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Clinical Study

Spinal cord detethering procedures in children: A 5 year retrospective cohort study of the early post-operative course

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ABSTRACT

Tethered spinal cord can cause neurological, orthopaedic and sphincteric problems in children and detethering surgery may prevent or reverse these problems. This 5 year retrospective cohort study aimed to review our experience of detethering surgery at The Children's Hospital at Westmead, Sydney, Australia, particularly examining the early post-operative complications of this procedure. Between 2007 and 2012, 61 children underwent 63 detethering procedures. The median age at detethering surgery was 1.4 years old (interquartile range: 0.7–5.6 years). Fifty-five children (90.1%) had lumbosacral procedures, 31 (50.8%) were asymptomatic from tethering, 11 (18.0%) had motor or gait disturbance, 11 (18.0%) sphincteric disturbance, eight (13.1%) lower limb orthopaedic deformities, eight (13.1%) scoliosis, six (9.8%) back or leg pain and two (3.3%) sensory disturbance. The most common tethering pathologies were spinal lipomas in 32 children (52.5%), filum abnormalities in 23 (37.7%), dorsal sinus tracts in eight (13.1%) and diastematomyelia in seven (11.5%). Twenty-six children (42.6%) had either a syrinx or central canal dilatation preoperatively. The most common complications were wound infection and cerebrospinal fluid leak. Six children (9.8%) required reoperation for wound issues and two patients (3.3%) required subsequent reoperation for cord retethering during the study period. There were no deaths and no new neurological deficits. Of the children with the above preoperative deficits, 26.7% were documented to have improvement or resolution of their symptoms post-operatively. The highest rate of improvement occurred in children with motor or gait disturbance (36.4%) or sphincteric disturbance (27.3%).

1. Introduction

This study aimed to review the experience of detethering procedures and complication rates at The Children's Hospital at Westmead. The anchoring of the spinal cord to the vertebral column by inelastic tissue attachments causes tethered spinal cord (TSC). Most commonly, tethering occurs in the lower thoracic, lumbar or sacral spine. The causes of tethering in children can be roughly defined into three different categories: post-myelomeningocoele and post-meningocoele repair, occult spinal dysraphism (OSD) and sacral agenesis. In children with post-myelomeningocoele or meningocoele repair the etiology of tethering is usually fibrous adhesions. Often these children already have significant, chronic neurological deficits not due to tethering, however, when progressive deficits occur, exploration and detethering can be of value. OSD encompasses a large number of tethering pathologies. These include tight, thickened or fatty filum terminale, split cord malformations (diastematomyelia), non-filum spinal lipomas (such as lipomyelomeningocoele), dermoid tracts, cysts and tumours, atretic meningocoele, meningocoele manqué and neurenteric cysts. The earliest manifestation of OSD can be cutaneous with hypertrichosis, capillary hemangiomas, lumbosacral appendages, dimples and lipomas leading to the diagnosis in asymptomatic children. Babies with undiagnosed dermal sinus tracts can occasionally present with meningitis from their connection with the thecal sac. Sometimes OSD related TSC is only detected after problems develop, such as during investigation of scoliosis, incidentally, or is not detected until adulthood. Sacral agenesis can be present on its own or related to a syndrome. Syndromes associated with agenesis and tethering include the OEIS syndrome (omphalocoele, exstrophy of cloaca, imperforate anus and spinal defects), VATER syndrome (vertebrae, anus, trachea, oesophagus, and renal), Sirenoid malformation and the Currarino triad. Associated external manifestations can include stenotic or imperforate anus, rectovaginal or rectourethral fistulae or a persistent cloaca with common urinary, genital and intestinal outflow, although these are not necessarily present.

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Tethered cord syndrome (TCS) is the progressive development of neurological, sphincteric or orthopaedic dysfunction due to tethering. Neurological manifestations of TCS include upper and/or lower motor neuron weakness, pain, sensory loss, wasting and gait disturbance, orthopaedic manifestations including cavovarus deformities, claw toes, foot asymmetry, leg length discrepancy and scoliosis, sphincteric manifestations including urinary or faecal incontinence, neurogenic bladder and/or frequent urinary infections. TCS may be due to stretching of the spinal cord during vertebral column growth, flexion movements of the spine on an already stretched cord, or both.

Detethering surgery usually involves a relatively conservative operation which involves a one or two level laminectomy and division of the filum and adhesions and/or debulking or excision of tethering pathologies such as lipomas. Sometimes this can be achieved via preexisting posterior bony defects or limited laminotomies. Some authors have proposed other approaches in the literature, including mini-open using expandable tubular retraction systems [1] or duraplasties to reduce the rate of retethering [2]. In adults, for difficult cases of recurrent symptomatic TCS despite multiple detethering procedures, instrumented posterior vertebral column reduction procedures at the thoracolumbar junction have been proposed as a potential option and authors report some success in reducing symptoms in these cases [3–5]. Intraoperative neurophysiological monitoring of the lower lumbar and sacral nerve roots can be performed in cases where the anatomy is difficult to distinguish [6].

