipline following receipt of referral?

Tick one

- |   |                                   |
|---|-----------------------------------|
| <input type="radio"/> No waiting list         | <input type="radio"/> 6-9 months  |
| <input type="radio"/> 0-3 months              | <input type="radio"/> 9-12 months |
| <input type="radio"/> 3-6 months              | <input type="radio"/> 12 + months |
| <input type="radio"/> Other (please describe) |                                   |



## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**11. How long do children have to wait to transition to the next service after discharge from yours (e.g. from EIT to SAT or children to adult services)?**

**Tick one**

- |                                  |                                   |
|----------------------------------|-----------------------------------|
| <input type="radio"/> 0-3 months | <input type="radio"/> 9-12 months |
| <input type="radio"/> 3-6 months | <input type="radio"/> 12 + months |
| <input type="radio"/> 6-9 months | <input type="radio"/> Other       |

Please explain transition stage

### Assessment and Interventions

This section investigates the assessment and intervention practices currently in place nationally in order to establish where gaps in services may be evident. Questions ask about a range of potentially available services such as seating assessment and prescription, developmental therapy, pressure relief, bowel and bladder management and psychological interventions. This information will assist with developing guidelines and recommendations to assist with future services and practices related to children with Spina Bifida.

**\*12. How often are the following functional categories of children with Spina Bifida seen in your service by your profession (CURRENTLY)?**

**Tick one from each row**

	Weekly	Every 2 weeks	Monthly	Every 3 months	Every 6 months	Annually	Not seen
Infants	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Too young to tell mobility level yet	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Uses wheelchair most of the time	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Uses splints and walking aids sometimes, wheelchair sometimes	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Uses splints and sometimes needs help	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Walks without difficulty (with splints/aids)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Walks without difficulty (without splints/aids)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other, please explain



## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**\*13. In your professional opinion how often should the following categories of children with Spina Bifida be seen by your profession (RECOMMENDED)?**

**Tick one from each row**

	Weekly	Every 2 weeks	Monthly	Every 3 months	Every 6 months	Annually	Not seen
Infants	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Too young to tell mobility level yet	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Uses wheelchair most of the time	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Uses splints and walking aids sometimes, wheelchair sometimes	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Uses splints and sometimes needs help	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Walks without difficulty (with splints/aids)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Walks without difficulty (without splints/aids)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Further Comments

## Assessment and Interventions

**14. Which professional(s) are involved with assessment or prescription of wheelchairs and/or seating in your service? (please include your own profession if you are involved)**

**Tick all that apply**

- ☐ Assessed in another specialist service (i.e. seating clinic)
- ☐ Occupational Therapist
- ☐ Physiotherapist
- ☐ Engineer/Technician
- ☐ Company Representative
- ☐ Unsure
- ☐ Other

Other, please list. Not available, please detail

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**\*15. Are you directly involved with assessing for wheelchairs and/or seating systems?**

**Tick one**

- ☐ Yes GO TO QUESTION 16
- ☐ No GO TO QUESTION 19

Further Comments

### Assessment and Interventions

**\*16. In your service at what age on average is the first seating system and wheelchair assessments completed for a child with Spina Bifida?**

**Tick one**

	0-6 months	6-9 months	9-12 months	12-18 months	18-24 months	24+ months
Seating System Assessment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Wheelchair Assessment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Further Comments

**\*17. In your professional opinion at what age would you RECOMMEND a child with Spina Bifida should be assessed for their first seating system and wheelchair?**

**Tick one**

	0-6 months	6-9 months	9-12 months	12-18 months	18-24 months	24+ months
Seating System Assessment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Wheelchair Assessment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Further Comments

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**\*18. Following assessment and recommendation how long does it take for seating equipment to arrive?**

**Tick one**

	0-3 months	3-6 months	6-12 months	12+ months	Unsure (explain below)	Other
Seating System	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Wheelchair	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Please expand

**\*19. Are you involved with assessment and/or prescription of standers?**

**Tick one**

☐ Yes GO TO QUESTION 20

☐ No GO TO QUESTION 23

Further Comments

## Assessment and Interventions

**\*20. In your service at what age on average is an initial stander assessment completed for a child with Spina Bifida?**

**Tick one**

☐ 6-12 months

☐ 12-18 months

☐ 18 months – 2 years

☐ 2 – 2 ½ years

☐ 2 ½ years – 3 years

☐ Other

Further Comments

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**\*21. In your professional opinion at what age would you RECOMMEND a child with Spina Bifida should be assessed for their first stander?**

**Tick one**

- |   |   |
|---|---|
| <input type="radio"/> 6-12 months         | <input type="radio"/> 2 – 2 ½ years       |
| <input type="radio"/> 12-18 months        | <input type="radio"/> 2 ½ years – 3 years |
| <input type="radio"/> 18 months – 2 years | <input type="radio"/> Other               |

Further Comments

**\*22. Approximately how long following assessment and stander recommendation does equipment arrive?**

**Tick one**

- |                                   |                                  |
|-----------------------------------|----------------------------------|
| <input type="radio"/> 0-3 months  | <input type="radio"/> 12+ months |
| <input type="radio"/> 3-6 months  | <input type="radio"/> Unsure     |
| <input type="radio"/> 6-12 months | <input type="radio"/> Other      |

Further Comments

**\*23. Are you involved in advising or prescribing equipment related to sleep positioning (e.g. sleep systems) for children with Spina Bifida?**

**Tick one**

- ☐ Yes GO TO QUESTION 24
- ☐ No GO TO QUESTION 26

Further Comments

### Assessment and Interventions

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

### \*24. Why would you prescribe a sleep system for a child with Spina Bifida?

Tick all that apply

☐ Spinal alignment

☐ Limb positioning

☐ Pressure relief

☐ Hydrocephalus management

☐ Physiological need (e.g. respiratory, gastrointestinal)

☐ Other

Other (please specify)

### \*25. Approximately how long following recommendation does it take for a sleep system to arrive?

Tick one

☐ 0-3 months

☐ 3-6 months

☐ 6-12 months

☐ 12+ months

☐ Unsure

☐ Other

Other (please expand)

### \*26. Are you involved in the provision of splints and/or orthoses?

Tick one

☐ Yes GO TO QUESTION 28

☐ No GO TO QUESTION 27

Further Comments

## Assessment and Interventions

## \*27. What are the main reasons you do not provide splints?

Tick all that apply

- ☐ Splints/Orthoses provided by another professional within my team
- ☐ Lack of facilities (e.g. splint pan or materials)
- ☐ Lack of training/skills
- ☐ Completed by another specialist service
- ☐ Orthoses provided by externally contracted Orthotist
- ☐ Other (please specify)

GO TO QUESTION 29

## Assessment and Interventions

## \*28. Approximately how long do children with Spina Bifida in your service have to wait for their splints/orthoses following recommendation?

Tick one from each row

	0-3 months	3-6 months	6-12 months	12+ months	Unsure
Splints	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthoses	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Please expand



## PRINTED VERSION Spina Bifida Service Provider Questionnaire

### \*29. Who provides the following types of interventions for children with Spina Bifida within your service?

Tick all that apply from each row

	Provided Personally by Me	Provided by someone else in my team	Not provided	N/A
Splinting	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Pressure relief	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Positioning/Posture advice	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Feeding	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Mobility training	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Hydrotherapy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Chest Physio	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Learning and Attainment	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Strengthening	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Fine Motor Development	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Gross Motor Development	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Self Care (e.g. dressing, washing, toileting)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Communication	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Transfers (e.g. bed to chair)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Sexual functioning	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Other	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Other (please specify)

### 30. In your professional capacity how often do you visit children with Spina Bifida in their home environment?

Tick one

☐ Frequently (50+% of your time)

☐ Rarely (Less than 25% of your time)

☐ Occasionally (25+% of your time)

☐ Never

Further Comments

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**31. In your professional capacity how often do you visit children with Spina Bifida in their school environment?**

**Tick one**

☐ Frequently (50+% of your time)

☐ Rarely (Less than 25% of your time)

☐ Occasionally (25+% of your time)

☐ Never

Further Comments

**\*32. Do children with Spina Bifida have access to a Hydrotherapy pool in your service?**

**Tick one**

☐ Yes

☐ No

If yes please detail the service available

**\*33. In your opinion, how beneficial are Hydrotherapy services for children with Spina Bifida?**

**Tick one**

☐ Very beneficial

☐ Not beneficial at all

☐ Beneficial

☐ Unsure as not involved

☐ Somewhat beneficial

Further Comments



## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**\*34. Which professionals are involved in pressure relief advice and/or support in your service?**

**Tick all that apply**

- |  |  |
|--|--|
| <input type="checkbox"/> Occupational therapist    | <input type="checkbox"/> Paediatrician             |
| <input type="checkbox"/> Physiotherapist           | <input type="checkbox"/> Orthopaedic Surgeon       |
| <input type="checkbox"/> Liaison Nurse             | <input type="checkbox"/> Clinical nurse specialist |
| <input type="checkbox"/> General Practitioner (GP) | <input type="checkbox"/> Public Health Nurse       |
| <input type="checkbox"/> Other (please specify)    |  |

**\*35. If you have access to pressure mapping equipment, how frequently is this used for children with Spina Bifida children?**

**Tick one**

- |   |  |
|---|--|
| <input type="radio"/> Never               | <input type="radio"/> Sometimes                      |
| <input type="radio"/> Rarely              | <input type="radio"/> Occasionally                   |
| <input type="radio"/> Frequently          | <input type="radio"/> Don't have access to equipment |
| <input type="radio"/> On every assessment | <input type="radio"/> Completed in another service   |

Further Comments

**\*36. Which (if any) of the following Bladder or Bowel continence management services do you personally provide?**

**Tick all that apply**

- |  |
|--|
| <input type="checkbox"/> Prescriptions and treatments for Urinary Tract Infections (UTIs). |
| <input type="checkbox"/> Catheterisation advice/support                                    |
| <input type="checkbox"/> Toilet Adaptation Advice  |
| <input type="checkbox"/> Ultrasounds   |
| <input type="checkbox"/> Washouts advice   |
| <input type="checkbox"/> Toilet Training   |
| <input type="checkbox"/> None of the above (provided elsewhere)                            |
| <input type="checkbox"/> Other (please specify)  |

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**\*37. Within your service which professionals are involved in addressing the bowel and bladder continence needs?**

**Tick all that apply**

- |   |   |
|---|---|
| <input type="checkbox"/> Urologist              | <input type="checkbox"/> Public Health Nurse    |
| <input type="checkbox"/> Nurse specialist       | <input type="checkbox"/> Company Representative |
| <input type="checkbox"/> Occupational Therapist | <input type="checkbox"/> Not required           |
| <input type="checkbox"/> Family Support Worker  | <input type="checkbox"/> Other                  |
| <input type="checkbox"/> Paediatrician          |   |

Other (please specify)

**\*38. Are you involved with height and weight monitoring?**

**Tick one**

- ☐ Yes GO TO QUESTION 39
- ☐ No GO TO QUESTION 40

Further Comments

### Assessment and Interventions

**\*39. How often do you measure the height and weight of children with Spina Bifida?**

**Tick one from each row**

	More than once per month	Every 1-3 months	Every 3-6 months	Every 6 months to 1 year	Less than once a year	Other
Height	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Weight	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Please expand

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

### \*40. Are you involved with head circumference monitoring?

Tick one

- ☐ Yes GO TO QUESTION 41
- ☐ No GO TO QUESTION 42

Further Comments

## Assessment and Interventions

### \*41. How often do you measure the head circumference of children with Spina Bifida at the following ages?

Tick one from each row

	More than once per month	Every 1-3 months	Every 3-6 months	Every 6 months to 1 year	Less than once per year	Other
Infant	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
1-5 years	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
5+ years	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Further Comments

## Assessment and Interventions

### \*42. In your experience, are any of the following social, emotional or psychological concerns prevalent with children with Spina Bifida?

Tick all that apply

- |  |   |
|--|---|
| <input type="checkbox"/> Low self-esteem         | <input type="checkbox"/> Low mood/affect            |
| <input type="checkbox"/> Reduced confidence      | <input type="checkbox"/> Attention or concentration |
| <input type="checkbox"/> Bullying/isolation      | <input type="checkbox"/> Learning and attainment    |
| <input type="checkbox"/> Reduced quality of life |   |
| <input type="checkbox"/> Other (please specify)  |   |

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

### \*43. Where can a child in your service access psychology?

Tick one from each row

	On team psychologist	National Educational Psychology Service (NEPS)	Referral to another team	Private psychologist	None available	Other
Cognitive Assessment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Psychological support or intervention	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please specify)

### \*44. In your experience, are any of the following speech, language or swallowing needs prevalent with children with Spina Bifida?

