

Impact of tethered cord release on symptoms of Chiari II malformation in children born with a myelomeningocele

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Abstract

Purpose The role of distal traction in the form of a tethered spinal cord in exacerbating anatomical findings or symptoms of Chiari II malformation (CIIM) has been debated for decades. Despite the association of Chiari II malformation with myelomeningocele, the impact of tethered cord release on CIIM symptoms in patients has not been explored.

Methods A retrospective review of 59 patients born with a myelomeningocele was performed. A total of 92 untethering procedures were performed in which symptoms of CIIM were present in 29 cases. In 57 out of 92 cases, the patients did not have symptoms of CIIM prior to untethering. Six cases were excluded because cervicomedullary decompression was performed prior to untethering. The response of CIIM symptoms, syrinx size, and cerebellar tonsil position were examined before and after spinal cord untethering.

Results Forty-four characteristic signs and symptoms of CIIM were present prior to 29 untetherings. Thirty-three of 44 (75%) symptoms improved following spinal cord untethering, though no symptom resolved completely. Syrinx size and cerebellar tonsil position were unchanged following untethering.

Conclusion The authors conclude that mild to moderate symptoms of CIIM may respond positively to spinal cord untethering, potentially by normalization cerebrospinal fluid flow dynamics. Symptom improvement occurs despite the lack of radiographic evidence of CIIM resolution.

Keywords Myelomeningocele · Tethered spinal cord · Chiari malformation · Pediatric · Outcome

Introduction

Nearly all children born with a myelomeningocele will have radiographic evidence of Chiari II malformation (CIIM) [2, 5], though only 20–30% will have symptoms related to lower cranial nerve and brainstem dysfunction [13]. In these children, these symptoms are the major determinant of quality of life and are the major cause of death [3, 7, 9]. Many theories of the pathoanatomic relationship between spinal cord tethering and CIIM have been proposed, including the traction theory, which many regard as disproved. This theory states that the vermian and brainstem herniation that occurs in CIIM is a result of caudal traction of the hindbrain by a tethered spinal cord [8, 14]. This theory as the *causative* mechanism of CIIM has been challenged with evidence in animals [6] and cadavers [16], but its role in potentially exacerbating or relieving symptoms of CIIM has not yet been examined.

More recent reports have shown dramatic improvements in the degree of hindbrain herniation with intrauterine myelomeningocele repair, raising new questions regarding the role of distal traction in the symptomatology of CIIM. This has led many to postulate that relief of distal tethering has some decompressive effect at the cervicomedullary junction, despite the fact that symptom response to untethering has not previously been explored. In this retrospective analysis, we evaluate the symptomatic and radiographic response of Chiari malformation type II to spinal cord untethering in 55 children born with a myelomeningocele who developed symptomatic spinal cord tethering.

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Materials and methods

Approval for this retrospective analysis was obtained from the Johns Hopkins Hospital (IRB #00016159). A total of 59 patients who underwent spinal cord untethering following myelomeningocele repair were identified from 119 patients who underwent tethered cord release due to any TCS etiology from 1997 to 2008. The medical records of these patients were reviewed, which included all hospital records, pre- and post-operative clinic notes and pre- and post-operative imaging studies. Relevant variables recorded included patient demographics, presenting symptoms, and neurological deficits occurring immediately prior to and after spinal cord untethering, which always extended beyond the section of the filum terminale. Signs and symptoms recorded included swallowing difficulties, poor feeding, cyanosis during feeding, nasal regurgitation, prolonged feeding time, poor oral secretion, decreased gag reflex, apneic spells, stridor, aspiration, arm numbness, arm weakness, upper extremity myelopathy, upper extremity coordination difficulty, loss of temperature sensation, “cape” pattern sensory loss, opisthotonos, nystagmus, weak or absent cry, facial weakness, facial paresthesias, and headache of a suboccipital or cervical variety. Symptom status was judged to be improved, maintained, or worsened as compared to the immediate pre-operative status. The assessment of signs before and after each un-tethering by the attending physician was recorded. The status of the post-operative CIIM symptoms was assessed at the first post-operative clinic visit, which occurred 3–6 weeks after surgery. Symptoms occurring in the setting of a suspected or confirmed shunt malfunction were not recorded. Children who underwent cervicomedullary decompression (CMD) for severe CIIM symptoms prior to tethered cord release were excluded to isolate the effect of untethering on symptoms. In patients who underwent CMD after tethered cord release, symptom status was recorded immediately prior to decompression. All patients were evaluated for the presence of a syrinx involving the cervical spinal cord. The presence and size of a cervical syrinx was recorded from the radiology reports and the status of the syrinx following tethered cord release was evaluated. The extent of cerebellar tonsil extension into the foramen magnum before and after surgery was also recorded.

