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RESEARCH

Recurrent tethered cord: outcome and follow-up of 20 de-thetering for symptomatic spina bifida: choort study

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Abstract

Objective The objective of this study was to evaluate the prevalence of tethered cord among patients in the postoperative period of open and occult spina bifida. To identify warning signs for its early diagnosis, as well as outcomes after the new surgical approach.

Methods Retrospective study of patients followed at the Pediatric Neurosurgery Department of the Federal University of São Paulo with spinal dysraphism. Signs and symptoms indicating reoperation were collected, and postoperative results were classified as improved, unchanged, or worsened.

Results 222 medical records of patients diagnosed with spinal dysraphism were evaluated. Symptomatic Tethered Cord Syndrome (STCS) was identified in 30 patients (13.51%), with clinical manifestations related to orthopedic deformities (66.7%), neurological deficits (56.7%), urological dysfunction (50%), and intestinal dysfunction (40%). 20 cases underwent surgery for tethered cord release. The mean age at the time of surgery was 7.7 ± 4.9 years, with 13 female patients (65%). In the postoperative evaluation, improvement in low back pain (90.9%), urological pattern, and urinary tract infection episodes (45.4%) were particularly noteworthy. 3 patients (33.3%) with constipation showed improvement, and one worsened (11.1%). Improvement in ambulation was seen in two cases (16.7%). Low back pain was the first symptom to improve after surgery, with an average time of 1.3 months, followed by changes in the urological pattern at 15.6 months. Improvement in constipation was observed in the first month in 2 cases (66.7%), positive changes in ambulation were observed around 7 months after surgery, and only one case showed improvement in clubfoot correction.

Conclusions The prevalence of tethered cord recurrence after primary correction surgery for open or occult neural tube closure defects was similar to that found in the literature. The results were encouraging, with good postoperative evolution of patients, especially in the improvement of low back pain and urological symptoms.

Keywords Neurogenic bladder · Symptomatic tethered cord syndrome · Spinal dysraphisms

Introduction

The most common neurological malformations in the spinal cord, also called spinal dysraphisms, result from the failure of fusion of the neural arches of the vertebrae that begins in the fourth week after conception [1]. In this sense, neural tube defects can also compromise adjacent tissues, such as meninges, muscles, and skin, and manifest themselves in an evident or even silent manner [2]. Therefore, spinal dysraphisms are divided into open and occult, with myelomeningocele (MMC)

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being the major representative of open defects, where there is protrusion and exposure of neural tissue in the lumbosacral region, while for occult defects, such as lipomyelomeningocele, the protrusion of neural tissue is covered with intact skin, with or without any stigma [1]. The treatment for these congenital anomalies is surgical, and it is performed as early as possible with the aim of minimizing infection and associated neurological deficits, such as alterations in the musculoskeletal system, genitourinary and gastrointestinal systems, and the presence of low back pain [3].

Symptomatic Tethered Cord Syndrome (STCS) is one of the main complications after primary surgical repair of spinal dysraphisms, such as MMC and lipomas, because the healing process promotes adhesion of the spinal cord to adjacent structures, and thus, during children's growth, symptoms related to it may arise, such as neurological deficits and sphincter dysfunction, as well as lower limb deformity and low back pain [4].

Magnetic resonance imaging (MRI) is the gold standard for detecting spinal dysraphisms, especially for occult dysraphisms [5], being useful to confirm low-lying conus medullaris, defined as the medullary cone below the L2-L3 level. However, low-lying conus medullaris on MRI is not synonymous with reoperation of the lipoma or MMC, since, due to the nonspecificity of radiological examinations, the diagnosis of STCS is eminently clinical, based on the recurrence or progression of symptoms suggestive of tethered cord [6]. Furthermore, due to adhesion to adjacent structures, there is the possibility of new deficits emerging, as well as the regression of previously acquired skills, such as walking without support [4].

In this sense, bladder dysfunction is one of the most common symptoms related to the condition and can present with various manifestations, from urinary incontinence to hypoactivity of the detrusor muscle of the bladder, promoting urine retention, such urological pattern favors episodes of urinary infection and vesicoureteral reflux [7]. The management of these symptoms is extremely important for the protection of renal function and social quality of life. Thus, understanding these signs enables an early and adequate diagnosis and surgical treatment, as the associated symptoms have a high impact on daily activities and the well-being of patients. In this study, we evaluated the criteria for surgical indication for reoperation of patients with operated spinal dysraphisms, after birth, i.e., myelomeningocele or lipomas, and the short- and long-term results.