Tick all that apply

- |   |  |
|---|--|
| <input type="checkbox"/> Feeding difficulties   | <input type="checkbox"/> Social Skills             |
| <input type="checkbox"/> Communication          | <input type="checkbox"/> Articulation or phonology |
| <input type="checkbox"/> Language skills        |  |
| <input type="checkbox"/> Other (please specify) |  |

### \*45. Where can a child in your service access speech and language therapy?

Tick all that apply

- ☐ On team speech and language therapist
- ☐ Refer to Primary Care/Community speech and language therapy service
- ☐ Ask parents to access private speech and language therapy
- ☐ Don't know / Not Available
- ☐ Refer to Early Intervention Team or School Age Team/Service (provided more than one need is present)
- ☐ Other (please specify)

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**\*46. In your experience, are any of the following visual difficulties prevalent with children with Spina Bifida?**

**Tick all that apply**

- |  |  |
|--|--|
| <input type="checkbox"/> Difficulty copying off the whiteboard                   | <input type="checkbox"/> Difficulties with hand eye-coordination                   |
| <input type="checkbox"/> Difficulty with visual tracking (e.g. following a ball) | <input type="checkbox"/> Difficulties with jigsaws and/or staying inside the lines |
| <input type="checkbox"/> Gazing at lights  | <input type="checkbox"/> Difficulty noticing details (e.g. in pictures)            |
| <input type="checkbox"/> Avoiding light (Photophobia)                            | <input type="checkbox"/> Fatiguing easily from visual tasks                        |
| <input type="checkbox"/> Loss of visual range (visual field loss)                | <input type="checkbox"/> Other   |

Other (please specify)

**\*47. In your experience, how frequently are the following aids or interventions used with children with Spina Bifida?**

**Tick one from each row**

	Very frequently	Frequently	Occasionally	Seldom	Not Applicable
Ankle Foot Orthosis (AFO)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Reciprocating Gait Orthosis (RGO)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Knee Ankle Foot Orthosis (KAFO)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hip Knee Ankle Foot Orthosis (HKAFO)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Mobility Aids	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthopaedic surgery	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Harnessing	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Casting	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Botox	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please specify)

**\*48. Which components of family education are you involved with for families of children with Spina Bifida?**

**Tick all that apply**

- |  |   |
|--|---|
| <input type="checkbox"/> Education on splinting                              | <input type="checkbox"/> Education regarding available benefits                         |
| <input type="checkbox"/> Education on equipment (e.g. chairs/walkers)        | <input type="checkbox"/> Education on bladder and bowel management                      |
| <input type="checkbox"/> Education on stretching or strengthening programmes | <input type="checkbox"/> Education regarding learning (e.g. numeracy, literacy, memory) |
| <input type="checkbox"/> Sexuality/Sexual functioning                        | <input type="checkbox"/> Wound management   |
| <input type="checkbox"/> Pressure Relief Advice                              | <input type="checkbox"/> Shunt management   |
| <input type="checkbox"/> Visual skills advice                                | <input type="checkbox"/> Medical Management   |
| <input type="checkbox"/> Diet  | <input type="checkbox"/> Other  |

Other (please specify)

**\*49. Which areas of family education are you not currently providing, but would like to be able to provide?**

**Tick all that apply**

- |  |   |
|--|---|
| <input type="checkbox"/> Education on splinting                              | <input type="checkbox"/> Education regarding available benefits                         |
| <input type="checkbox"/> Education on equipment (e.g. chairs/walkers)        | <input type="checkbox"/> Education on bladder and bowel management                      |
| <input type="checkbox"/> Education on stretching or strengthening programmes | <input type="checkbox"/> Education regarding learning (e.g. numeracy, literacy, memory) |
| <input type="checkbox"/> Sexuality/Sexual functioning                        | <input type="checkbox"/> Wound management   |
| <input type="checkbox"/> Pressure Relief Advice                              | <input type="checkbox"/> Shunt management   |
| <input type="checkbox"/> Visual skills advice                                | <input type="checkbox"/> Medical Management   |
| <input type="checkbox"/> Diet  | <input type="checkbox"/> Other  |

Other (please specify)



## PRINTED VERSION Spina Bifida Service Provider Questionnaire

### \* 50. What methods do you most frequently use to assist with family education?

Tick all that apply

- |   |  |
|---|--|
| <input type="checkbox"/> Group education                | <input type="checkbox"/> Electronic information (e.g. email, websites or Facebook) |
| <input type="checkbox"/> Individual education           | <input type="checkbox"/> Coffee mornings or meetings                               |
| <input type="checkbox"/> Visual education               | <input type="checkbox"/> Family events (e.g. fun days/camps etc)                   |
| <input type="checkbox"/> Written information (leaflets) | <input type="checkbox"/> Other   |

Other (please specify)

### \* 51. In your professional opinion, which service areas would you prioritise in relation to the management of a child with Spina Bifida?

Tick one from each row

	High priority	Medium priority	Low priority
Mobility	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Equipment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Postural management	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Psychological well-being	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Cognitive ability	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Visual abilities	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Speech and Language skills	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Feeding	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Self care (e.g. dressing)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Pressure relief	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Hand function	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other (specify)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please specify)

## Transition, Shared Care and Service Development

This final section investigates the transition for children with Spina Bifida into adulthood and some of the services involved in this important process. It also examines shared care between community care and local services with tertiary services such as Multidisciplinary (MDT) Spina Bifida Clinics. One example of an existing MDT Spina Bifida Clinic is the monthly clinic in Temple Street Children's University Hospital, the team present at this clinic consists of a Neurosurgeon, Paediatrician, Nurse Specialist, Physiotherapist and Occupational Therapist. Finally this section leaves space for you to highlight your goals and desires for future services for children with Spina Bifida in Ireland.

## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**\*52. Are you involved in the transition to adult services for children with Spina Bifida?**

**Tick one**

- ☐ Yes GO TO QUESTION 53
- ☐ No GO TO QUESTION 54

Further Comments

### Transition, Shared Care and Service Development

**\*53. Please explain some of your input or interventions during the transition to adulthood for children with Spina Bifida and how you feel these could be further developed.**

### Transition, Shared Care and Service Development

**\*54. If any of your clients have attended a specialist Spina Bifida clinic, how beneficial to your work have you found the following feedback?**

**Choose one from each row**

	Extremely Beneficial	Beneficial	Somewhat Beneficial	Not at all Beneficial	Not received
Written Communication (letters)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Electronic Communication (emails)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Telephone Communication	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Parental Feedback	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Overall impact of clinic on your work	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Further Comments



## PRINTED VERSION Spina Bifida Service Provider Questionnaire

**\*55. In your opinion how important do you feel the presence of the following professionals is for the effective running of a specialised Spina Bifida clinic?**

**Tick one from each row**

	Very Important	Important	Moderately Important	Of Little Importance	Unimportant
Neurosurgeon	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Paediatrician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Nurse Specialist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Physiotherapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Occupational Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Orthopaedic Surgeon	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Urologist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Psychology	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Social Worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Speech and Language Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Dietician	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Family Support Worker	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Play Therapist	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Other (please specify)

**56. Do you have any further comments or recommendations regarding MDT Spina Bifida Clinic's?**

Please add an extra sheet if required

**57. If you have any further comments which might support the health and therapy needs of children with Spina Bifida please include them below**

Please add an extra sheet if required

**THANK YOU**

Thank you very much for completing our questionnaire!

In order to gather more in depth information and to further probe issues highlighted by the questionnaires we will be conducting interviews around the country between September and November 2013.

If you would be interested in finding out more information about completing INTERVIEWS please contact the researchers on:

Sarah Governey, Occupational Therapist email [sarah.governey@cuhi.ie](mailto:sarah.governey@cuhi.ie) (01) 8784542

OR

Eimear Culligan, Physiotherapist on email [eimear.culligan@cuhi.ie](mailto:eimear.culligan@cuhi.ie) (01) 8784563.

# APPENDIX 4

# Questionnaire for children/adolescents

ID: \_ \_ \_ \_ \_



## Module: long-term illness/staying in hospital

Hello there!

We would like to know how you have been feeling during the past week, so we have worked out a few questions which we would like you to answer.

- ⇒ Please read each question carefully.
- ⇒ Think about how things have been for you over the past week.
- ⇒ Choose the answer that fits you best in each line and put a cross in the box.

There are no right or wrong answers. It's what you think that matters.

For example: 	never	seldom	some-times	often	all the time
During the past week, I liked to listen to music.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input checked="" type="checkbox"/>	<input type="checkbox"/>

Date of fill out:

\_\_\_\_\_

(day / month / year)

Please tell us something about you. Please put a cross or fill in!



I am a ☐ girl ☐ boy

Age: \_\_\_\_\_ years old

How many siblings do you have? ☐ 0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ more than 5

Which type of school do you go to? \_\_\_\_\_

<i>During the past week...</i>	<b>never</b>	<b>seldom</b>	<b>some- times</b>	<b>often</b>	<b>all the time</b>
1. ... I was afraid that my illness might get worse	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
2. ... I was sad because of my illness	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
3. ... I was able to cope well with my illness	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
4. ... My parents treated me like a baby because of my illness	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
5. ... I wanted nobody to notice my illness	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
6. ... I missed something at school because of my illness	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

**Thank you for helping us!**



# APPENDIX 5

## Questionnaires

### Parent / Guardian Interviews – Guide

Today we are interested in hearing your story about your experiences with your child \_\_\_\_ with spina bifida. We will start at the beginning when you first discovered that your child had spina bifida and will also discuss your hospital experience, your current services, how spina bifida has impacted on your child and family and any recommendations you may have for the future development of the service

Main Question	Additional questions	Clarifying Questions
<b>DIAGNOSIS</b>		
1. Can you describe your experience and feelings during the period of time that you discovered your child had spina bifida?	<ul style="list-style-type: none"> <li>When did you find out?</li> <li>How were you informed?</li> <li>Who informed you?</li> <li>Who was present at the time of the diagnosis?</li> <li>What information or support was offered</li> <li>How prepared did you feel when your child was born?</li> </ul>	<ul style="list-style-type: none"> <li>Can you expand on that a little?</li> <li>How did that make you feel?</li> </ul>
<b>MEDICAL CARE – Dr's, consultants, nurse specialists</b>		
2. Has your child had their spina bifida lesion repaired, and if so how did you find the hospital experience?	<ul style="list-style-type: none"> <li>Which hospital did your child attend?</li> <li>What interventions did your child receive (shunt / back surgery)?</li> <li>Did you feel you had adequate support while in hospital? How prepared were you for bringing your child home?</li> </ul>	<p><u>If positive:</u></p> <ul style="list-style-type: none"> <li>Who influenced your experience?</li> </ul> <p><u>If negative</u></p> <ul style="list-style-type: none"> <li>What could have improved your experience?</li> </ul>

3. Where does your child access medical services ( <i>Paeds, ortho, urologist, neurosurgeon, nurse specialist....</i> ), and How satisfied are you with your child's medical care to date?	<ul style="list-style-type: none"> <li>• Which hospital (s) does your child attend?</li> <li>• Are you concerned with waiting times for appointments in the hospital?</li> <li>• How supported do you feel in managing your child's bladder and bowel issues?</li> <li>• Do you have any concerns about access to medical specialists?</li> <li>• How coordinated do you feel the care is between your medical professionals?</li> </ul>	<ul style="list-style-type: none"> <li>• Examples of satisfaction or dissatisfaction?</li> <li>• How does that make you feel?</li> <li>• Is this an ongoing problem?</li> <li>• Suggestions for improvement?</li> </ul>
4. Does your child attend a specialist Spina Bifida clinic (MDT)?	<ul style="list-style-type: none"> <li>• Where?</li> <li>• How satisfied?</li> <li>• How could it be improved?</li> </ul>	
<b>SERVICES – therapy</b>		
5. How satisfied are you with your child's therapy service to date in relation to meeting their needs?	<ul style="list-style-type: none"> <li>• What type of team does your child attend?</li> <li>• How long did it take you to access these services?</li> <li>• What professional have been involved? Do they work as a team?</li> <li>• Are you satisfied with the quality and frequency of therapy interventions?</li> <li>• How have these services impacted positively or negatively on your family? family?</li> <li>• <i>Private??</i></li> </ul>	<ul style="list-style-type: none"> <li>• Examples of satisfaction or dissatisfaction?</li> <li>• I'm not familiar with that service, can you tell me more about it?</li> <li>• Can you give me more details?</li> <li>• Have you been able to highlight these concerns at a higherlevel?</li> </ul>



6. Do you feel fully informed of your child's potential and what are your main concerns for them in the future?	<ul style="list-style-type: none"> <li>• <b>Physical</b></li> <li>• Educational</li> <li>• Emotional</li> <li>• Social</li> <li>• <b>Transition</b> / becoming an adult</li> </ul>	<ul style="list-style-type: none"> <li>• Who can you talk to about this?</li> <li>• How do you feel about that?</li> </ul>
7. What do you feel are the greatest barriers to your child achieving their potential (for example equipment, delays, funding....)?	<ul style="list-style-type: none"> <li>• How could these barriers be overcome?</li> </ul>	
<b>QUALITY OF LIFE</b>		
8. Overall, how do you feel your child having spina bifida has impacted on your family life?	<ul style="list-style-type: none"> <li>• Family relationships – siblings, parental.</li> <li>• Financial implications</li> <li>• School</li> <li>• Community supports</li> <li>• Integration</li> <li>• Self-esteem, confidence, friends, happiness, participation with peers.</li> </ul>	<ul style="list-style-type: none"> <li>• How does that make you feel?</li> <li>• How have you coped with this?</li> <li>• What supports are in place and what should be in place?</li> </ul>
<b>IDEAS FOR THE FUTURE</b>		
9. Could you tell us some of your child's strengths or the good things about X?		

