Closed Neural Tube Defects: Neurologic, Orthopedic, and Gait Outcomes

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Purpose: The purpose of our study was to obtain a clear understanding of the various diagnoses within the closed neural tube defect (NTD) groups included in the large database of clients in our Spina Bifida Clinic and a clear picture of the outcomes for the various NTD groups. **Methods:** One hundred and four clients with closed NTD were categorized using a classification system devised by Tortori-Donati et al. Various clinical markers were investigated, including ambulation and the need for orthoses and wheelchairs. **Results:** Most clients are ambulatory, requiring an orthoses, but not a wheelchair, despite the high incidence of ankle/foot abnormalities. **Conclusions:** This classification system has enhanced our knowledge of this group of clients, provided a greater understanding of the varied outcomes of these children and clinical management required. **(Pediatr Phys Ther 2007;19:288–295)** *Key words: ambulation, children, classification, diagnosis, outcome study, rehabilitation, retrospective study, spinal dysraphism, wheelchair*

INTRODUCTION

At Bloorview Kids Rehab, a pediatric rehabilitation tertiary care facility in Ontario, Canada, physiotherapists in the Spina Bifida Clinic work with children who have open or closed neural tube defects (NTDs). Since 1995, there has been a reduction in the number of babies referred to this facility with myelomeningocele, a type of open NTD. Despite this decrease, the total number of referrals received on a yearly basis to the clinic has remained constant since the referrals for clients with closed NTD have increased. For example, in the first six months of 2003 when this study was conducted, the number of referrals for open and closed NTD was equal. However, before 1995, the ratio of open to closed NTD was approximately five to one. The decreasing incidence of babies born with open NTD is similarly reported for North America.[1,2] This change has likely occurred because of the increased use of folic acid and genetic counseling.[1,2] The observed increase in the number of children with closed NTD prompted our investigation.

The general terms of spina bifida and spinal dysraphism are often given as the diagnosis for children with closed NTD. Some authors use the term NTD to describe the entire group of neural tube anomalies,[2–4] while others divide the anomalies into open and closed NTD.[5,6] The lack of specificity can be a source of confusion in the understanding and management of these children. A literature search was conducted using the Medline and CINAHL databases for the period from 1983 to February 2006. Key words included the various types of closed neural tube defects (as outlined in the NTD classification system that follows), "gait," "mobility," "ambulation," and "function." No publications were found by the authors that covered outcomes across the breadth of closed NTD diagnostic categories although some information was found about the functional outcomes within specific categories. Thus, the purpose of this study was to apply a more detailed NTD classification system to better understand the multiple types of NTD within this group, and the associated implications for clinical management and outcomes.

METHOD

This study consisted of a retrospective chart review of clients with closed NTD. The first two authors collected

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data from the physiotherapy assessments, magnetic resonance imaging, and neurosurgical reports found in the clients' health records. Each client was then categorized using the classification system of Tortori-Donati et al.[6]

Classification Instrument

Three classification systems were explored: one embryology-based approach proposed by Tortori-Donati et al[6] and two clinically based systems, one by Tortori-Donati et al,[6] and the other by McComb.[5] The clinically based system of Tortori-Donati et al[6] was selected for use in this study because it was the most clinically comprehensive and linked the neuroradiologic and clinical findings with current embryologic theory. To develop this classification system, Tortori-Donati et al reviewed the neuroradiologic features of spinal dysraphism for 986 patients referred to their centre over a 24-year period. The Tortori-Donati classification system divides the closed NTD into two categories: those with a subcutaneous mass and those without a subcutaneous mass. Closed NTD with a subcutaneous mass are divided into lumbosacral and cervical categories. The category of closed NTD without a subcutaneous mass is divided into simple and complex dysraphic states. Table 1 outlines all categories found in the Tortori-Donati classification system.

Sample

The authors selected 110 charts (71%) at random from among the 145 clients with closed NTD seen in the Spina Bifida Clinic in 2003. Six of the 110 charts selected were considered ineligible because of difficulties determining a clear diagnosis; thus, the chart review was completed for 104 clients. Given the broad geographic catchment area of our clinic and the use of random sampling methods, the sample is expected to be representative of the wider group of children in our province with spina bifida.