Methods

Retrospective data from patients treated at the outpatient Pediatric Neurosurgery Department of the Paulista School of Medicine/UNIFESP with a diagnosis of neural tube closure defects from 2018 to 2023 were evaluated.

Data related to symptoms and neurological deficits immediately before and immediately after surgery were collected based on clinical notes from outpatient follow-up. The diagnosis of STCS was considered in the worsening or onset of symptoms related to urinary, intestinal, and/or motor dysfunction, pain at the level and below the lesion, and sensory deficits. Clinical complaints were considered even in the absence of radiological findings.

Changes in symptoms were assessed in the immediate postoperative period and during regular outpatient follow-up of patients and classified as improved, unchanged, or worsened compared to the last preoperative consultation.

All patients in this study have neurogenic bladder and were categorized according to urine loss between relief catheterizations; thus, patients who perform intermittent catheterization but do not have urine loss between catheterizations were named socially continent, while those who use diapers are intermittently continent.

The chi-square test was used for qualitative variables, with Fisher's exact test used when the expected frequency < 5. A p-value < 0.05 was considered significant.

Results

Among the 222 patients with neural tube closure defects treated at the outpatient clinic of this institution from 2018, even those who were previously operated on, 30 were diagnosed with symptomatic tethered cord syndrome (STCS) (13.51%), and 20 patients underwent a new surgical approach (Table 1). The other ten symptomatic patients are awaiting availability of hospital resources for the procedure (waiting list for elective surgeries).

The average age at the time of surgery was 7.7 ± 4.9 years with a median of 6.5 years, with 13 female patients (65%). The causes of tethering included previous repair of myelomeningocele in 14 patients (70%), spinal lipoma (25%), and filum lipomatosis (5%).

The most common clinical manifestations present in the 30 patients indicated for surgical correction included orthopedic deformities (66.7%), neurological deficits (56.7%), urological dysfunction (50%), and intestinal dysfunction (40%). Six patients (20%) had scoliosis and syringomyelia. Among the neurological deficits, motor or sensory alterations were present in 4 (13.3%) and 2 (6.7%)

Table 1 Summary of the clinical characteristics of 20 patients in the preoperative period and their respective outcomes following the new surgical approach

No	Aetiology	Gender	Age at surger(yr)	Follow-up time	Urological dysfunction	Orthopedic deformities	Intestinal dysfunction	Neurological déficits	Follow-up conditions
1	Myelomeningocele	F	7	11 mo	Worsening bladder compliance	Clubfoot with ulcerated lesion		Paresthesia in lower limbs	Improvement in urological pattern
2	Myelomeningocele	F	4	9 yr 4 mo	Recurrent urinary tract infection		Intestinal constipation	Lower back pain	Improvement in lower back pain
3	Myelomeningocele	F	5	4 yr 4 mo	Worsening bladder compliance			Lower back pain	Improvement in lower back pain, urinary continence and worsening of fecal incontinence
4	Myelomeningocele	F	5	7 yr 5 mo	Recurrent urinary tract infection		Intestinal constipation	Lower limb paresis and low back pain	Urinary incontinence and worsening intestinal constipation
5	Myelomeningocele	F	6	13 yr	Worsening bladder compliance			Lower back pain	Improvement in lower back pain
6	Myelomeningocele	F	9	1 yr 6 mo	Recurrent urinary tract infection	Worsening scoliosis and recurrence of clubfoot		Lower back pain	Improvement in lower back pain
7	Myelomeningocele	F	12	9 mo	Recurrent urinary tract infection		Intestinal constipation	Lower limb paresis and low back pain	Improvement in lower back pain and intestinal constipation
8	Myelomeningocele	F	11	5 yr 3 mo			Intestinal constipation	Lower limb paresis and lower back pain	Improvement in lower back pain
9	Myelomeningocele	F	6	13 yr 5 mo	Recurrent urinary tract infection	Multiple orthopedic surgical			Improvement in urological pattern and reduction in urinary tract infection episodes
10	Myelomeningocele	F	10	1 yr 6 mo	Recurrent urinary tract infection	Multiple orthopedic surgical	Intestinal constipation	Lower back pain	Improvement in lower back pain, urological pattern and reduction in urinary tract infection episodes
11	Myelomeningocele	M	3	4 yr 3 mo	Recurrent urinary tract infection		Intestinal constipation		Without sphincter alteration
12	Myelomeningocele	M	8	3 yr 11 mo	Urinary incontinence and recurrent urinary tract infection	Multiple orthopedic surgical	Intestinal constipation		Improvement in intestinal constipation and urological pattern
13	Myelomeningocele	M	2	10 mo		Recurrence of clubfoot			Without sphincter alteration
14	Myelomeningocele	M	2	2 mo		Recurrence of clubfoot			Improvement in intestinal constipation
15	Filum lipomatosis	F	7	5 yr 9 mo		Difficulty in walking with frequent falls			Without sphincter alteration