## Service Providers Interview – Guide

Today we want to understand a bit about your experience of working with children with spina bifida.

We're interested in your opinions of the accessibility of services, how they meet children's needs and the types of interventions that you personally provide. Finally we want to gather your recommendations which might help improve services in the future

Main Question	Main Question	Clarifying Questions
<b>PROFESSION AND EXPERIENCE</b>		
1. What is your current role with children with spina bifida?	<ul style="list-style-type: none"> <li>• Profession</li> <li>• How long are you qualified?</li> <li>• How many years have you been working with children with spina bifida? (Staff/Senior)</li> <li>• What type of service do you work in?</li> <li>• Who else is in your team?</li> <li>• How do you feel about the staff availability in your team?</li> </ul>	<ul style="list-style-type: none"> <li>• Can you expand on that a little?</li> <li>• And how do you think this works?</li> <li>• Do you feel confident working with children with spina bifida?</li> </ul>

SERVICE ACCESS		
<p>2. How easy or difficult is it for child with spina bifida to access your service?</p>	<ul style="list-style-type: none"> <li>How long roughly does a child have to wait to access your profession within your service?</li> <li>What do you think influences the waiting times?</li> </ul> <p><i>On reviewing the questionnaires, we were pleasantly surprised by the low waiting times to access services. Do you feel this accurately represents your service for children with spina bifida?</i></p> <p><i>*42% no waiting list, 41% less than 3 months wait</i></p>	<ul style="list-style-type: none"> <li>Do you have any recommendations on how this could be improved?</li> </ul>
<p>3. Do you feel your service adequately meets the needs of children with spina bifida? **Professional opinion</p>	<p>a. Frequency b. Quality c. Availability of professionals</p> <p><i>Noted disparity between how often children are currently seen and how often professionals feel they should be seen. Comments on this?</i></p> <p><i>*Infants, pre mobility, wheelchair users, walkers with aids and without</i></p>	<ul style="list-style-type: none"> <li>Do you have any recommendations on how this could be improved?</li> <li>What's hindering that?</li> </ul>

INTERVENTIONS		
4. Are you involved in providing any equipment for children with spina bifida?	<p><b>If providing equipment:</b></p> <ul style="list-style-type: none"> <li>• What types of equipment do you prescribe? (<i>stander, seating, orthoses, sleep systems, medical devices – catheters etc</i>)</li> <li>• How confident do you feel in prescribing or giving advice about specialist equipment for children with spina bifida?</li> <li>• How beneficial do you feel the equipment you prescribe is for children with spina bifida? <ul style="list-style-type: none"> <li>- <i>If standers</i>: What is your professional opinion on the use of <b>standers</b> for children with spina bifida?</li> <li>- <i>If sleep systems</i>: How beneficial do you feel a <b>sleep system</b> is if indicated for use with a child with spina bifida?</li> </ul> </li> <li>• Does a child have to wait long to receive the equipment you prescribe?</li> <li>• What do you think influences this wait time?</li> <li>• Does this wait time have an impact on your work?</li> </ul> <p><i>Noted disparity between when ax is completed and when professionals feel they should be completed.</i></p>	<p><b>If not prescribing equipment:</b></p> <ul style="list-style-type: none"> <li>• Do you know who to contact if you feel a piece of equipment is required?</li> <li>• How easy or difficult do you find it to liaise with other agencies involved in prescribing equipment?</li> <li>• Can you explain how this works further?</li> </ul>
5. Do children with spina bifida attending your service also get equipment from other agencies? (e.g. CRC. Private, Enable)	<ul style="list-style-type: none"> <li>• How easy or difficult do you find it to <b>liaise with other agencies</b> involved in prescribing equipment?</li> </ul>	

6. Do you have any comments on the bladder and bowel needs of children with spina bifida and how this impacts on their function?	a. Do you have anyone/where that you can refer children to when they are presenting with these difficulties? b. Please describe your involvement to assist with these difficulties (if any)?	
7. Do you have any ideas on how to improve the social, emotional and psychological well being of children with spina bifida?	a. Is there anyway this could be incorporated into your role? b. How do you feel these children could be better supported at home and in school? c. Where would you refer these children to for assistance?	
8. How do you assist children with visual difficulties as a result of spina bifida?	a. How would you identify these needs? b. How would you adapt your interventions to meet this need? c. Is there anywhere / one that you can refer a child with visual difficulties to?	<i>I don't:</i>  Do you know who you could refer a child to if they present with visual difficulties (i.e. CVI or visual perceptual difficulties)
<b>DEVELOPMENT</b>		
9. How do you feel care is co-ordinated between the community services and the paediatric hospitals?	<ul style="list-style-type: none"> <li>• Communication following appointment visits</li> <li>• Referrals</li> <li>• Professional advice and / or expertise</li> </ul>	Any recommendations on how this could be improved to better support your role and caseload
10. Have you had any children on your caseload who attend the spina bifida MDT Clinic in Temple Street?	Comments on: <ul style="list-style-type: none"> <li>• Communication</li> <li>• Liaison with professionals</li> <li>• Feedback received</li> </ul>	Any recommendations on how this could be improved to better support your role and caseload

<p>11. Do you have any recommendations on how services for children with spina bifida nationally could be improved or developed?</p>	<ul style="list-style-type: none"> <li>• Service access</li> <li>• Quality</li> <li>• Availability</li> <li>• Professional Training (CPD)</li> <li>• Liaison</li> <li>• MDT</li> </ul>	<p>Can you explain that in more detail</p>
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## Child Focus Group – Guide

Main Question	Additional Questions	Resources required
<b>INTRODUCTIONS – 10 MINS</b>		
<p>1. INTRODUCTION WITH PARENTS</p> <p>Intro, Ice Breaker</p> <p>Group Rules flip chart – let the children decide on rule with prompting if required.</p> <p><b>This is your group and a safe place for you to air your views. You can say what you like but if you tell us something that would make us worried about you, we can't guarantee secrecy</b> (danger, unsafe, very upset).</p> <ul style="list-style-type: none"> <li>• There are no right or wrong answers, say what you want</li> <li>• You can say pass if you don't want to answer</li> <li>• Take time to think before your answer</li> <li>• Tell me if I don't understand you or if you don't understand me</li> <li>• Take turns talking -</li> <li>• No teasing or making fun</li> </ul>	<ul style="list-style-type: none"> <li>• Aims for today are to discuss what it's like to have spina bifida.</li> <li>• Give Name Tags on entrance</li> <li>• Explain parents staying for first 5 minutes then just kids and facilitators just kids and facilitators</li> <li>• Ice Breaker – ball game.</li> <li>• Group Rules (flip chart)</li> <li>• Explain the need for activity calendar encourage them to include all therapy and extracurricular and social activities also on an average school going week.</li> </ul>	<p><b>Flipchart</b></p> <p><b>Name badges / stickers</b></p> <p><b>Ball</b></p>

KINDL – 10 MINS		
<p>2. KINDL</p> <p>As a group complete KINDL SB Questionnaire</p>	<p>We would like to know how you have been feeling during the past week, so we have worked out a few questions which we would like you to answer.</p> <ul style="list-style-type: none"> <li>• Please listen to or read each question carefully.</li> <li>• Think about how things have been for you over the past week.</li> <li>• Choose the answer that fits you best in each line and put a cross in the box.</li> <li>• <b>There are no right or wrong answers. It's what you think that matters</b></li> <li>• Read through individual questions, providing clarification if needed.</li> </ul>	<p>Explain the terms:</p> <ul style="list-style-type: none"> <li>• never</li> <li>• seldom (not very often)</li> <li>• sometimes</li> <li>• often (a lot of the time)</li> <li>• all the time</li> </ul> <p><b>KINDL chronic disease questionnaire</b></p> <p><b>Clipboards</b></p> <p><b>Pens</b></p>
ACTIVITY		
<p>3. How do you feel about how your body works?</p>	<p>Flip chart with body chart – indicate what parts of your body do not work so well</p> <ul style="list-style-type: none"> <li>• How does that make you feel?</li> <li>• How do you feel about having to use equipment to help you?</li> </ul>	<ul style="list-style-type: none"> <li>• Wheelchair</li> <li>• Stander Catheter</li> <li>• Splints Walking aids</li> </ul> <p>?show of hands</p> <ul style="list-style-type: none"> <li>• <b>Flip chart</b></li> <li>• <b>Body picture / chart</b></li> <li>• <b>Stickers</b></li> </ul>
<p>4. Tell us the good and bad things about the appointments you have to go to... (e.g. physio, doctors, nurses, OT)</p>	<ul style="list-style-type: none"> <li>• How do you feel about having to go to therapy?</li> <li>• How could these appointments be made better?</li> <li>• How are appointments different in the hospital and in your local team/service?</li> </ul>	



5. Tell me something that you like about yourself?	a. Can everyone take turns to tell us something that makes you feel good about or proud of yourself?	<b>Flipchart</b>
6. How does your spina bifida affect your family life?	a. If you could change anything in your family what would it be? b. Are you treated differently than your brother and sisters? c. Does attending appointments bother you or your family? d. Do you enjoy spending time with your family?	
7. Use your faces to tell us how happy you are when you are in school?	a. Why / what do you like / dislike b. What would you like to change?	<b>Faces on sticks (5 faces scale)</b>  (Very happy, happy, in between, unhappy, very unhappy)
8. How do you feel about making new friends?	a. How many? b. How nice or how they treat you? c. How easy it is to make new friends? d. Do you feel included? e. Have you ever been bullied? (why?)	<b>Faces on sticks (3 faces scale)</b>  (Not good, Ok, or Excellent)
Life Ladder	0-10 ladder scale. Best life =10, worst life =0, Please sticker where you see yourself at the moment	<b>Life ladder on colour A4 page</b>  <b>Stickers</b>
9. How do you feel about growing up?	<ul style="list-style-type: none"> <li>• Education / Jobs</li> <li>• Friends/ Relationships</li> <li>• Changing hospital</li> <li>• Having a family</li> </ul>	

10. How happy does having spina bifida make you feel?	Faces scale 1-5. Circle.  Does anyone want to say about that?	<b>Faces scales on A4 colour page</b>
<b>QUESTIONS</b>		
<b>IDEAS FOR THE FUTURE</b>		

Notes from meeting Social worker, Temple Street Children's University Hospital on 16<sup>th</sup> September 2013:

- Give the children ownership of the group
- Safe place but if anything that we are worried about we may have to disclose
- What would require disclosure:
  - o Unsafe / dangerous behaviour
  - o Inappropriate behaviour
  - o If child becomes very upset
    - Advised to explain to child the need to disclose this information
    - Talk to parents
    - Refer to Family Support Worker / Local social worker
- What circumstances would we encourage children to disclose:
  - o Admitting to bullying for first time
  - o Admitting to emotional upset not yet disclosed to parents
    - Advised to talk with child and encourage them to talk to parents, provide additional support if required to facilitate this
    - Talk with FSW
- Normalise the childrens answers, even if nobody else agrees with them. Especially the negatives.
- If a child is oversharing or overexposing themselves, ask them to stop and that we can talk about that later after the group.
- If a child becomes very upset, give them the option to stop or step outside
- If a child requires to leave the room, have an additional staff member to accompany them.