Results

A total of 119 children underwent spinal cord untethering between 1990 and 2007, of which 59 had myelomeningocele as a tethering etiology. There were 17 (28.8%) males and 42 (71.2%) females with an average \pm SD age of 8.92 \pm 4.44 years (range, 2–18). The average time from myelome-

ningocele repair to index untethering was 6.39 years. Six patients were excluded from the analysis as multiple instances of shunt malfunction and revision did not allow for a proper evaluation of CIIM symptoms, tethering, and the response to tethered cord release. A total of 92 untetherings were performed in these 59 patients (average of 1.6 untetherings per patient) for progressive symptoms of spinal cord tethering, including bowel/bladder dysfunction, motor and sensory deficits, or pain. Symptoms of CIIM were present prior to untethering in 29/92 cases. Six out of 29 cases were excluded from analysis because CMD for severe symptoms of brainstem and lower cranial nerve dysfunction occurred prior to untethering. These symptoms included progressive gagging, headache, snoring, decrease in upper extremity strength, episodes of desaturation, and rapidly progressing syrinx. In 57 out of 92 cases, there were no symptoms of CIIM prior to tethering cord release, but these cases were included in the pre- and post-untethering radiographic analysis. A total of seven children underwent CMD for progressive CIIM symptoms following tethered cord release, at a mean of 11.2 months following the most recent untethering.

A total of 44 symptoms of Chiari II malformation were present prior to 29 un-tethering operations. Post-operatively, 33 out of 44 symptoms were improved, eight out of 44 were maintained and three out of 44 were worsened. Of the 33 symptoms that improved, all had some persistent degree of symptoms with no symptoms being completely resolved. The most common symptoms were headache, nystagmus, and upper extremity sensory deficit, which showed a 69%, 66%, and 100% response to untethering, respectively. All three cases in which symptoms were worse following untethering required subsequent CMD. A complete list of symptom responses is provided in Table 1. A syrinx involving the cervical spinal cord was present in 13/92 cases, with an average pre-operative size

Table 1 Summary of Chiari II malformation symptom presentation and responses to spinal cord untethering

	Number	Improvement	Worsened	Same
Upper extremity				
Sensory deficit	6	6	0	0
Weakness	4	3	0	1
Bulbar features				
Difficulty swallowing	2	2	0	0
Facial weakness	1	1	0	0
Poor feeding	4	3	0	1
Apnea	3	2	0	1
Nystagmus	11	7	1	3
Headache (cervical/ suboccipital)	13	9	2	2
Total	44	33 (75%)	3 (6.8%)	8 (18%)

of 3.70 mm. Following un-tethering, syrinx persisted in all cases with an average size of 3.57 mm, which was not significantly changed from the pre-operative size ($p=0.24$). The average extension of the cerebellar tonsils below the level of the foramen magnum pre-operatively was 5.73 mm (range, 2.0–7.6 mm). Post-operatively, the extension was 5.46 mm, which was not a statistically significant difference from the pre-operative value ($p=0.43$).

Discussion

Forty-four symptoms of CIIM were present prior to 29 untetherings, of which 33 (75%) improved after tethered cord release. Eight (18%) did not change and three (7%) worsened after surgery. No patients experienced complete relief of CIIM symptoms following tethered cord release. There was no observed change in syrinx size or any cranial–caudal position change of the cerebellar tonsils as a result of untethering.

The long known association between spinal cord tethering and Chiari malformation led many early investigators to attempt to find a causal link between the two disorders. Penfield and Coburn [14], and later Lichtenstein [8], were the first to suggest that the cerebellum and medulla might be pulled into the cervical canal as a result of distal tethering occurring at the site of the myelomeningocele. Many years later, a significantly more eloquent and elaborate theory based on the lack of distention in the embryonic ventricular system was proposed which accounted not only for the cerebellar herniation but the myriad of other malformations observed in CIIM [10]. This theory was more recently supported by evidence that restoring the normal ventricular pressure by intrauterine myelomeningocele closure could dramatically resolve hind-brain herniation and even completely correct the Chiari malformation [17].