The first and second authors, both of whom are PTs, initially worked together to review the diagnostic information in the health record and classify the clients according to the Tortori-Donati system. Subsequently, each of these two authors was responsible for determining the Tortori-Donati classification category for approximately half of the clients. If the rater was uncertain about a child's group allocation as a result of perceived ambiguity in the health record, she then conferred with the other rater, and they arrived at a consensus as to the most suitable category.

Clinical Markers and Gait Outcomes

The following clinical markers were considered to gain an understanding of how the clients presented in each of the classification categories: incidence and age of tethered cord release, the need for repeat tethered cord release, the presence of hydrocephalus, Chiari II malformation, scoliosis, kyphosis, the occurrence of spine and foot surgery, and ankle/foot abnormalities. Spinal surgeries included tethered cord release, posterior and anterior spinal instrumentation, in situ spinal fusion, and laminectomy. Ankle/foot abnormalities included varus, valgus, equinovarus, cavovarus, and claw toes. Ankle/foot surgeries included clubfoot repair, tendoachilles lengthening, tibialis anterior transfer, calcaneal osteotomy, and tibialis posterior release.

To assess ambulation, we used the Hoffer et al[7] classification for functional gait outcomes of clients with myelomeningocele: specifically community ambulator, household ambulator, nonfunctional ambulator, or nonambulator (full time wheelchair user). In the group of ambulators, the numbers of clients who used a reciprocating gait orthosis (RGO), knee-ankle-foot orthosis (KAFO), ankle-foot orthosis (AFO), and shoe inserts were identified.

After the collection of data, frequency counts were summarized for each of the variables listed above. The findings were then compared descriptively across the diagnostic categories appearing in the sample. In this article, counts of the number of cases are presented for each variable studied, and also summarized as percentages when there were at least 10 clients in the diagnostic category.

Interrater Reliability

The interrater reliability of the Tortori-Donati classification system was established using the *kappa* agreement analysis. A minimum *kappa* of 0.80 was set a priori as the level of agreement required for the primary reliability analysis, as an indicator of excellent interrater reliability.[8]

Each of the raters independently classified 34 health records that had been randomly selected from the total

TABLE 1

Tortori-Donati Cliniconeuroradiologic Classification of Spinal Dysraphism*

Closed with a Subcutaneous Mass		Closed Without a Subcutaneous Mass	
Lumbosacral	Cervical	Simple Dysraphic States	Complex Dysraphic States
Lipomyelomeningocele	Cervical myelocystocele	Posterior spina bifida	Dorsal enteric fistula
Lipomyeloschisis	Cervical myelomeningocele	**Intradural lipoma**	Neurenteric cysts
		Intramedullary lipoma	
Terminal myelocystocele	**Meningocele**	**Filum terminale lipoma**	**Split cord malformation** (diastematomyelia and diplomyelia)
Meningocele		Tight filum terminale	**Dermal sinus**
		Abnormally long spinal cord	**Caudal regression syndrome** (sacral agenesis)
		Persistent terminal ventricle	Segmental spinal dysgenesis

* Bolded categories are discussed in this article.

group of 104 charts by the third author. Using the same 34 health records, each rater identified independently the child's Tortori-Donati classification category and recorded this on a structured response sheet. The rater then noted on the response sheet whether she was certain of the rating for that child or whether she would confer with the other rater to come to consensus as to the appropriate grouping. This methodology was used to test interrater reliability since it was identical to the way in which the ratings had been done for the study itself. A stratified sampling approach was taken to make certain that each of the eight classification categories was represented by at least three charts. This process ensured that there would be entries within each of the cells of the [*kappa*] agreement analysis. The raters were unaware of the sampling procedure used, and thus did not have any expectations as to the number of charts that they would review in each category.