# APPENDIX 6



Dr. Jane Leonard  
Consultant Paediatrician  
Temple Street Children's University Hospital  
Temple Street  
Dublin 1

24<sup>th</sup> June 2013

**Re: 13.021. Investigating the health and therapy needs of children with Spina Bifida in Ireland**

Dear Dr. Leonard,

Your proposal was reviewed by the Ethics Committee at its recent meeting held on Tuesday, 11<sup>th</sup> June 2013.

No issues were raised and therefore the Committee is happy to grant ethics approval for your project.

Yours sincerely,



Prof Philip D Mayne  
Medical Secretary Ethics Committee  
MCRN: 06935

c.c. Department of Research

# REFERENCE LIST

ABRESCH, R. T., MCDONALD, D. A., WIDMAN, L. M., MCGINNIS, K. & HICKEY, K. J. 2007. Impact of spinal cord dysfunction and obesity on the health-related quality of life of children and adolescents. *The Journal Of Spinal Cord Medicine*, 30 Suppl 1, S112-S118. [LINK](#)

ADZICK, N. S., THOM, E. A., SPONG, C. Y., BROCK, J. W., 3RD, BURROWS, P. K., JOHNSON, M. P., HOWELL, L. J., FARRELL, J. A., DABROWIAK, M. E., SUTTON, L. N., GUPTA, N., TULIPAN, N. B., D'ALTON, M. E. & FARMER, D. L. 2011. A randomized trial of prenatal versus postnatal repair of myelomeningocele. *The New England Journal Of Medicine*, 364, 993-1004. [LINK](#)

AKBAR, M., BRESCH, B., RAISS, P., FÜRSTENBERG, C. H., BRUCKNER, T., SEYLER, T., CARSTENS, C. & ABEL, R. 2010. Fractures in myelomeningocele. *Journal of Orthopaedics & Traumatology*, 11, 175-182. [LINK](#)

ALDANA, P. R., WOOD, D. L., POSTLETHWAIT, R. A. & JAMES, H. E. 2010. Initiating, developing and evaluating a comprehensive spinal defects clinic: a clinical report. *Pediatric Neurosurgery*, 46, 329-334. [LINK](#)

ALRIKSSON-SCHMIDT, A. I., THIBADEAU, J. K., SWANSON, M. E., MARCUS, D., CARRIS, K. L., SIFFEL, C. & WARD, E. 2013. The natural history of spina bifida in children pilot project: research protocol. *JMIR Research Protocols*, 2, e2-e2. [LINK](#)

AMERICA., S. B. A. O. 2005. Health Guide for Adults Living With Spina Bifida. In: AMERICA, S. B. A. O. (ed.). Washington D.C. [LINK](#)

ANGOLD, A., MESSER, S. C., STANGL, D., FARMER, E. M. Z., COSTELLO, E. J. & BURNS, B. J. 1998. Perceived parental burden and service use for child and adolescent psychiatric disorders. *American Journal of Public Health*, 88, 75-80. [LINK](#)

ASCANI, E., BARTOLOZZI, P., LOGROSCINO, C. A., MARCHETTI, P. G., PONTE, A., SAVINI, R., TRAVAGLINI, F., BINAZZI, R. & DI SILVESTRE, M. 1986. Natural history of untreated idiopathic scoliosis after skeletal maturity. *Spine*, 11, 784-789. [LINK](#)

ASHTON, R. 2008. Improving the transfer to secondary school: How every child's voice can matter. *Support for Learning*, 23, 176-182. [LINK](#)

AU, K. S., ASHLEY-KOCH, A. & NORTHRUP, H. 2010. Epidemiologic and genetic aspects of spina bifida and other neural tube defects. *Developmental Disabilities Research Reviews*, 16, 6-15. [LINK](#)

BACON 2013. Does a Postural Management Training Programme Improve Understanding of the Importance of Postural Management for Children with Complex Movement Disorders? *Journal of the Association of Paediatric Chartered Physiotherapists*, 4, 27-36. [LINK](#)

BARF, H. A., VERHOEF, M., JENNEKENS-SCHINKEL, A., POST, M. W. M., GOOSKENS, R. H. J. M. & PREVO, A. J. H. 2003. Cognitive status of young adults with spina bifida. *Developmental medicine And Child Neurology*, 45, 813-820. [LINK](#)

- BARTONEK, Å. 2010. Motor development toward ambulation in preschool children with myelomeningocele---a prospective study. *Pediatric Physical Therapy*, 22, 52-60. [LINK](#)
- BARTONEK, A., ERIKSSON, M. & GUTIERREZ-FAREWIK, E. M. 2007. Effects of carbon fibre spring orthoses on gait in ambulatory children with motor disorders and plantarflexor weakness. *Developmental Medicine & Child Neurology*, 49, 615-620. [LINK](#)
- BARTONEK, Å. & SARASTE, H. 2001. Factors influencing ambulation in myelomeningocele: a cross-sectional study. *Developmental Medicine & Child Neurology*, 43, 253-260. [LINK](#)
- BATAVIA, M., GIANUTSOS, J. G., VACCARO, A. & GOLD, J. T. 2001. A do-it-yourself membrane-activated auditory feedback device for weight bearing and gait training: a case report. *Archives Of Physical Medicine And Rehabilitation*, 82, 541-545. [LINK](#)
- BAUER, S. & Joseph, D. 1990. Management of the obstructed urinary tract associated with neurogenic bladder dysfunction. *Urologic Clinic of North America*, 17, 395-406.
- BAUER, S. B., AUSTIN, P. F., RAWASHDEH, Y. F., DE JONG, T. P., FRANCO, I., SIGGARD, C. JORGENSEN, T. M. 2012. International Children's Continence Society's recommendations for initial diagnostic evaluation and follow-up in congenital neuropathic bladder and bowel dysfunction in children. *Neurourology And Urodynamics*, 31, 610-614. [LINK](#)
- BAX, M. 2001. Does 'therapy' have a future? *Developmental Medicine And Child Neurology*, 43, 3-3. [LINK](#)
- BELLIN, M. H. & RICE, K. M. 2009. Individual, family, and peer factors associated with the quality of sibling relationships in families of youths with spina bifida. *Journal of Family Psychology*, 23, 39-47. [LINK](#)
- BELLIN, M. H., ZABEL, T. A., DICIANNO, B. E., LEVEY, E., GARVER, K., LINROTH, R. & BRAUN, P. 2010. Correlates of depressive and anxiety symptoms in young adults with spina bifida. *Journal Of Pediatric Psychology*, 35, 778-789. [LINK](#)
- BOULTER, P. 1992. Learning disabilities. Using hydrotherapy: maximising benefits. *Nursing Standard (Royal College Of Nursing (Great Britain): 1987)*, 7, 25-27. [LINK](#)
- BOWMAN, R. M. & MCLONE, D. G. 2010. Neurosurgical management of spina bifida: research issues. *Developmental Disabilities Research Reviews*, 16, 82-87. [LINK](#)
- BOWMAN, R. M., MCLONE, D. G., GRANT, J. A., TOMITA, T. & ITO, J. A. 2001. Spina bifida outcome: a 25-year prospective. *Pediatric Neurosurgery*, 34, 114-120. [LINK](#)
- BOYD, P. A., DEVIGAN, C., KHOSHNOOD, B., LOANE, M., GARNE, E., DOLK, H. & GROUP, E. W. 2008. Survey of prenatal screening policies in Europe for structural malformations and chromosome anomalies, and their impact on detection and termination rates for neural tube defects and Down's syndrome. *BJOG*, 115, 689-96. [LINK](#)

- BRAUN, V. & CLARKE, V. 2006. Using thematic analysis in psychology. *Qualitative Research in Psychology*, 3, 77-101. [LINK](#)
- BREI, T. J. 2007. The future of multidisciplinary clinic. *The Scientific World. Special Issue: Challenges in Spina Bifida.*, 7, 1752-1756.
- BROUGHTON, N. S., MENELAUS, M. B., COLE, W. G. & SHURTLEFF, D. B. 1993. The natural history of hip deformity in myelomeningocele. *The Journal Of Bone And Joint Surgery. British Volume*, 75, 760-763. [LINK](#)
- BROWN, J. P. 2001. Orthopaedic care of children with spina bifida: you've come a long way, baby! *Orthopaedic Nursing*, 20, 51-58. [LINK](#)
- BRUSTROM, J., THIBADEAU, J., JOHN, L., LIESMANN, J. & ROSE, S. 2012. Care Coordination in the Spina Bifida Clinic Setting: Current Practice and Future Directions. *Journal of Pediatric Healthcare*, 26, 16-26. [LINK](#)
- BUFFART, L. M., ROEBROECK, M. E., ROL, M., STAM, H. J. & VAN DEN BERG-EMONS, R. J. G. 2008a. Triad of physical activity, aerobic fitness and obesity in adolescents and young adults with myelomeningocele. *Journal Of Rehabilitation Medicine: Official Journal Of The UEMS European Board Of Physical And Rehabilitation Medicine*, 40, 70-75.
- BUFFART, L. M., VAN DEN BERG-EMONS, R. J., BURDORF, A., JANSSEN, W. G., STAM, H. J. & ROEBROECK, M. E. 2008b. Cardiovascular disease risk factors and the relationships with physical activity, aerobic fitness, and body fat in adolescents and young adults with myelomeningocele. *Archives Of Physical Medicine And Rehabilitation*, 89, 2167-2173.
- BUFFART, L. M., WESTENDORP, T., VAN DEN BERG-EMONS, R. J., STAM, H. J. & ROEBROECK, M. E. 2009. Perceived barriers to and facilitators of physical activity in young adults with childhood-onset physical disabilities. *Journal Of Rehabilitation Medicine: Official Journal Of The UEMS European Board Of Physical And Rehabilitation Medicine*, 41, 881-885. [LINK](#)
- BURAN, C. F., MCDANIEL, A. M. & BREI, T. J. 2002. Needs assessment in a spina bifida program: a comparison of the perceptions by adolescents with spina bifida and their parents. *Clinical Nurse Specialist: The Journal for Advanced Nursing Practice*, 16, 256-262. [LINK](#)
- BURMEISTER, R., HANNAY, H. J., COPELAND, K., FLETCHER, J. M., BOUDOUSQUIE, A. & DENNIS, M. 2005. Attention problems and executive functions in children with spina bifida and hydrocephalus. *Child Neuropsychology: A Journal On Normal And Abnormal Development In Childhood And Adolescence*, 11, 265-283. [LINK](#)
- BYRNE, J. 2010. Birth defects among maternal first cousins in Irish families with a neural tube defect. *Irish Journal Of Medical Science*, 179, 375-380. [LINK](#)



- BYRNE, J. & CAROLAN, S. 2006. Adverse reproductive outcomes among pregnancies of aunts and (spouses of) uncles in Irish families with neural tube defects. *American Journal Of Medical Genetics. Part A*, 140, 52-61. [LINK](#)
- CAHILL, R. A. & KIELY, E. A. 2003. The spectrum of urological disease in patients with spina bifida. *Irish Journal Of Medical Science*, 172, 180-184. [LINK](#)
- CARDENAS, D. D., TOPOLSKI, T. D., WHITE, C. J., MCLAUGHLIN, J. F. & WALKER, W. O. 2008. Sexual functioning in adolescents and young adults with spina bifida. *Archives Of Physical Medicine And Rehabilitation*, 89, 31-35. [LINK](#)
- CARE QUALITY COMMISSION 2012. Health Care for disabled children and Young People. United Kingdom. [LINK](#)
- CARROLL, C., MURPHY, G. & SIXSMITH, J. 2013. The Progression of Early Intervention Disability Services in Ireland. *Infants & Young Children: An Interdisciplinary Journal of Special Care Practices*, 26, 17-27. [LINK](#)
- CARROLL, N. C. 1987. Assessment and management of the lower extremity in myelodysplasia. *The Orthopedic Clinics Of North America*, 18, 709-724. [LINK](#)
- CDC 1995. Surveillance for Anencephaly and Spina Bifida and the Impact of Prenatal Diagnosis United States, 1985-1994 Surveillance Summaries. [LINK](#)
- CHAKRABORTY, A., CRIMMINS, D., HAYWARD, R. & THOMPSON, D. 2008. Toward reducing shunt placement rates in patients with myelomeningocele. *Journal of Neurosurgery: Pediatrics*, 1, 361-365. [LINK](#)
- CHARNEY, E. B., ROSENBLUM, M. & FINEGOLD, D. 1981. Linear growth in a population of children with myelomeningocele. *Zeitschrift Für Kinderchirurgie: Organ Der Deutschen, Der Schweizerischen Und Der Österreichischen Gesellschaft Für Kinderchirurgie = Surgery In Infancy And Childhood*, 34, 415-419. [LINK](#)
- CHEN, C. P. 2005. Maternal diabetes and neural tube defects: prenatal diagnosis of lumbosacral myelomeningocele, ventriculomegaly, Arnold-Chiari malformation and foot deformities in a pregnancy with poor maternal metabolic control, and review of the literature. *Genetic Counseling (Geneva, Switzerland)*, 16, 313-316. [LINK](#)
- CHRISTAKIS, D. A., WRIGHT, J. A., ZIMMERMAN, F. J., BASSETT, A. L. & CONNELL, F. A. 2003. Continuity of Care Is Associated With Well-Coordinated Care. *Ambulatory Pediatrics*, 3, 82-86. [LINK](#)
- CLAYTON, D. B., BROCK, J. W., 3RD & JOSEPH, D. B. 2010. Urologic management of spina bifida. *Developmental Disabilities Research Reviews*, 16, 88-95. [LINK](#)
- CLEVE, L. V. 1989. Parental coping in response to their child's spina bifida. *Journal Of Pediatric Nursing*, 4, 172-176. [LINK](#)

COMMITTEE TO REVIEW NEUROSURGICAL SERVICES 2005. Report of the committee to review neurosurgical services in Ireland. *In*: NOSPIDÉAL, C. N. (ed.). Ireland.