2. Methods

This study was designed as a retrospective cohort study. To identify children from 2007–2012, neurosurgical operation lists and World Health Organization International Classification of Diseases-10 (ICD-10) codes relating to spinal cord detethering were searched. Children were eligible if they had a diagnosis of spinal cord tethering and underwent detethering surgery. We excluded children who primarily underwent myelomeningocoele or meningocoele repairs as the purpose of this surgery is generally not detethering, and excluded patients undergoing detethering after these repairs as we felt they represented a different subset of patients. Information was gathered from medical records regarding preoperative neurological status, cause of spinal cord tethering (for example, lipomyelomeningocoele or sometimes multiple pathologies), imaging findings, perioperative and early post-operative complications. Neurological, orthopaedic or sphincteric improvement was obtained from available notes, usually from the treating neurosurgeon, spina bifida service, physiotherapist or orthopaedic surgeon. Ethics and site specific approval was obtained from the Sydney Children's Hospital Network Human Research Ethics Committee.

3. Results

Sixty-one children were identified, 59 of whom had their first detethering surgery performed in the study period and two who were readmitted for repeat detethering surgery (Table 1). Twenty-eight patients were boys and 33 patients were girls. The median age at detethering surgery was 1.4 years old (interquartile range: 0.7–6.3 years). Fifty-five children (90.2%) had lumbosacral procedures, four (6.6%) thoracolumbar procedures and two (3.3%) cervical procedures. The most common tethering pathologies were spinal lipomas in 32 children (52.5%), filum abnormalities in 23 (37.7%), dorsal sinus tracts in eight (13.1%) and diastematomyelia in seven (11.5%). The conus was low lying (L3 or below) in 41 children (67.2%). Twenty-three children (37.7%) had either a syrinx or central canal dilatation preoperatively. Syrinxes occurred most frequently between the T11 and L2 vertebral levels (Fig. 1). Thirty-one children (50.8%) were asymptomatic from tethering, 11 (18.0%) had motor or gait disturbance, 11 (18.0%) sphincteric disturbance, eight (13.1%) lower limb orthopaedic deformities, eight (13.1%) scoliosis, six (9.8%) back or leg pain and two (3.3%) sensory disturbance (Table 2).

Table 1
Demographics of children who underwent spinal cord detethering procedures

Study variable	Number (n, %)
Demographics	
Male	28, 45.9
Female	33, 54.1
Median age	1.4 years
Age range	10 days–16.1 years
Level of procedure	
Lumbosacral	55, 90.1
Thoracolumbar	4, 6.6
Cervical	2, 3.3
Tethering pathologies	
Spinal lipoma	32, 52.5*
Lipomyelomeningocoele	20, 32.8
Filum abnormalities	23, 37.7*
Tight filum	9, 14.8
Thickened filum	8, 13.1
Fatty filum/lipoma	8, 13.1
Dorsal sinus tract	8, 13.1
Diastematomyelia	7, 11.5
Type 1 diastematomyelia	3, 4.9
Type 2 diastematomyelia	4, 6.6
Atretic meningocoele	5, 8.2
Dermoid, epidermoid, neurenteric cyst, teratoma	5, 8.2
Caudal regression	2, 3.3
Other**	2, 3.3
Other imaging findings	
Low lying conus (L3 or lower)	41, 67.2
Syrinx	21, 34.4
Central canal dilatation without syrinx	8, 13.1

* Spinal lipomas include Fatty filum/lipoma.
** Other pathologies are dynamic cord tethering, syndromic related causes (eg. Currarino Triad), vertebral dislocation causing tethering.

Sixty-three detethering procedures were performed, four of which were redo operations. The most common complications were wound infection and cerebrospinal fluid (CSF) leak (Table 3). Eight detethering procedures (12.7%) were complicated by wound infection, three (4.8%) by CSF leak and two (3.2%) by both wound infection and CSF leak. Six detethering procedures (9.5%) required at least one return to theatre for wound repair or CSF diversion. The majority of wound infections (75%) were able to be managed conservatively without return to theatre, however the majority of CSF leaks (67%) required return to theatre and all children with both wound infection and CSF leak required return to theatre (Table 4). Two children had allergic reactions to

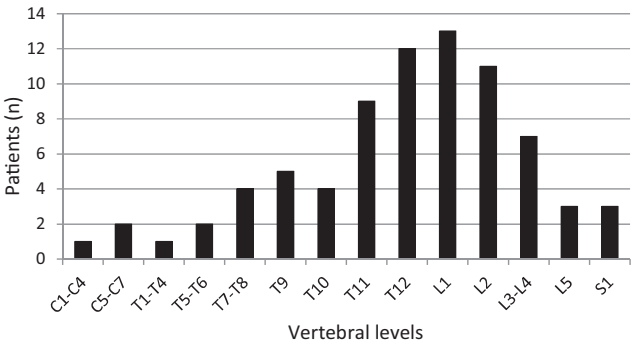

Fig. 1. Number of patients with syrinx at certain vertebral levels.