RESULTS

Interrater Reliability

The main interrater reliability analysis indicated that there was excellent agreement between the two raters ($kappa = 0.84$) on the Tortori-Donati classification system for the 23 charts on which they both indicated confidence in their independent ratings. When all 34 reliability sample charts were considered in a secondary reliability analysis, the agreement was moderate ($kappa = 0.67$). For six of the 11 charts that were identified as candidates for consensus rating with the other PT, the uncertainty related to whether the category should be an intradural lipoma or lipomyelomeningocele.

Tortori-Donati Classifications

After using the Tortori-Donati Classification System and reviewing the 104 charts, it was determined that eight categories were seen at this facility (Table 2). These eight categories are described below.

Closed NTD with a Subcutaneous Mass

Lipomyelomeningocele and meningocele are the two types of closed NTD from the lumbosacral category seen at this facility. *Lipomyelomeningocele* is the most common and severe of the closed NTD with a subcutaneous mass.[9,10] Lipomyelomeningocele is usually described as a fatty subcutaneous mass that passes through the open bifid vertebrae and attaches to the spinal cord. The lesion is not exposed to the air and there is no drainage of cerebral spinal fluid (CSF). The neural elements remain within the spinal canal and it is only the lipoma or fatty mass that protrudes through the vertebral defect to appear as a subcutaneous mass. Lipomyelomeningocele develops during the embryologic stage of neurulation. Children with lipomyelomeningocele usually present with varying degrees of lower extremity paralysis, decreased sensation and neurogenic bowel and bladder. Hydrocephalus and a Chiari II malformation are generally not associated with lipomyelomeningocele.[10]

A *meningocele* consists of a skin-covered sac filled with CSF that has herniated through a vertebral defect along with the meninges. This occurs most commonly in the lumbar or sacral area. There is not a clear differentiation between meningoceles and myelomeningoceles in the literature or when a diagnosis is applied; therefore, the true incidence of meningocele is difficult to determine.[5,11] Neurologic examination of a neonate with a meningocele is usually normal although neurologic problems can occur with growth.[5] Hydrocephalus can be associated with meningoceles but a Chiari II malformation rarely occurs.[11]

Cervical meningocele was the only type of NTD from the cervical category seen at this facility. They are rarely seen and poorly understood.[12] The lesion is covered with skin or a fibrous membrane and is associated with a vertebral defect. A thin stalk of neural tissue passes out of the spinal canal through a small fascial defect and ends along one side of the meningocele sac. Neurologic deficits depend on the extent of involvement of the posterior part of the spinal cord. Hydrocephalus and Chiari II malformation can be found in this population.[12]

Closed NTD Without a Subcutaneous Mass

Closed NTDs without subcutaneous masses can be divided into simple and complex dysraphic states. Simple dysraphic states (intradural and filum terminale lipoma) appear during the embryologic stages of primary and secondary neurulation.[6] The complex dysraphic states (split cord malformation, dermal sinus, and caudal regression syndrome) appear during gastrulation.[6] Children born with one of the complex dysraphic states usually present with more involved abnormalities, which may include other organ systems.

Simple Dysraphic States. *Intradural lipoma* is diagnosed when the fatty mass is entirely within the dura, or entirely intraspinal. Clinically this can cause varying degrees of neurologic involvement of the lower extremities, bowel and

TABLE 2

Number of Subjects in the Study Classified According to the Tortori-Donati Cliniconeuroradiologic Classification of Spinal Dysraphism

Closed with a Subcutaneous Mass		Closed Without a Subcutaneous Mass	
Lumbosacral	Cervical	Simple Dysraphic States	Complex Dysraphic States
Lipomyelomeningocele (63) Meningocele (9)	Meningocele (3)	Intradural lipoma (5) Filum terminale lipoma (7)	Split cord malformation (diastematomyelia and diplomyelia) (4) Dermal sinus (5) Caudal regression syndrome (sacral agenesis) (8)

All values inside parentheses indicate n values.

bladder. Children with an intradural lipoma are typically less involved than children with a lipomymelomeningocele.[5,11]

Filum terminale lipoma occurs when there is fibrolipomatous thickening of the filum terminale in association with a tethered cord.[6] As with intradural lipoma, this can result in various degrees of neurologic involvement of the lower extremities, bowel and bladder. These children are typically less involved than those with an intradural lipoma.[5]