CONNOR-KUNTZ, F. J., DUMMER, G. M. & PACIOREK, M. J. 1995. Physical education and sport participation of children and youth with spina bifida myelomeningocele. *Adapted Physical Activity Quarterly*, 12, 228-238. [LINK](#)

CORNEGÉ-BLOKLAND, E., JANSEN, H. E., DE JONG-DE VOS VAN STEENWIJK, C. C. E. & POENARU, D. 2011. Quality of life of children with spina bifida in Kenya is not related to the degree of the spinal defects. *Tropical Medicine & International Health: TM & IH*, 16, 30-36. [LINK](#)

COT. 2014. *OT as a Career* [Online]. London. Available: <http://www.cot.co.uk/study-ot/study-ot> [Accessed 13th August 2014]. [LINK](#)

COTTALORDA, J., VIOLAS, P. & SERINGE, R. 2012. Neuro-orthopaedic evaluation of children and adolescents: a simplified algorithm. *Orthopaedics & Traumatology, Surgery & Research: OTSR*, 98, S146-S153. [LINK](#)

CROSTHWAITE, A., CHUCHYARD, A., WILLIAMS, R., WELLS, J., DONELLEN, S. & TRUMBLE, S. 2001. Spina Bifida Overview. Victoria, Australia: Spina Bifida Foundation of Victoria.

CRESWELL, J. W. & PIANO-CLARK, V. L. 2007. *Designing and conducting mixed methods research.*, London, Sage Publications. [LINK](#)

CURTIN, C. 2001. Eliciting Children's Voices in Qualitative Research. *The American Journal of Occupational Therapy*, 55, 295-302. [LINK](#)

CURTIS, K., ROBERTS, H., COPPERMAN, J., DOWNIE, A. & LIABO, K. 2004. 'How come I don't get asked no questions?' Researching 'hard to reach' children and teenagers. *Child & Family Social Work*, 9, 167-175. [LINK](#)

CZAPRAN, P., GIBBON, F., BEATTIE, B., WILSON-JONES, N. & LEACH, P. 2012. Neural tube defects in Wales: changing demographics from 1998 to 2009. *British Journal Of Neurosurgery*, 26, 456-459. [LINK](#)

CZEIZEL, A. E. 2000. Primary prevention of neural-tube defects and some other major congenital abnormalities: recommendations for the appropriate use of folic acid during pregnancy. *Paediatric Drugs*, 2, 437-449. [LINK](#)

CZEIZEL, A. E. 2011. Periconceptional Folic Acid-Containing Multivitamin Supplementation for the Prevention of Neural Tube Defects and Cardiovascular Malformations. *Annals of Nutrition & Metabolism*, 59, 38-40. [LINK](#)

- D'EATH, M. S., JANE.CANNON , ROSEANNE.KELLY, LOUISE. 2005. The experience of people with disabilities in accessing health services in Ireland: do inequalities exist? Galway:Centre for Health Promotion Studies. Department of Health Promotion. National University of Ireland, Galway. [LINK](#)
- DAGENAIS, L. M., LAHAY, E. R., STUECK, K. A., WHITE, E., WILLIAMS, L. & HARRIS, S. R. 2009. Effects of electrical stimulation, exercise training and motor skills training on strength of children with meningomyelocele: a systematic review. *Physical & Occupational Therapy in Pediatrics*, 29, 445-463. [LINK](#)
- DANIELSSON, A. J., BARTONEK, A., LEVEY, E., MCHALE, K., SPONSELLER, P. & SARASTE, H. 2008. Associations between orthopaedic findings, ambulation and health-related quality of life in children with myelomeningocele. *Journal Of Children's Orthopaedics*, 2, 45-54 [LINK](#).
- DE JONG, T. P. V. M., CHRZAN, R., KLIJN, A. J. & DIK, P. 2008. Treatment of the neurogenic bladder in spina bifida. *Pediatric Nephrology (Berlin, Germany)*, 23, 889-896. [LINK](#)
- DE KORT, L. M. O., BOWER, W. F., SWITHINBANK, L. V., MARSCHALL-KEHREL, D., DE JONG, T. P. V. M. & BAUER, S. B. 2012. The management of adolescents with neurogenic urinary tract and bowel dysfunction. *Neurourology And Urodynamics*, 31, 1170-1174. [LINK](#)
- DEL BIGIO, M. R. 2010. Neuropathology and structural changes in hydrocephalus. *Developmental Disabilities Research Reviews*, 16, 16-22. [LINK](#)
- DELMARVA FOUNDATION 2006. Spina Bifida Clinic Survey Analysis Report.FOUNDATION, D. (ed.). [LINK](#)
- DENNIS, M. & BARNES, M. A. 2010. The cognitive phenotype of spina bifida meningomyelocele. *Developmental Disabilities Research Reviews*, 16, 31-39. [LINK](#)
- DENNIS, M., FLETCHER, J. M., ROGERS, T., HETHERINGTON, R. & FRANCIS, D. J. 2002. Object-based and action-based visual perception in children with spina bifida and hydrocephalus. *Journal Of The International Neuropsychological Society: JINS*, 8, 95-106. [LINK](#)
- DENNIS, M., JEWELL, D., HETHERINGTON, R., BURTON, C., BRANDT, M. E., BLASER, S. E. & FLETCHER, J. M. 2008. Verb generation in children with spina bifida. *Journal Of The International Neuropsychological Society: JINS*, 14, 181-191. [LINK](#)
- DENNIS, M., ROGERS, T. & BARNES, M. A. 2001. Children with spina bifida perceive visual illusions but not multistable figures. *Brain And Cognition*, 46, 108-113. [LINK](#)
- DEPARTMENT OF HEALTH AND CHILDREN 2011. Growing up in Ireland, National longitudinal study of children. Ireland. [LINK](#)

- DES 2007. Inclusion of students with special educational needs post primary guidelines. *In: SCIENCE, D. O. E. A. (ed.). Dublin: Stationary Office.* [LINK](#)
- DICKIE, V. A. 2003. Data analysis in qualitative research: A plea for sharing the magic and the effort. *American Journal of Occupational Therapy January/February, 57*, 49-56. [LINK](#)
- DIK, P., KLIJN, A. J., VAN GOOL, J. D., DE JONG-DE VOS VAN STEENWIJK, C. C. E. & DE JONG, T. P. V. M. 2006. Early start to therapy preserves kidney function in spina bifida patients. *European Urology, 49*, 908-913. [LINK](#)
- DOHC. 2001. *Quality and fairness—A health system for you.* *In: CHILDREN., D. O. H. A. (ed.). Dublin, Ireland.* [LINK](#)
- DOHC. 2003. Audit of the structure and functions in the health system. Prospectus putting strategy to work. *In: CHILDREN, D. O. H. A. (ed.). Dublin, Ireland:.* [LINK](#)
- DOSA, N. P., ECKRICH, M., KATZ, D. A., TURK, M. & LIPTAK, G. S. 2007. Incidence, prevalence, and characteristics of fractures in children, adolescents, and adults with spina bifida. *Journal of Spinal Cord Medicine, 30*, S5-9. [LINK](#)
- DOSA, N. P., FOLEY, J. T., ECKRICH, M., WOODALL-RUFF, D. & LIPTAK, G. S. 2009a. Obesity across the lifespan among persons with spina bifida. *Disability & Rehabilitation, 31*, 914-920. [LINK](#)
- DUNLEAVY, M. J. 2007. The role of the nurse coordinator in spina bifida clinics. *ScientificWorldJournal, 7*, 1884-9. [LINK](#)
- DUTTON, G. N., SAAED, A., FAHAD, B., FRASER, R., MCDAID, G., MCDADE, J., MACKINTOSH, A., RANE, T. & SPOWART, K. 2004. Association of binocular lower visual field impairment, impaired simultaneous perception, disordered visually guided motion and inaccurate saccades in children with cerebral visual dysfunction-a retrospective observational study. *Eye (London, England), 18*, 27-34. [LINK](#)
- DUVAL-BEAUPÈRE, G., KACI, M., LOUGOVOY, J., CAPONI, M. F. & TOUZEAU, C. 1987. Growth of trunk and legs of children with myelomeningocele. *Developmental Medicine And Child Neurology, 29*, 225-231 [LINK](#).
- ESSNER, B. S. & HOLMBECK, G. N. 2010. The Impact of Family, Peer, and School Contexts on Depressive Symptoms in Adolescents With Spina Bifida. *Rehabilitation Psychology, 55*, 340-350. [LINK](#)
- EUROCAT 1991. Working Group. Prevalence of neural tube defects in 20 regions of Europe and the impact of prenatal diagnosis, 1980–1986. *Journal of Epidemiology Community Health, 45*, 52–8. [LINK](#)

FARLEY, R., CLARK, J., DAVIDSON, C., EVANS, G., MACLENNAN, K., MICHAEL, S., MORROW, M. & THORPE, S. 2003. What is the evidence for the effectiveness of postural management?... including commentary by Roxborough L. *International Journal of Therapy and Rehabilitation*, 10, 449-455. [LINK](#)

FELDMAN, D., SWAINE, B., GOSSELIN, J., MESHEFEDJIAN, G. & GRILLI, L. 2008. Is waiting for rehabilitation services associated with changes in function and quality of life in children with physical disabilities? *Physical & Occupational Therapy in Pediatrics*, 28, 291-304 [LINK](#).

FILLER, G., GHARIB, M., CASIER, S., LÖDIGE, P., EHRICH, J. H. H. & DAVE, S. 2012. Prevention of chronic kidney disease in spina bifida. *International Urology And Nephrology*, 44, 817-827. [LINK](#)

FINLAY, L. 1999. Applying phenomenology in research: problems, principles and practice. *British Journal of Occupational Therapy*, 62, 299-306. [LINK](#)

FLETCHER, J. M., BARNES, M. & DENNIS, M. 2002. Language development in children with spina bifida. *Seminars In Pediatric Neurology*, 9, 201-208. [LINK](#)

FLETCHER, J. M. & BREI, T. J. 2010. Introduction: Spina bifida--a multidisciplinary perspective. *Developmental Disabilities Research Reviews*, 16, 1-5. [LINK](#)

FOOD SAFETY AUTHORITY OF IRELAND 2008. Report of implementation group on folic acid food fortification to the department of health and children. Dublin. [LINK](#)

FSAI 2013. FSAI Advises Against Any Further Folic Acid Food Fortification. [LINK](#)

GABRIELI, A. P. T., VANKOSKI, S. J., DIAS, L. S., MILANI, C., LOURENCO, A., FILHO, J. L. & NOVAK, R. 2003. Gait analysis in low lumbar myelomeningocele patients with unilateral hip dislocation or subluxation. *Journal Of Pediatric Orthopedics*, 23, 330-334. [LINK](#)

GAMÉ, X., MOSCOVICI, J., GAMÉ, L., SARRAMON, J.-P., RISCHMANN, P. & MALAUAUD, B. 2006. Evaluation of sexual function in young men with spina bifida and myelomeningocele using the International Index of Erectile Function. *Urology*, 67, 566-570. [LINK](#)

GANNON, B. & NOLAN, B. 2005. Disability and social inclusion in Ireland. In: INSTITUTE, E. A. S. R. (ed.). National Disability Authority and Equality Authority. [LINK](#)

GARDNER, M. F. 1996. *TVPS-R: Test of visual-perceptual skills (n-m) revised—Manual.*, Hydesville, CA, Psychological and Educational Publications.