Table 1 (continued)

No	Aetiology	Gender	Age at surger(yr)	Follow-up time	Urological dysfunction	Orthopedic deformities	Intestinal dysfunction	Neurological déficits	Follow-up conditions
16	Spinal lipoma	F	19	14 yr 3 mo				Lower back pain	Improvement in lower back pain
17	Spinal lipoma	F	12	13 yr		Recurrence of clubfoot		Lower back pain	Improvement in lower back pain and clubfoot
18	Spinal lipoma	M	2	3 yr		Recurrence of clubfoot	Intestinal constipation	Lower back pain	Improvement in lower back pain, urinary and fecal incontinence
19	Spinal lipoma	M	6	5 yr 8 mo		Difficulty in walking with frequent falls	Intestinal constipation	Pain in the right lower limb	Improvement in intestinal constipation and ambulation
20	Spinal lipoma	M	18	4 yr 8 mo		Multiple orthopedic surgical			Without sphincter alteration

yr = year; mo = month; F = female; M = male

Table 2 Summary of post surgical symptom status in 20 patients based on the type of spinal malformation

Aetiology	Total (n = 20)	Follow-up status	
		Improved	Stationary
Myelomeningocele	14	71,4%	28,6%
Filum lipomatoso	1	0	100%
Spinal lipoma	5	80%	20%

patients, respectively, while low back pain was reported in 14 (46.7%). Intestinal constipation was present in 40%, as was recurrence of clubfoot; 4 patients underwent multiple orthopedic surgical approaches for correction without improvement. Additionally, worsening scoliosis (10%) was also a clinical sign considered during evaluation. Signs and symptoms related to the genitourinary system included worsening bladder compliance (13.3%), recurrent urinary tract infection (30%), and urinary incontinence (3.3%).

Seventeen patients underwent preoperative voiding cystourethrography indicating the presence of vesicoureteral reflux in 41.2% of cases, ranging from grade III (28.6%) to grade IV (48.8%) and grade V (14.3%). Vesicoureteral reflux was mainly unilateral (85.7%), and in the case of grade V, the involvement was bilateral.

All surgical procedures were performed with microsurgery and intraoperative electrophysiological monitoring. There were no intraoperative complications related to spinal cord release, and postoperative complications such as cerebrospinal fluid fistula were present in only one case (5%). The 20 patients were followed up for 5.6 ± 4.6 years, ranging from approximately two months to fourteen years. In the postoperative evaluation, improvement in low back pain (90.9%) was particularly noteworthy, accompanied by improvement in urological pattern and episodes of urinary tract infection (45.4%). Among patients with complaints of constipation, 3 showed improvement (33.3%), and only one worsened (11.1%). There was improvement in ambulation and clubfoot in two cases (16.7%).

Almost half of the reoperated cases (40%) had a change in urological pattern, with six (46.17%) of patients who had intermittent continence shifting to social continence, and 2 patients worsening. Low back pain was the first symptom to improve after surgery, with an average time of 1.3 months, followed by changes in the urological pattern at 15.6 months. Improvement in constipation was observed in the first month in 2 cases (66.7%), positive changes in ambulation were observed around 7 months after surgery, and only one case showed improvement in clubfoot correction.

Patients with myelomeningocele had better symptom resolution (71.4%) compared to occult defect malformations ($p = 0.483$), with all reoperated cases from five years of age onwards showing improvement postoperatively

($p=0.015$) (Table 2). Symptom improvement was observed in (84.6%) of female patients and in (57.1%) of male patients ($p=0.122$).

Discussion

Patients undergoing primary correction of spinal dysraphisms may present throughout their lives, especially during the growth phase, with clinical manifestations of tethered cord, indicating the need for a new neurosurgical intervention [8]. Therefore, understanding the symptoms of STCS can provide a more early and timely intervention, allowing for better outcomes and increased quality of life for patients. In the literature, the prevalence of reoperation for tethered cord is 10 to 40% [4, 8, 9]. The incidence found in this study was 15.6%.