Evidence from anatomic experiments in animals and cadavers provided additional evidence against the traction theory as the *causative* mechanism of CIIM in myelomeningocele patients. Early work by Goldstein and Kepes in rats demonstrated that caudal traction at the distal spinal cord dissipated within the 3–4 proximal lumbar segments and did not produce any cervicomedullary herniation [6]. This traction also did not produce the medullary kink or vermian peg that is characteristic of CIIM. Tubbs et al. later demonstrated in cadavers that distal cord tension produced no caudal movement of the cerebellar tonsils [16]. Neither of these experiments examined cerebrospinal fluid (CSF) flow before or after traction was applied. While the evidence for the unified theory of CIIM and against traction theory are available, it is important to recognize that they do not completely discount distal traction contributing to CIIM-related symptoms later in life.

In a large series describing late neurological deterioration in this population, an experienced group of authors describe associated findings at the time of symptomatic tethered cord presentation. In 26/26 patients, evidence of Chiari II malformation was present at the time of late neurological deterioration, with two patients complaining primarily of upper extremity weakness, which resolved completely with posterior fossa decompression [4]. The association of higher spinal cord abnormalities with spinal cord tethering led them to suggest “that a spinal cord dysfunction related to tethering phenomena may be revealed or decompensated by ‘distal’ pathological condition, the correction of which may lead to an improvement in the clinical picture, rather than an operation aimed at untethering the spinal cord [4].”

Isolated cases of acquired Chiari malformation in the setting of spinal tethering have been reported, suggesting that events occurring later in life at the distal spinal cord may contribute to this pathology. Waldau et al. reported on a young girl who developed acquired Chiari malformation with a large cervical syrinx due to an untreated lipomyelomeningocele [19]. Similarly, Abel et al. reported on a young girl who acquired Chiari malformation due to a fatty terminal filum [1]. The child in their report had magnetic resonance (MR) imaging prior to the onset of cerebellar tonsil herniation documenting the absence of any congenital abnormality. Interestingly, both of these children were asymptomatic as a result of their acquired deformity. Further implicating spinal cord tethering with symptomatic Chiari malformations is the report by Ng and Seow who describe a lower thoracic (lumbar) syrinx developing after the onset of a tethered cord [12]. With cord untethering, the syrinx resolved completely, suggesting that improved CSF flow dynamics occurs as a result of the removal of the distal cord traction; however, it should be noted that we did not appreciate any change in cervical syrinx in this study. The finding of a higher than expected incidence of CIM with normal posterior cranial fossa volume in patients with a lipomyelomeningocele might also suggest that events at the distal cord contribute to brainstem compression [15].

A particular challenge to understanding symptom improvement in this series is the absence of any radiographic improvement following untethering; however, prior studies have shown that anatomy may not necessarily dictate symptoms as precisely as previously thought. In a large review of 73 children with CIIM, Narayan et al. observed that neither presenting symptoms nor improvement after surgical decompression were related to the initial level of the cervicomedullary deformity [11]. Based on these results, they concluded that “the cervicomedullary deformity is not a reliable marker to determine which patients may become symptomatic and require decompression or to determine prognosis.” In evaluating their experience with

surgical decompression for symptomatic CIIM in children born with a myelomeningocele, Vandertop et al. reflected on their experience and commented that “the fact that most patients develop symptoms that are not present at birth supports the concept that intrinsic abnormalities do not play a role in the pathogenesis of symptomatic Chiari II malformation” [18]. They went on to suggest that compressive forces causing a secondary neuronal destruction was most important for the development of symptomatic CIIM. These combined observations allow for theories other than the uniform theory to explain symptomatic CIIM developing later in life.

In an attempt to explain the findings in this report, we theorize that a tethered cord plays a role in CSF dynamics and that even subtle improvement in CSF flow can relieve symptoms in patients with CIIM. Further investigation, including pre- and post-operative MR CSF flow studies would be helpful in substantiating such a claim. Although spinal cord tethering is likely not the direct cause of CIIM, these data demonstrate that tethered cord release is associated with improvement in symptoms of Chiari malformation.

Conclusion

Mild to moderate symptoms of CIIM may respond positively to spinal cord untethering. Radiographic evidence of improvement is often not apparent following untethering as the malformation is thought to be a congenital, rather than an acquired, anatomic deformity. Despite our findings of CIIM symptom responses, tethered cord release should not be considered the primary treatment for a severely symptomatic Chiari malformation, where CMD or VP shunt revision may be required to maintain neurologic status and preserve quality of life.

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