GOLDSMITH, S. 2000. The Mansfield Project: postural care at night within a community setting: a feedback study. *Physiotherapy*, 86, 528-534. [LINK](#)

GOVERNMENT OF IRELAND 2004. Education for Persons with Special Educational Needs Act (EPSEN) 2004. *Number 30 of 2004*. Dublin: Government Stationary Office. [LINK](#)

- HADDAD, M. A. O., SEI, M., SAMPAIO, M. W. & KARA-JOSÉ, N. 2007. Causes of visual impairment in children: a study of 3,210 cases. *Journal Of Pediatric Ophthalmology And Strabismus*, 44, 232-240. [LINK](#)
- HARKNESS, S., MOSCARDINO, U., BERMÚDEZ, M. R., ZYLICZ, P. O., WELLES-NYSTRÖM, B., BLOM, M. & SUPER, C. M. 2006. Mixed Methods in International Collaborative Research: The Experiences of the International Study of Parents, Children, and Schools. *Cross-Cultural Research*, 40, 65-82. [LINK](#)
- HARVEY, B. 2007. *Evolution of health services and health policy in Ireland*. In: AGENCY, C. P. (ed.). Dublin, Ireland. [LINK](#)
- HAYES-ALLEN, M. C. & TRING, F. C. 1973. Obesity: another hazard for spina bifida children. *British Journal Of Preventive & Social Medicine*, 27, 192-196. [LINK](#)
- HETHERINGTON, R., DENNIS, M., BARNES, M., DRAKE, J. & GENTILI, F. 2006. Functional outcome in young adults with spina bifida and hydrocephalus. *Child's Nervous System: Chns: Official Journal Of The International Society For Pediatric Neurosurgery*, 22, 117-124. [LINK](#)
- HOLMBECK, G. N., DELUCIA, C., ESSNER, B., KELLY, L., ZEBRACKI, K., FRIEDMAN, D. & JANDASEK, B. 2010. Trajectories of psychosocial adjustment in adolescents with spina bifida: a 6-year, four-wave longitudinal follow-up. *Journal Of Consulting And Clinical Psychology*, 78, 511-525. [LINK](#)
- HOLMBECK, G. N. & DEVINE, K. A. 2010. Psychosocial and family functioning in spina bifida. *Developmental Disabilities Research Reviews*, 16, 40-46. [LINK](#)
- HOLMBECK, G. N., WESTHOVEN, V. C., PHILLIPS, W. S., BOWERS, R., GRUSE, C., NIKOLOPOULOS, T., TOTURA, C. M. W. & DAVISON, K. 2003. A multimethod, multi-informant, and multidimensional perspective on psychosocial adjustment in preadolescents with spina bifida. *Journal Of Consulting And Clinical Psychology*, 71, 782-796. [LINK](#)
- HORWATH CONSULTING IRELAND 2008. Review of Current Neurosurgery Services Final Report. Health Services Executive; Dublin Ireland. [LINK](#)
- HOULISTON, M. J., TAGURI, A. H., DUTTON, G. N., HAJIVASSILIOU, C. & YOUNG, D. G. 1999. Evidence of cognitive visual problems in children with hydrocephalus: a structured clinical history-taking strategy. *Developmental Medicine And Child Neurology*, 41, 298-306. [LINK](#)
- HSE. 2010. *National Service Plan*. Health Service Executive: Dublin, Ireland. [LINK](#)
- HUBER-OKRAINEC, J., DENNIS, M., BRETTSCHEIDER, J. & SPIEGLER, B. J. 2002. Neuromotor speech deficits in children and adults with spina bifida and hydrocephalus. *Brain And Language*, 80, 592-602. [LINK](#)



HUFF, C. W. & RAMSEY, P. L. 1978. Myelodysplasia. The influence of the quadriceps and hip abductor muscles on ambulatory function and stability of the hip. *The Journal Of Bone And Joint Surgery. American Volume*, 60, 432-443. [LINK](#)

HUMPHREYS, G. & POUNTNEY, T. 2006. The development and implementation of an integrated care pathway for 24-hour postural management: a study of the views of staff and carers. *Physiotherapy*, 92, 233-239. [LINK](#)

HUNT, G. M. 1981. Spina bifida: implications for 100 children at school. *Developmental Medicine And Child Neurology*, 23, 160-172. [LINK](#)

HUNT, G. M., OAKESHOTT, P. & KERRY, S. 1999. Link between the CSF shunt and achievement in adults with spina bifida. *Journal of Neurology, Neurosurgery & Psychiatry*, 67, 591-595. [LINK](#)

HÜSLER, M. R., DANZER, E., JOHNSON, M. P., BEBBINGTON, M., SUTTON, L., ADZICK, N. S. & WILSON, R. D. 2009. Prenatal diagnosis and postnatal outcome of fetal spinal defects without Arnold-Chiari II malformation. *Prenatal Diagnosis*, 29, 1050-1057. [LINK](#)

IBORRA, J., PAGÈS, E. & CUXART, A. 1999. Neurological abnormalities, major orthopaedic deformities and ambulation analysis in a myelomeningocele population in Catalonia (Spain). *Spinal Cord*, 37, 351-357. [LINK](#)

J, W. 2000. *Positioning and handling*, St Louis.

JEANNEROD, M. 2006. *Motor cognition*, Oxford, Oxford University Press. [LINK](#)

JOHNSON, K. L., DUDGEON, B., KUEHN, C. & WALKER, W. 2007. Assistive technology use among adolescents and young adults with spina bifida. *American Journal of Public Health*, 97, 330-336. [LINK](#)

JOHNSON, R. B., ONWUEGBUZIE, A. J. & TURNER, L. A. 2007b. Toward a definition of mixed methods research. *Journal of Mixed Methods Research*, 1, 112-133. [LINK](#)

JOSEPH, D. B. 2008. Current approaches to the urologic care of children with spina bifida. *Current Urology Reports*, 9, 151-157. [LINK](#)

KATRANCHA, E. D. 2008. Clean intermittent catheterization in the school setting. *Journal of School Nursing (Sage Publications Inc.)*, 24, 197-204. [LINK](#)

KATZ-LEURER, M., WEBER, C., SMERLING-KEREM, J., ROTTEM, H. & MEYER, S. 2004. Prescribing the reciprocal gait orthosis for myelomeningocele children: a different approach and clinical outcome. *Pediatric Rehabilitation*, 7, 105-109. [LINK](#)

- KAUFMAN, B. A., TERBROCK, A., WINTERS, N., ITO, J., KLOSTERMAN, A. & PARK, T. S. 1994. Disbanding a multidisciplinary clinic: effects on the health care of Myelomeningocele patients. *Pediatric Neurosurgery*, 21, 36-44. [LINK](#)
- KENNEDY, D., CHITAYAT, D., WINSOR, E. J., SILVER, M. & TOI, A. 1998. Prenatally diagnosed neural tube defects: ultrasound, chromosome, and autopsy or postnatal findings in 212 cases. *American Journal Of Medical Genetics*, 77, 317-321. [LINK](#)
- KENNY, S. & GOWRAN, R. J. 2014. Outcome measures for wheelchair and seating provision: a critical appraisal. *British Journal of Occupational Therapy*, 77, 67-77. [LINK](#)
- KINAVEY, C. 2007. Adolescents born with spina bifida: experiential worlds and biopsychosocial developmental challenges. *Issues in Comprehensive Pediatric Nursing*, 30, 147-164. [LINK](#)
- KING, G., LAW, M., PETRECHNIK, T. & HURLEY, P. 2013. Psychosocial determinants of out of school activity participation for children with and without physical disabilities. *Physical & Occupational Therapy In Pediatrics*, 33, 384-404. [LINK](#)
- KING, G., TUCKER, M. A., BALDWIN, P. J., LOWRY, K., LAPORTA, J. A. & MARTENS, L. 2002. A Life Needs Model of Pediatric Service Delivery. *Physical & Occupational Therapy in Pediatrics*, 22, 53-77. [LINK](#)
- KING, G. A., TUCKER, M. A., BALDWIN, P. J. & LAPORTA, J. A. 2006. Bringing the Life Needs Model to Life. *Physical & Occupational Therapy in Pediatrics*, 26, 43-70. [LINK](#)
- KINSMAN, S. L., LEVEY, E., RUFFING, V., STONE, J. & WARREN, L. 2000. Beyond Multidisciplinary Care: A New Conceptual Model for Spina Bifida Services. *Eur J Pediatr Surg*, 10, 35-38. [LINK](#)
- KULKARNI, A. V., DRAKE, J. M. & LAMBERTI-PASCULLI, M. 2001. Cerebrospinal fluid shunt infection: a prospective study of risk factors. *Journal of Neurosurgery*, 94, 195-201. [LINK](#)
- LAM, F. C., LOVETT, F. & DUTTON, G. N. 2010. Cerebral Visual Impairment in Children: A Longitudinal Case Study of Functional Outcomes Beyond the Visual Acuties. *Journal of Visual Impairment & Blindness*, 104, 625-635. [LINK](#)
- LAW, M., KING, G., KING, S., KERTOY, M., HURLEY, P., ROSENBAUM, P., YOUNG, N. & HANNA, S. 2006. Patterns of participation in recreational and leisure activities among children with complex physical disabilities. *Developmental Medicine And Child Neurology*, 48, 337-342. [LINK](#)
- LEHMAN, S. S. 2012. Cortical visual impairment in children: identification, evaluation and diagnosis. *Current Opinion In Ophthalmology*, 23, 384-387. [LINK](#)
- LEVEY, E. 2007. Addressing sexuality in spina bifida. *Pediatric News*. Special needs: realizing potential ed. Baltimore.: Kennedy Krieger Institute. [LINK](#)



LIPTAK, G. 2003. Evidence-Based Practice in Spina Bifida: Developing a research agenda. Spina Bifida Association of America (ed.). Centers for Disease Control and Prevention, Atlanta, Georgia Agency for Healthcare Research and Quality, Rockville, Maryland National Institutes of Health, Bethesda, Maryland Department of Education, Washington, DC. [LINK](#)

LIPTAK, G. S. & EL SAMRA, A. 2010. Optimizing health care for children with spina bifida. *Developmental Disabilities Research Reviews*, 16, 66-75. [LINK](#)

LOEBIG, M. 1990. Mothers' assessments of the impact of children with spina bifida on the family. *Maternal-Child Nursing Journal*, 19, 251-264. [LINK](#)

LOLLAR, D. J. 1994. Learning among children with spina bifida. In: SBAA (ed.) *Spina Bifida Spotlight*. [LINK](#)

LOMAX-BREAM, L. E., BARNES, M., COPELAND, K., TAYLOR, H. B. & LANDRY, S. H. 2007. The impact of spina bifida on development across the first 3 years. *Developmental Neuropsychology*, 31, 1-20. [LINK](#)

LORBER, J. 1972. Spina bifida cystica. Results of treatment of 270 consecutive cases with criteria for selection for the future. *Archives Of Disease In Childhood*, 47, 854-873. [LINK](#)

LYNCH, C. & MALONE, F. D. 2007. Prenatal screening and diagnosis. *Irish Medical Journal*, Mar;100, 405-7. [LINK](#)

LYNCH, S. A. 2005. Non-multifactorial neural tube defects. *American Journal Of Medical Genetics. Part C, Seminars In Medical Genetics*, 135C, 69-76. [LINK](#)

MAGNAN, M. A. 1996. Psychological considerations for patients with acute wounds. *Critical Care Nursing Clinics of North America*, 8, 183-193. [LINK](#)

MALAS, B. S. 2011. What Variables Influence the Ability of an AFO to Improve Function and When Are They Indicated? *Clinical Orthopaedics & Related Research*, 469, 1308-1314. [LINK](#)

MARGARON, F. C., POENARU, D., BRANSFORD, R. & ALBRIGHT, A. L. 2010. Timing of ventriculoperitoneal shunt insertion following spina bifida closure in Kenya. *Child's Nervous System: Chns: Official Journal Of The International Society For Pediatric Neurosurgery*, 26, 1523-1528. [LINK](#)

MARINI, N. J., HOFFMANN, T. J., LAMMER, E. J., HARDIN, J., LAZARUK, K., STEIN, J. B., GILBERT, D. A., WRIGHT, C., LIPZEN, A., PENNACCHIO, L. A., CARMICHAEL, S. L., WITTE, J. S., SHAW, G. M. & RINE, J. 2011. A genetic signature of spina bifida risk from pathway-informed comprehensive gene-variant analysis. *PLoS One*, 6, e28408. [LINK](#)

MAYFIELD, J. 1991. *Comprehensive orthopedic management in myelomeningocele*, Boston, CRC Press.