Similar to our data, Bettegowda et al. identified symptoms in 54 patients undergoing reoperation after myelomeningocele repair, including urinary dysfunction (87%), motor dysfunction (80%), and sensory deficits (61%) [10]. Similarly, Tseng et al. showed clinical manifestations in 31 symptomatic patients with primary correction of occult spinal dysraphism, mainly involving neurological deficits (83.9%), urinary dysfunction (77.4%), and orthopedic deformities (38.7%) [11].

Therefore, surgery to release the tethered cord has been performed more frequently in recent years, with indications based on clinical signs suggestive of spinal cord traction, which notably have a significant impact on the quality of life of these patients. Outpatient follow-up, in cases of spinal column malformations, is of great value for the identification and management of these symptoms, since magnetic resonance imaging will show spinal cord tethering in these patients, but the radiological signs of STCS are still obscure [12].

After correction of the tethered cord, pain is the symptom with the best positive outcome, present in our study in 90.9% of cases and in 87% in the analysis by Bettegowda et al. [10]. Additionally, we observed satisfactory improvement in the urological pattern accompanied by a decrease in the frequency of urinary tract infections (45.4%), reducing the use of prophylactic antibiotics among patients. On the other hand, 84.6% of patients with symptoms related to orthopedic deformities did not improve but stabilized deficits after surgery. Therefore, we conclude that even with a lower rate of improvement compared to other symptoms, orthopedic signs help in the identification of STCS, and surgical correction prevents the progression of these deformities.

The best outcomes of reoperations occurred in cases of myelomeningocele compared to occult defect malformations. Patients operated on after five years of age had better symptom resolutions compared to younger ages.

The treatment of STCS also impacts genitourinary care, contributing to the preservation of renal function and increased sociability of patients with spinal cord malformations. Balkan et al. [13], in their evaluation of 20 cases, observed alterations in the detrusor muscle bladder contractility pattern in 17 (85%) cases, with improvement after spinal cord release in 7 (35%). Reduced bladder compliance was observed in 85% of patients, with improvement and stability in the postoperative period in 10 (50%) and 8 (40%) respectively. In the same study, the incidence of vesicoureteral reflux was 42%, with the degree of involvement ranging from III to V according to the International Reflux Study in Children classification, and 50% of cases had renal scarring associated with reflux.

In this study, vesicoureteral reflux preoperatively was identified in 41.2% of our cases, with the majority classified as grade IV (48.8%). There was no indication for kidney transplantation in any of the cases during the follow-up period. Satisfactory changes in the urological pattern provided urinary continence after surgery in 46.17% of cases, ensuring greater autonomy and sociability that positively impacts the quality of life of these patients.

The limitations of this study were the lack of correlation between the results and access to postoperative rehabilitation. Details of the surgical technique and pre- and postoperative images were also not analyzed in this study. Furthermore, we understand that STCS has a different pathophysiology depending on whether the dysraphism is open or closed, and to compare them, we must consider their particularities.

The indication for STCS treatment requires recognition of its clinical symptoms, and therefore, multidisciplinary follow-up is essential for surgery to be indicated at an opportune moment.

Conclusion

The indication for reoperation of tethered cord is based on neurological, urological, and orthopedic clinical alterations. Our data corroborate the relevant literature: surgery interrupts the progression of symptoms, improving especially low back pain and urological alterations, positively impacting quality of life.

Author contributions V.C.A contributed to the study by collecting data from medical records, conducting a review of data available in the literature, and drafting the final manuscript.

M.T.C.L contributed to the study by planning and designing the study, as well as analyzing the data collected from medical records.

P.A.D contributed to the study by designing the study, analyzing data collected from medical records, and reviewing data gathered from available literature.

R.A.W contributed to the study by collecting literature data, analyzing the results, and performing the final translation of the text.

N.Z.C contributed to the study by designing the study, analyzing collected data, drafting the manuscript, and performing the final translation of the text.

Data availability The data supporting the findings of this study are not openly available for reasons supported by Brazilian Law 13,709/18, which aims to ensure the privacy and protection of patients' personal data. Data may be made available by the corresponding author upon reasonable request. The data is located in the controlled access data storage provided by the Federal University of Sao Paulo.

Declarations

Conflict of interest The authors declare no competing interests.

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