Complex Dysraphic States. *Split cord malformation*, a complex dysraphic state, is divided into two categories: diplomyelia and diastematomyelia. Diplomyelia, the most common type, results when the two spinal cord segments are completely duplicated within a single dural sleeve with four sets of posterior and anterior nerve roots.[11] Diplomyelia is often associated with hydromyelia, tethered cord, and vertebral anomalies, such as butterfly vertebrae. Diastematomyelia, on the other hand, describes the malformation where the spinal cord is split into two hemicords, each contained within its own dural sleeve. Each neural tube has its own single set of posterior and anterior nerve roots.[11] Clinically, children usually present with scoliosis, tethered cord syndrome, and cutaneous birthmarks, such as hairy tufts, hemangioma, and dyschromic patches.[11,13] Vertebral anomalies are always present, and include block, butterfly, or hemivertebrae. Progressive deterioration can occur with age in orthopedic, urologic, and neurologic areas.[13]

A *dermal sinus* presents as a midline cutaneous pit or skin dimple above the intergluteal crease. This midline pit communicates with the subarachnoid space, and is therefore a portal for infections, meningitis, or spinal abscess. An epithelium-lined fistula extends inwards to the spinal cord or filum from the skin surface, and sometimes connects the CSF and its meningeal coating.[6] A dermal sinus is usually found in the lumbosacral area, but can be found anywhere from the intergluteal fold to the occiput. There may be cutaneous stigmata, including skin tags and hemangiomas. This altered disjunction allows a dermal sinus to run from the subcutaneous tissues to the intramedullary space. Children with a dermal sinus are treated surgically once the diagnosis is determined. These children usually have intact neurologic function at birth, but due to the possibility of tethered cord, may experience neurologic deterioration with age.

In *caudal regression syndrome,* the spectrum of abnormalities can range from agenesis of the coccyx to the absence of the sacral, lumbar, or thoracic vertebrae. The degree of involvement has been categorized from Type I, which is total sacral agenesis with some lumbar vertebrae missing, to Type V, which is the least severe with only coccygeal agenesis. Because the sacrum is typically the main region involved, the term sacral agenesis has been used synonymously with caudal agenesis or caudal regression.[14] Clinical presentation varies from major neurologic deficits associated with the lumbar or thoracic malformations to no symptoms when associated with isolated coccygeal agenesis. For those with extensive neurologic deficits, multiple visceral anomalies, such as anal imperforation, genital anomalies, bilateral renal dysplasia, and pulmonary hypoplasia can occur.

Tethered cord is often considered a malformation, but according to Tortori-Donati et al,[6] it is a clinical syndrome seen in the presentation of some of the closed NTD diagnoses, including lipomyelomeningocele, intradural lipoma, filum terminale lipoma, split cord malformation, and caudal regression syndrome. Tethered cord occurs as a result of stretching and distortion of the vessels as well as nerve fibers in the spinal cord, causing metabolic impairment. This results in the gradual deterioration of spinal function.[15] The signs and symptoms of a tethered cord can be present at birth or occur at any time in the development of the child.[15] Signs and symptoms include sensory changes and pain in the upper and lower extremities, muscle atrophy/weakness, spasticity, changes in gait, orthopedic deformities, scoliosis, and urinary incontinence.

Clinical Markers

Tables 3 to 5 present the data related to clinical markers and gait outcomes. Lipomyelomeningocele was the most common group of clients with closed NTD. The clients in the sample varied in age from 0.9 to 22.9 years with a median age of 10.4 years. The breakdowns of the specific

TABLE 3
The Incidence of Tethered Cord Release (TCR), Hydrocephalus, and Chiari II Malformation