- MAZUR, J. M. & KYLE, S. 2004. Efficacy of bracing the lower limbs and ambulation training in children with myelomeningocele. *Dev Med Child Neurol*, 46, 352-6. [LINK](#)
- MAZUR, J. M. & MENELAUS, M. B. 1991. Neurologic status of spina bifida patients and the orthopedic surgeon. *Clinical Orthopaedics And Related Research*, 54-64. [LINK](#)
- MAZUR, J. M., SHURTLEFF, D., MENELAUS, M. & COLLIVER, J. 1989. Orthopaedic management of high-level spina bifida. Early walking compared with early use of a wheelchair. *The Journal Of Bone And Joint Surgery. American Volume*, 71, 56-61. [LINK](#)
- MCDONALD, C. M., JAFFE, K. M., MOSCA, V. S. & SHURTLEFF, D. B. 1991. Ambulatory outcome of children with myelomeningocele: effect of lower-extremity muscle strength. *Developmental Medicine & Child Neurology*, 33, 482-490. [LINK](#)
- MCDONNELL, G. V. & MCCANN, J. P. 2000. Issues of medical management in adults with spina bifida. *Child's Nervous System: Chns: Official Journal Of The International Society For Pediatric Neurosurgery*, 16, 222-227. [LINK](#)
- MCDONNELL, R., DELANY, V., O'MAHONY, M. T., MULLANEY, C., LEE, B. & TURNER, M. J. 2014. Neural tube defects in the Republic of Ireland in 2009–11. *Journal of Public Health*. [LINK](#)
- MCGONNELL, M. 2009. Changing needs for children and adults with spina bifida and hydrocephalus in Northern Ireland. *Cerebrospinal Fluid Research* 6, S45. [LINK](#)
- MCLONE, D. & DIAS, M. 2003. The Chiari II malformation: cause and impact. *Child's Nervous System*, 19, 540-550. [LINK](#)
- MCPHERSON, A. C., SWIFT, J. A., YUNG, E., LYONS, J. & CHURCH, P. 2013. The assessment of weight status in children and young people attending a spina bifida outpatient clinic: a retrospective medical record review. *Disability & Rehabilitation*, 35, 2123-2131. [LINK](#)
- MERKENS, M. J. 2006. Guidelines for Spina Bifida health care services throughout the lifespan. In: COUNCIL, P. A. (ed.). Washington DC: Spina Bifida Association. [LINK](#)
- MERRIAM, S. B. 2009. *Qualitative research: a guide to design and implementation*, San Francisco, Jossey-Bass. [LINK](#)
- MITA, K., AKATAKI, K., ITOH, K., ONO, Y., ISHIDA, N. & OKI, T. 1993. Assessment of obesity of children with spina bifida. *Developmental Medicine And Child Neurology*, 35, 305-311. [LINK](#)
- MORROW, V. & RICHARDS, M. 1996. The Ethics of Social Research with Children: An Overview. *Children & Society*, 10, 90-105. [LINK](#)
- MRC 1991. Prevention of neural tube defects: Results of the Medical Research Council Vitamin Study. *The Lancet*, 338, 131-137. [LINK](#)

- MULLIGAN, H. & POLKINGHORNE, A. 2013. Community use of a hospital pool by people with disabilities. *Disability And Health Journal*, 6, 385-390. [LINK](#)
- NATIONAL DISABILITY AUTHORITY 2011. *Report on the Practice of Assessment of Need under Part 2 of the Disability Act 2005*. [LINK](#)
- NETER, J. E., SCHOKKER, D. F., DE JONG, E., RENDERS, C. M., SEIDELL, J. C. & VISSCHER, T. L. 2011. The Prevalence of Overweight and Obesity and Its Determinants in Children with and without Disabilities. *Journal of Pediatrics*, 158, 735-739. [LINK](#)
- NEUROLOGICAL ALLIANCE OF IRELAND 2002. The future for neurological conditions in Ireland: A challenge for health care; an opportunity for change. [LINK](#)
- NHS. 2012. *Spina bifida - Treatment* [Online]. NHS. Available: <http://www.nhs.uk/conditions/spina-bifida/pages/treatment.aspx> [Accessed 18th August 2014]. [LINK](#)
- NHS. 2011. Children's Neurosurgical Specification Standards. National Health Service (ed.). UK [LINK](#).
- NORTHRUP, H. & VOLCIK, K. A. 2000. Spina bifida and other neural tube defects. *Current Problems in Pediatrics*, 30, 317-332. [LINK](#)
- OAKESHOTT, P., HUNT, G. M., POULTON, A. & REID, F. 2010. Expectation of life and unexpected death in open spina bifida: a 40-year complete, non-selective, longitudinal cohort study. *Developmental Medicine & Child Neurology*, 52, 749-753. [LINK](#)
- PADUA, L., RENDELI, C., RABINI, A., GIRARDI, E., TONALI, P. & SALVAGGIO, E. 2002. Health-related quality of life and disability in young patients with spina bifida. *Archives of Physical Medicine & Rehabilitation*, 83, 1384-1388. [LINK](#)
- PALLIJA, G., MONDOZZI, M. & WEBB, A. A. 1999. Skin care of the pediatric patient. *Journal of Pediatric Nursing*, 14, 80-87. [LINK](#)
- PARKES, J., DONNELLY, M., DOLK, H. & HILL, N. 2002. Use of physiotherapy and alternatives by children with cerebral palsy: a population study. *Child: Care, Health & Development*, 28, 469-477. [LINK](#)
- PARKES, J., HILL, N., DOLK, H. & DONNELLY, M. 2004. What influences physiotherapy use by children with cerebral palsy? *Child: Care, Health & Development*, 30, 151-160. [LINK](#)
- PASQUIER, J. C., MORELLE, M., BAGOUET, S., MORET, S., LUO, Z. C., RABILLOUD, M., GAUCHERAND, P. & ROBERT-GNANSIA, E. 2007. Effects of residential distance to hospitals with neonatal surgery care on prenatal management and outcome of pregnancies with severe fetal malformations. *Ultrasound in Obstetrics and Gynecology*, 29, 271-275. [LINK](#)

PENY-DAHLSTRAND, M., KRUMLINDE-SUNDHOLM, L. & GOSMAN-HEDSTROM, G. 2013. Patterns of participation in school-related activities and settings in children with spina bifida. *Disability & Rehabilitation*, 35, 1821-1827. [LINK](#)

PHILIP, S. S. & DUTTON, G. N. 2014. Identifying and characterising cerebral visual impairment in children: a review. *Clinical & Experimental Optometry*, 97, 196-208. [LINK](#)

PORTER, P., OBST, B. & ZABEL, A. 2009. A guide for school personnel working with students with Spina Bifida Kennedy Krieger Institute (ed.). Maryland: Maryland State Department of Education, Division of Special Education/Early Intervention Services. [LINK](#)

POUNTNEY, T., MANDY, A., GREEN, E. & GARD, P. 2002. Management of hip dislocation with postural management. *Child: Care, Health and Development*, 28, 179-185. [LINK](#)

PSIHOGIOS, A. M. & HOLMBECK, G. N. 2013. Discrepancies in mother and child perceptions of spina bifida medical responsibilities during the transition to adolescence: associations with family conflict and medical adherence. *Journal Of Pediatric Psychology*, 38, 859-870. [LINK](#)

RAWASHDEH, Y. F., AUSTIN, P., SIGGAARD, C., BAUER, S. B., FRANCO, I., DE JONG, T. P. & JORGENSEN, T. M. 2012. International Children's Continence Society's recommendations for therapeutic intervention in congenital neuropathic bladder and bowel dysfunction in children. *Neurourology And Urodynamics*, 31, 615-620. [LINK](#)

REINEHR, T., DOBE, M., WINKEL, K., SCHAEFER, A. & HOFFMANN, D. 2010. Obesity in disabled children and adolescents: an overlooked group of patients. *Deutsches Arzteblatt International*, 107, 268-75, 2p. [LINK](#)

RENDELI, C., NUCERA, E., AUSILI, E., TABACCO, F., RONCALLO, C., POLLASTRINI, E., SCORZONI, M., SCHIAVINO, D., CALDARELLI, M., PIETRINI, D. & PATRIARCA, G. 2006. Latex sensitisation and allergy in children with myelomeningocele. *Child's Nervous System: Chns: Official Journal Of The International Society For Pediatric Neurosurgery*, 22, 28-32. [LINK](#)

REVEN-SIEBERER, U., GOSCH, A., ABEL, T., AUQUIER, P., BELLACH, B. M., DUR, W., RAMJIL, L. & RAVENS-SIEBER, T. E. P. D. U. 2001. Quality of life in children and adolescents: a European public health perspective. *Social and preventative medicine*, 46, 297-302. [LINK](#)

RINTOUL, N. E., SUTTON, L. N., HUBBARD, A. M., COHEN, B., MELCHIONNI, J., PASQUARIELLO, P. S. & ADZICK, N. S. 2002. A new look at myelomeningoceles: functional level, vertebral level, shunting, and the implications for fetal intervention. *Pediatrics*, 109, 409-413. [LINK](#)

ROBERTS, D., SHEPHERD, R. W. & SHEPHERD, K. 1991. Anthropometry and obesity in myelomeningocele. *Journal Of Paediatrics And Child Health*, 27, 83-90. [LINK](#)

ROGERS, S. L., GORDON, C. Y., SCHANZENBACHER, K. E. & CASE-SMITH, J. 2001. Common diagnosis in pediatric occupational therapy practice. *In: CASE-SMITH, J. (ed.) Occupational Therapy for Children*. Fourth Edition ed. St. Louis: Mosby. [LINK](#)

ROLLAND-CACHERA, M. F. 2011. Childhood obesity: current definitions and recommendations for their use. *Int J Pediatr Obes*, 6, 325-31. [LINK](#)

ROSENBAUM, P. & STEWART, D. 2004. The World Health Organization International Classification of Functioning, Disability, and Health: a model to guide clinical thinking, practice and research in the field of cerebral palsy. *Seminars in Pediatric Neurology*, 11, 5-10. [LINK](#)

ROUSSOS, N., PATRICK, J. H., HODNETT, C. & STALLARD, J. 2001. A long-term review of severely disabled spina bifida patients using a reciprocal walking system. *Disability & Rehabilitation*, 23, 239-244. [LINK](#)

RYAN, K. D., PLOSKI, C. & EMANS, J. B. 1991. Myelodysplasia -- the musculoskeletal problem: rehabilitation from infancy to adulthood. *Physical Therapy*, 71, 935-946. [LINK](#)

SAVOYE, M., NOWICKA, P., SHAW, M., SUNKYUNG, Y., DZIURA, J., CHAVENT, G., O'MALLEY, G., SERRECCHIA, J. B., TAMBORLANE, W. V. & CAPRIO, S. 2011. Long-term Results of an Obesity Program in an Ethnically Diverse Pediatric Population. *Pediatrics*, 127, 402-410. [LINK](#)

SAWYER, S. M. & MACNEE, S. 2010. Transition to adult health care for adolescents with spina bifida: Research issues. *Developmental Disabilities Research Reviews*, 16, 60-65. [LINK](#)

SAWYER, S. M. & ROBERTS, K. V. 1999. Sexual and reproductive health in young people with spina bifida. *Developmental Medicine And Child Neurology*, 41, 671-675. [LINK](#)

SCAL, P. 2002. Transition for youth with chronic conditions: primary care physicians' approaches. *Pediatrics*, 110, 1315-21. [LINK](#)

SCHOENMAKERS, M. A. G., UITERWAAL, C. S. P., GULMANS, V. A. M., GOOSKENS, R. H. J. & HELDERS, P. J. M. 2005. Determinants of functional independence and quality of life in children with spina bifida. *Clinical Rehabilitation*, 19, 677-685. [LINK](#)

SCHOENMAKERS, M. A. G. C., DE GROOT, J. F., GORTER, J. W., HILLAERT, J. L. M., HELDERS, P. J. M. & TAKKEN, T. 2009. Muscle strength, aerobic capacity and physical activity in independent ambulating children with lumbosacral spina bifida. *Disability And Rehabilitation*, 31, 259-266. [LINK](#)