Diagnosis	No. Cases	Median Age in years (Range)	Total TCR: % of Cases	Age of Tethered Cord Release: 0–6 mo: % of TCR	7–12 mo: % of TCR	1–2 y: % of TCR	>2 y: % of TCR	Repeat TCR % of Cases	Hydrocephalus % of Cases	Chiari II Malformation % of Cases
Lipomyelomeningocele	63	11.7 (1.4–22.9)	94 (59)	47 (28)	22 (13)	12 (7)	19 (11)	17 (10)	0	0
Meningocele	9	4.5 (1.2–17.2)	33 (3)	33 (1)	0	33 (1)	33 (1)	0	11 (1)	22 (2)
Caudal regression	8	5.8 (0.9–20.1	0	0	0	0	0	0	12 (1)	0
Filum terminale lipoma	7	10.5 (4.7- 22.6)	100 (7)	0	14 (1)	28 (2)	57 (4)	0	0	0
Intradural lipoma	5	6.5 (4.7 – 13.7)	100 (5)	40 (2)	40 (2)	0	20 (1)	0	0	0
Dermal sinus	5	9.8 (8.8–13.4)	40 (2)	50 (1)	0	0	50 (1)	0	0	0
Split cord malformation	4	10.4 (6.5–17.1)	100 (4)	25 (1)	0	0	75 (3)	0	0	0
Cervical meningocele	3	5.0 (0.9–8.8)	100 (3)	100 (3)	0	0	0	0	33 (1)	0

All values inside parentheses, excluding median age, indicate n values.

TABLE 4
The Incidence of Spinal and Ankle/Foot Abnormalities and Spine and Ankle/Foot Surgeries

Diagnosis	No. Cases	Scoliosis/Kyphosis %	Spine Surgery %	Ankle/Foot Abnormalities* %	Clubfoot Surgery %	Other Foot Surgery %†
Lipomyelomeningocele	63	28 (18)	11 (7)	73 (46)	19 (12)	21 (13)
Meningocele	9	11 (1)	0	22 (2)	0	0
Caudal regression	8	37 (3)	0	62 (5)	37 (3)	0
Filum terminale lipoma	7	43 (3)	14 (1)	57 (4)	14 (1)	14 (1)
Intradural lipoma	5	40 (2)	0	80 (4)	0	20 (1)
Dermal sinus	5	0	0	60 (3)	0	20 (1)
Split cord malformation	4	100 (4)	0	75 (3)	0	0
Cervical meningocele	3	0	0	33 (1)	0	0

All values inside parentheses indicate n values.
* Examples include: club foot, calcaneovarus, pes planus, planovalgus, equinus, calcaneus.
† Examples include: tendoachilles lengthening, tibialis anterior transfer, triple arthrodeses, calcaneal osteotomy, tibialis posterior release.

TABLE 5
The Incidence of Ambulation with and Without Orthoses

Diagnosis	No. Cases	Ambulators (% of Total Cases)	% of Ambulators Without Orthoses	% of Ambulators with Shoe Inserts	% of Ambulators with AFOs	% of Ambulators with KAFOs/RGOs	% Full-Time Wheelchair Users
Lipomyelomeningocele	63	100 (63)	67 (42)	2 (1)	25 (16)	6 (4)	0
Meningocele	9	100 (9)	67 (6)	0	33 (3)	0	0
Caudal regression	8	37 (3)	67 (2)	0	0	33 (1)	62 (5)
Filum terminale lipoma	7	100 (7)	57 (4)	0	28 (2)	14 (1)	0
Intradural lipoma	5	100 (5)	40 (2)	20 (1)	40 (2)	0	0
Dermal sinus	5	100 (5)	100 (5)	0	0	0	0
Split cord malformation	4	100 (4)	25 (1)	25 (1)	50 (2)	0	0
Cervical meningocele	3	100 (3)	67 (2)	0	33 (1)	0	0

All values inside parentheses indicate n values.
AFO indicates ankle-foot orthoses; KAFO, knee-ankle-foot orthoses; RGO, reciprocating gait orthoses.

types within the Tortori-Donati categories and ages of the children are provided in Table 3.

The frequency of tethered cord release, the age at which this surgery took place, and the need for a repeat tethered cord release are also summarized in Table 3. Of the 104 cases, 83 (80%) required a tethered cord release. The age at which the surgery took place varied across the age groups.