SHAER, C. M. 1997. The infant and young child with spina bifida: major medical concerns. *Infants & Young Children: An Interdisciplinary Journal of Special Care Practices*, 9, 13-25. [LINK](#)

- SHAER, C. M., CHESCHEIR, N. & SCHULKIN, J. 2007. Myelomeningocele: a review of the epidemiology, genetics, risk factors for conception, prenatal diagnosis, and prognosis for affected individuals. *Obstetrical & Gynecological Survey*, 62, 471-479. [LINK](#)
- SHAMAN, D. 2009. A team approach to Cortical Visual Impairment (CVI) in schools. University of North Dakota. [LINK](#)
- SHEPHERD, K., ROBERTS, D., GOLDING, S., THOMAS, B. J. & SHEPHERD, R. W. 1991. Body composition in myelomeningocele. *The American Journal of Clinical Nutrition*, 53, 1-6. [LINK](#)
- SHIN, M., KUCIK, J. E., SIFFEL, C., LU, C., SHAW, G. M., CANFIELD, M. A. & CORREA, A. 2012. Improved survival among children with spina bifida in the United States. *Journal of Pediatrics*, 161, 1132-1137.e3. [LINK](#)
- SHURTLEFF, D. B., WALKER, W. O., DUGUAY, S., PETERSON, D. & CARDENAS, D. 2010. Obesity and myelomeningocele: anthropometric measures. *Journal of Spinal Cord Medicine*, 33, 410-419. [LINK](#)
- SIM, J. & WRIGHT, C. 2000. Research in health care: concepts, designs and methods. Cheltenham: Stanley Thomas. [LINK](#)
- SIMEONSSON, R. J., MCMILLEN, J. S. & HUNTINGTON, G. S. 2002. Secondary conditions in children with disabilities: spina bifida as a case example. *Mental Retardation and Developmental Disabilities Research Reviews*, 8, 198-205. [LINK](#)
- SIMMS, B. 1987. The route learning ability of young people with spina bifida and hydrocephalus and their able-bodied peers. *Zeitschrift Für Kinderchirurgie: Organ Der Deutschen, Der Schweizerischen Und Der Österreichischen Gesellschaft Für Kinderchirurgie = Surgery In Infancy And Childhood*, 42 Suppl 1, 53-56. [LINK](#)
- SPINABIFIDA HQ. 2014. *Occupational & Physical Therapy Treatment for Spina Bifida* [Online]. Available: <http://www.spinabifida.net/occupational-physical-therapy-treatment-for-spina-bifida.html> [Accessed 18th August 2014. [LINK](#)
- STALLARD, J., LOMAS, B., WOOLLAM, P., FARMER, I. R., JONES, N., POINER, R. & MILLER, K. 2003. New technical advances in swivel walkers. *Prosthetics And Orthotics International*, 27, 132-138. [LINK](#)
- STEERS, J. & STOWER, S. 2010. Safe and Sustainable Paediatric Neurosurgery: Report of the Unit Visits. London: NHS.
- SWANK, M. & DIAS, L. S. 1994. Walking ability in spina bifida patients: a model for predicting future ambulatory status based on sitting balance and motor level. *Journal Of Pediatric Orthopedics*, 14, 715-718. [LINK](#)



- TAYLOR, H. B., LANDRY, S. H., BARNES, M., SWANK, P., COHEN, L. B. & FLETCHER, J. 2010. Early information processing among infants with and without spina bifida. *Infant Behavior & Development*, 33, 365-372. [LINK](#)
- TEULIER, C., SMITH, B. A., KUBO, M., CHANG, C., MOERCHEN, V., MURAZKO, K. & ULRICH, B. D. 2009. Stepping responses of infants with myelomeningocele when supported on a motorized treadmill. *Physical Therapy*, 89, 60-72. [LINK](#)
- THOMPSON, D. N. 2009. Postnatal management and outcome for neural tube defects including spina bifida and encephaloceles. *Prenatal Diagnosis*, 29, 412-419. [LINK](#)
- THOMSON, J. D., OUNPUU, S., DAVIS, R. B. & DELUCA, P. A. 1999. The effects of ankle-foot orthoses on the ankle and knee in persons with myelomeningocele: an evaluation using three-dimensional gait analysis. *Journal Of Pediatric Orthopedics*, 19, 27-33. [LINK](#)
- THOMSON, J. D. & SEGAL, L. S. 2010. Orthopedic management of spina bifida. *Developmental Disabilities Research Reviews*, 16, 96-103. [LINK](#)
- TRIVEDI, J., THOMSON, J. D., SLAKEY, J. B., BANTA, J. V. & JONES, P. W. 2002. Clinical and radiographic predictors of scoliosis in patients with myelomeningocele. *The Journal Of Bone And Joint Surgery. American Volume*, 84-A, 1389-1394. [LINK](#)
- TSAI, P. Y., YANG, T. F., CHAN, R. C., HUANG, P. H. & WONG, T. T. 2002. Functional investigation in children with spina bifida -- measured by the Pediatric Evaluation of Disability Inventory (PEDI). *Child's Nervous System: Chns: Official Journal Of The International Society For Pediatric Neurosurgery*, 18, 48-53. [LINK](#)
- TURNER, S. L. 2007. Introduction to special issue: Transitional issues for K-16 students. *Professional School Counseling*, 10, 224-226. [LINK](#)
- UN. 2014. *Convention on rights of persons with disabilities* [Online]. Geneva: United Nations. Available: <http://www.un.org/disabilities/convention/conventionfull.shtml> [Accessed 15th October. [LINK](#)
- VAN DEN BERG-EMONS, H. J., BUSSMANN, J. B., BROBBEL, A. S., ROEBROECK, M. E., VAN MEETEREN, J. & STAM, H. J. 2001. Everyday physical activity in adolescents and young adults with meningomyelocele as measured with a novel activity monitor. *The Journal Of Pediatrics*, 139, 880-886. [LINK](#)
- VAN DEN BERG-EMONS, H. J. G., BUSSMANN, J. B. J., MEYERINK, H. J., ROEBROECK, M. E. & STAM, H. J. 2003. Body fat, fitness and level of everyday physical activity in adolescents and young adults with meningomyelocele. *Journal Of Rehabilitation Medicine: Official Journal Of The UEMS European Board Of Physical And Rehabilitation Medicine*, 35, 271-275. [LINK](#)
- VANKOSKI, S. J., MICHAUD, S. & DIAS, L. 2000. External tibial torsion and the effectiveness of the solid ankle-foot orthoses. *Journal Of Pediatric Orthopedics*, 20, 349-355. [LINK](#)

- VEGA-SANCHEZ, R., DE LA LUZ GOMEZ-AGUILAR, M., HAUA, K. & ROZADA, G. 2012. Weight-based nutritional diagnosis of Mexican children and adolescents with neuromotor disabilities. *BMC Research Notes*, 5, 218-218. [LINK](#)
- VERHOEF, M., BARF, H. A., POST, M. W. M., VAN ASBECK, F. W. A., GOOSKENS, R. H. J. M. & PREVO, A. J. H. 2006. Functional independence among young adults with spina bifida, in relation to hydrocephalus and level of lesion. *Developmental Medicine & Child Neurology*, 48, 114-119. [LINK](#)
- VERHOEF, M., BARF, H. A., VROEGE, J. A., POST, M. W., VAN ASBECK, F. W., GOOSKENS, R. H. & PREVO, A. J. 2005a. Sex education, relationships, and sexuality in young adults with spina bifida. *Archives Of Physical Medicine And Rehabilitation*, 86, 979-987. [LINK](#)
- VERHOEF, M., BARF, H. A., VROEGE, J. A., POST, M. W., VAN ASBECK, F. W., GOOSKENS, R. H. & PREVO, A. J. 2005b. Sex education, relationships, and sexuality in young adults with spina bifida. *Archives of Physical Medicine & Rehabilitation*, 86, 979-987. [LINK](#)
- VERHOEF, M., LURVINK, M., BARF, H. A., POST, M. W. M., VAN ASBECK, F. W. A., GOOSKENS, R. H. J. M. & PREVO, A. J. H. 2005c. High prevalence of incontinence among young adults with spina bifida: description, prediction and problem perception. *Spinal Cord*, 43, 331-340. [LINK](#)
- VERMAES, I. P. R., JANSSENS, J. M. A., MULLAART, R. A., VINCK, A. & GERRIS, J. R. M. 2008. Parents' personality and parenting stress in families of children with spina bifida. *Child: Care, Health & Development*, 34, 665-674. [LINK](#)
- VERMAES, I. P. R., JANSSENS, J. M. A. M., BOSMAN, A. M. T. & GERRIS, J. R. M. 2005. Parents' psychological adjustment in families of children with spina bifida: a meta-analysis. *BMC Pediatrics*, 5, 32-32. [LINK](#)
- VINCK, A., MAASSEN, B., MULLAART, R. & ROTTEVEEL, J. 2006. Arnold-Chiari-II malformation and cognitive functioning in spina bifida. *Journal Of Neurology, Neurosurgery, And Psychiatry*, 77, 1083-1086. [LINK](#)
- VINER, R. 2001. Barriers and good practice in transition from paediatric to adult care. *Journal of the Royal Society of Medicine*, 94, 2-4. [LINK](#)
- WAI, E. K., YOUNG, N. L., FELDMAN, B. M., BADLEY, E. M. & WRIGHT, J. G. 2005. The relationship between function, self-perception, and spinal deformity: Implications for treatment of scoliosis in children with spina bifida. *Journal Of Pediatric Orthopedics*, 25, 64-69. [LINK](#)
- WANDEL, J. 2000. *Positioning and handling*, St Louis.
- WARF, B., ONDOMA, S., KULKARNI, A., DONNELLY, R., AMPEIRE, M., AKONA, J., KABACHELOR, C. R., MULONDO, R. & NSUBUGA, B. K. 2009. Neurocognitive outcome and ventricular volume in children with myelomeningocele treated for hydrocephalus in Uganda. *Journal Of Neurosurgery. Pediatrics*, 4, 564-570. [LINK](#)



- WATSON, D. 1991. Occupational therapy intervention guidelines for children and adolescents with spina bifida. *Child: Care, Health and Development*, 17, 367-380. [LINK](#)
- WHO 2001. International classification of functioning, disability and health. Geneva: World Health Organisation. [LINK](#)
- WHO. 2008. Guidelines on the provision of Manual Wheelchairs in less resourced settings. Geneva: World Health Organisation. [LINK](#)
- WIDMAN, L. M., ABRESCH, R. T., STYNE, D. M. & MCDONALD, C. M. 2007. Aerobic fitness and upper extremity strength in patients aged 11 to 21 years with spinal cord dysfunction as compared to ideal weight and overweight controls. *The Journal Of Spinal Cord Medicine*, 30 Suppl 1, S88-S96. [LINK](#)
- WOODHOUSE, C. J. 2008. Myelomeningocele: neglected aspects. *Pediatric Nephrology*, 23, 1223-1231. [LINK](#)
- WORLD HEALTH ORGANISATION 2001. *International classification of Functioning, Disability and Health (ICF)*. [LINK](#)
- WRIGHT, C., CASEY, J. & PORTER-ARMSTRONG, A. 2010. Establishing best practice in seating assessment for children with physical disabilities using qualitative methodologies. *Disability & Rehabilitation: Assistive Technology*, 5, 34-47. [LINK](#)
- WRIGHT, J. G. 2011. Hip and Spine Surgery is of Questionable Value in Spina Bifida: An Evidence-based Review. *Clinical Orthopaedics & Related Research*, 469, 1258-1264. [LINK](#)
- WRIGHT, M., FOLEY, D. & STELL, D. 2009. *Spina Bifida Literature Review. Unpublished Thesis*.
- YOUNG, N. L., STEELE, C., FEHLINGS, D., JUTAI, J., OLMSTED, N. & WILLIAMS, J. I. 2005. Use of health care among adults with chronic and complex physical disabilities of childhood. *Disability & Rehabilitation*, 27, 1455-1460. [LINK](#)
- ZEGERS, B. S. H., WINKLER-SEINSTRAL, P. L. H., UITERWAAL, C. S. P., DE JONG, T. V. P., KIMPEN, J. L. L. & VAN STEENWIJK, C. C. E. 2009. Urinary tract infections in children with spina bifida: an inventory of 41 European centers. *Pediatric Nephrology*, 24, 783-788. [LINK](#)
- ZIMMET, P., KGM, M. M. A. & SERRANO RIOS, M. 2005. [A new international diabetes federation worldwide definition of the metabolic syndrome: the rationale and the results]. *Rev Esp Cardiol*, 58, 1371-6. [LINK](#)