Hydrocephalus occurred in two of nine children with meningocele, one of eight children with caudal regression, and one of three children with cervical meningocele (Table 3). It did not occur in any of the other groups. The meningocele group was the only one to have Chiari II malformation, and this occurred in two of nine cases.

The frequency of scoliosis/kyphosis, ankle/foot abnormalities, and surgical intervention was examined (Table 4). Overall, scoliosis or kyphosis occurred in 30% of the children with no occurrence in the dermal sinus and cervical meningocele categories. For the occurrence of scoliosis/kyphosis in the other diagnostic categories, see Table 4.

Ankle/foot abnormalities were present in 65% of the children (Table 4). The results of the need for clubfoot and other types of foot surgery are also presented in Table 4. Clubfoot surgery occurred in only the lipomyelomeningocele, caudal regression, and filum terminale lipoma groups. Other types of foot surgery took place in the lipomyelomeningocele, intradural lipoma, dermal sinus, and filum terminale lipoma categories.

Gait Outcomes

According to Hoffer's classification,[7] 98 clients were community ambulators, and of these, 92 (93.9%) walked full time and did not use a wheelchair, whereas six used a wheelchair for long distances (Table 5). In the rest of the sample, one child was classified as a household ambulator and five children were classified as nonambulators. All of the children who were nonambulators were in the caudal regression category.

Few of the children who were ambulatory used assistive devices such as crutches or canes to walk. For example, of the 63 children in the lipomyelomeningocele category, one client used a walker, two clients used forearm crutches, and one used a quad cane. One client in each of the meningocele and caudal regression categories used a walker for ambulation while one client in the filum terminale category used forearm crutches to ambulate. Walkers, crutches, and canes were not used by children in any of the other four categories.

AFOs and shoe inserts were the types of orthoses most commonly used by the ambulatory clients (Table 5). KAFOs or RGOs were used by a small number of clients with lipomyelomeningocele (6.3%), caudal regression (one of eight clients) and filum terminale (one of seven clients). Of the three children in the caudal regression category who were able to walk, one used an RGO and walker for indoor ambulation (household ambulator), while the other two clients ambulated without any aids or orthoses. None of the five children in the dermal sinus category wore orthoses when ambulating.

DISCUSSION

Numerous articles describe spinal dysraphism or spina bifida as a single diagnosis and do not give information on specific subgroup diagnoses.[1] Although the determination of a clear diagnosis for children with NTD can be challenging, the use of magnetic resonance imaging better equips physicians for this task.[16] The application of a more specific classification system for closed NTD has the potential to assist clinicians with treatment planning as it lets the team differentiate and recognize the specific needs of each diagnostic group.[16] In our study, this classification system allowed detailed breakdowns of the incidence of tethered cord and its subsequent release, hydrocephalus, Chiari II malformation, scoliosis/kyphosis, ankle/foot abnormalities, and gait outcomes within subgroups.

The Tortori-Donati classification system was easily applied in a retrospective manner by the authors for about 70% of the health records reviewed, and demonstrated excellent interrater reliability for these records. For the other 30% of records, in particular for those children with either an intradural lipoma or lipomyelomeningocele, there was evidence of ambiguity related to the scoring and the need for further detailed review of the health record, as well as consultation with the other rater to finalize the rating. The clinical team will be encouraged in the future to use the Tortori-Donati system prospectively for classifying new referrals. This would permit the opportunity for dialogue with the individual referring the client, in particular to differentiate between intradural lipoma and lipomyelomeningocele.

Since this was a retrospective chart review in which clients were considered at a single point in time rather than longitudinally, the impact of differing age ranges in the various classification categories needs to be taken into account in the interpretation of the data. Under-representation of older children was of particular relevance in the categories with smaller sample sizes and this was due, as mentioned in the introduction, to the proportionally lower number of referrals for clients with closed NTD to this centre before 1995 (ie, the oldest children in the meningocele, intradural lipoma, dermal sinus, split cord malformation, and cervical meningocele categories were no more than 17 years of age).

Implications of Tethered Cord

Lipomyelomeningocele was seen at the clinic more often than any of the other diagnoses. Almost all of these clients required surgical intervention for a tethered spinal cord. The age at the time of the tethered cord repair varied, but almost half of the surgeries were done between birth and six months of age. These findings are in line with Sutton[10] and McLone et al[15] who recommended early tethered cord repairs as they felt that the likelihood of irreversible neurologic damage is best avoided by having surgical intervention at a young age.

In the filum terminale lipoma and split cord malformation categories, it was observed that clients more commonly undergo a tethered cord release after the age of two years. Given the limited numbers of older adolescents in our sample, the data may under-represent the occurrence of tethered cord releases through the later teenage years.

None of the clients in the caudal regression category had a tethered cord repair. Rossi et al[16] indicated that in the type I caudal regression group, tethered cord did not occur, while it did occur with the type II group. Although the specific type of caudal regression was not identified in the charts for the clients in our database, it is likely they were in the type I group.

In light of the common occurrence of tethered cord in the closed NTD groups, it is important for clinicians to be aware of the signs of deterioration in function as the child ages, which could signify a tethered cord. Indications may include pain in the back or legs, a change in gait pattern, decreased lower extremity strength, and increased lower extremity tone possibly resulting in changes in the shape of the foot.[15–17] Changes in bowel and bladder routines may also occur. Parental education about the signs of tethered cord is an important consideration as the parents will likely be the first to notice the changes in daily life.

A small number of clients with lipomyelomeningocele (17%) had repeat tethered cord repairs. This frequency was similar to the results of Colak et al[17] who found that 20% retethering occurred, and to Sutton[10] who reported repeat tethered cord occurrences in the 10% range. The age at which the 10 clients in our sample with lipomyelomeningocele required a repeat tethered cord release varied from four to 13 years, with a mean age of 7.8 years. Retethering was not seen with any of the other clients in our sample. McLone et al[15] indicated that retethering is not seen in the dermal sinus category with good long-term preservation of the neurologic status once the initial repair is done. However, the lack of older adolescents in our other categories may have contributed to the absence of repeat tethered cord releases.

Other Neurologic and Spinal Implications

Sutton's work[10] supports our findings that hydrocephalus and Chiari II malformation are rare occurrences in the lipomyelomeningocele category. In our study, hydrocephalus or Chiari II malformation only occurred in the meningocele, cervical meningocele, and caudal regression groups. Steinbok[12] also found an association of Chiari II malformation with closed neural tube defects, specifically myelocystoceles and meningoceles. The presence of hydrocephalus in our sample of children with caudal regression was similar to the findings of Pang.[14]

Scoliosis and kyphosis were seen in all groups with the exception of dermal sinus and cervical meningocele. Pang[14] noted that 30% of children with caudal regression syndrome over two years of age had scoliosis. This was similar to our findings (three of eight children). Surgical correction was reported in only a few cases in our lipomyelomeningocele and filum terminale lipoma categories.

Both scoliosis and spine surgery can occur across the ages, and so for this reason, our data may not have captured those children who had not yet gone on to develop scoliosis or who might require surgery as they age.

Consequences of Ankle or Foot Abnormalities

The incidence of ankle or foot abnormalities was high across all categories with the exception of clients with meningocele and cervical meningocele. In the lipomyelomeningocele group, 46 of 63 children had some type of ankle or foot abnormality. These abnormalities were diverse and included varus, valgus, cavus, equinus, calcaneus, equinovarus, calcaneovalgus, pes planus, and planovalgus. The most frequent deformity in the lipomyelomeningocele category was equinovarus (20%), followed by cavovarus (17%). Sutton[10] also found that the most frequent orthopedic deformity in children with lipomyelomeningocele was cavovarus deformity of the foot with or without leg length discrepancy. Pang[14] noted that children with caudal regression had severe deformities in the ankle region including equinovarus and calcaneovalgus. These are similar to the types of severe deformities found in our sample.

Despite the large number of ankle or foot abnormalities in the whole sample, relatively few clients underwent foot surgery. Approximately one in five of our clients with lipomyelomeningocele required clubfoot surgery, and this usually took place during the infant or toddler years. As well, one in five went on to undergo other foot surgeries such as tendoachilles lengthening, tibialis anterior transfer, calcaneal osteotomy, and triple arthrodesis, and these other types of foot surgery occurred across the age spectrum. Our data may not have captured all clients who will require surgery in later adolescence.

Ambulation

As mentioned previously, almost all of the clients with closed NTDs were ambulators and two thirds of these children did not require any orthoses to ambulate. The philosophy in our clinic regarding orthotic management is to use the least amount of support required to ensure the most functional gait. AFOs were the most common orthoses, and were worn by more than two thirds of the children who required orthoses. The small number of clients who used KAFOs did so with the knee joints locked.

Gait outcomes for children with closed NTDs have not previously been reported in the literature; however, the results of our study show that clients with closed NTDs are likely to be full-time ambulators. The under-representation of older adolescents in several of the classification categories may have influenced our results related to ambulation. For example, ankle or foot abnormalities may progress through adolescence, meaning that clients may ultimately require more extensive orthoses or foot surgery. Spine surgery can also take place at any age. Any of these orthopedic issues can affect gait abilities.

The classification system by Hoffer et al[7] was developed for clients with open NTDs (eg, myelomeningocele) and the "community ambulator" category includes individuals who use a wheelchair for long distances. Ninety-two of the 104 clients in our sample had high levels of ambulatory ability, and did not require a wheelchair when out in the community. However, the Hoffer et al's classification system did not allow recognition of this advanced capability.

Clinical PT Management

The PT management of clients with closed NTD could be enhanced if there is an understanding of the diagnostic categories within NTD. The lipomyelomeningocele category is likely to be the largest closed NTD group that the clinician will see in the community. Our results showed that all were ambulators, despite some type of ankle or foot abnormality in 73% of these children. Almost one-third wore some type of orthotic device. This evidence will help the clinicians who are working with infants and young children with lipomyelomeningocele to project their likely mobility status as well as to plan interventions such as gait training, orthotic management, skin management, gross motor skills, high-level balance activities, and endurance activities. Given the incidence of scoliosis or kyphosis and spine surgery in this category, the PT needs to be aware of potential changes to the shape of the spine during the child's development, in particular during puberty. There is also the potential that the child with lipomyelomeningocele may require some type of foot surgery sometime during their development to optimize their function. The PT might then become involved with postoperative stretching, strengthening, and gait training.

In examining the data for the dermal sinus category, it was noted that all five of the clients were ambulators and three had ankle or foot abnormalities. Unlike the lipomyelomeningocele category, none of these children required an orthosis to walk. As well, none had scoliosis, kyphosis or underwent spine surgery. Despite having very few cases to examine, it does appear that these children are high functioning, and therefore, can be encouraged to participate in leisure, sport and endurance activities. As discussed at the beginning of this article, there is a possibility of tethered cord for clients with dermal sinus. The PT should be prepared that neurologic deterioration may occur with age and should regularly screen for signs of such deterioration.

In contrast, the majority of clients in the caudal regression category were nonambulators. Therefore, PTs will need to provide upper extremity strengthening as well as be involved in wheelchair prescription and skills training. One client in this category used an RGO for walking short distances. This was the only RGO used by any of the clients with closed NTD in our sample. A PT working with this group should be familiar with this more complex type of orthoses.

In the split cord malformation category, all of the children had scoliosis due to their vertebral anomalies. This is of significance for the clinician, as they should be monitoring these clients closely for scoliosis, particularly around the time of growth spurts.

Limitations

In addition to the limitations previously noted with respect to the retrospective nature of this study, the other key limitation was the small sample size that did not allow inferential statistical analyses for the various NTD category comparisons. A further study with more subjects and prospective data collection for the variables of interest would enable a more definitive assessment of status and outcomes in the various closed NTD categories.

CONCLUSIONS

The use of the Tortori-Donati et al classification has provided the clinical team at Bloorview Kids Rehab with a clearer framework for categorization of closed NTD and enhanced our knowledge of this complex group of diagnoses. Most importantly, it has given a greater understanding of associated neurologic, orthopedic, and gait outcomes that can be used to assist clinicians in clinical management of these children.

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