Is Surgery Necessary for Asymptomatic Tethered Cord in Anorectal Malformation Patients?

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Background: Evidence supporting routine surgery for asymptomatic tethered cord in patients with anorectal malformation (ARM) is, at best, speculative. The authors therefore examined whether untethering is indicated for asymptomatic tethered cord in patients with ARM.

Methods: A retrospective analysis of all patients with ARM (n = 223) between 1992 and 2002 was conducted. During the same period, 435 patients had surgery for tethered cord.

Results: Tethered cord was detected radiologically in 22 (9.8%); 8 patients with a low conus, and 14 with a low conus with and thickened filum. Seven of 22 patients underwent untethering; 3 prophylatic (14%) and 4 for neuro/motor function deficits (18%). All 4 symptomatic patients had significant clinical improvement in their neuro/motor functions after surgery. However, bowel and urinary functions remained unchanged in all 7 patients with a mean follow-up of 6.4 years (range, 4 to 8 years). Fifteen patients with radiologically diagnosed tethered cord remain asymptomatic with a mean follow-up of 2.7 years (range, 8 months to 10 years).

Conclusions: Neuro/motor functions clearly improved with surgery in symptomatic patients. However, bowel and urinary functions remained unchanged after surgery. Only 4 ARM patients with tethered cord required surgery, whereas prophylactic surgery appears to have minimal benefit. Expectant conservative approach in the management of asymptomatic tethered cord patient appears to be safe.

INDEX WORDS: Asymptomatic tethered cord, anorectal malformations, prophylactic surgery.

CONGENITAL ANOMALIES affecting the development of the anus and rectum are relatively common birth defects occurring 1 in 2,500 to 5,000 live births.[1] Malformation or maldevelopment of the region represents a developmental field defect frequently involving number of adjacent structures including the derivatives of notochord and neural tube.[2] Tethering of the spinal cord is one of the commonly recognized anomalies in association with congenital anorectal malformation (ARM).[3] Despite the long-standing interests in ARM, all issues regarding tethered cord in the setting of ARM, including definition, incidence, management, and clinical outcomes, remain controversial.[4,5]

Because the term *tethered cord* is morphologic and descriptive, the definition of tethered cord has been evolving, particularly since the introduction of more refined imagining modalities such as magnetic resonance imaging (MRI). Of course, increased detection changes the incidence of the anomaly and further compounds the issues involving its natural history and appropriate management. Moreover, it remains unclear as to whether tethered cord in the setting of ARM behaves in the same way as tethered cord in any spinal dysraphism. There is general agreement that symptomatic tethered cord should be treated surgically.[3,5-7] The clinical outcomes after untethering in both symptomatic and asymptomatic ARM patients, however, appear to be quite variable and unclear.

To clarify how the tethered cord detected in the setting of ARM should best be managed, we have analyzed the clinical outcomes of all ARM patients with symptomatic and asymptomatic tethered cord.

MATERIALS AND METHODS

A retrospective analysis was performed of patients treated for congenital anorectal malformation (ARM) between 1992 and 2002 by pediatric surgical service and all tethered cord patients who underwent surgery by neurosurgical service between 1986 and 2002. Two hundred twenty-three consecutive patients with ARM and 435 patients with the diagnosis of tethered cord were identified for the study. Cross referencing was done for all ARM patients to determine which patients had a history of tethered cord. Data collection included patient demographics, age at diagnosis, length of follow-up, presence or absence of tethered cord, motor symptoms, bowel or bladder incontinence, type of anorectal malformation (high or low), presence of associated anomalies, radiologic findings, indications for surgery, surgical outcome, and postsurgical complications. Statistical analysis was conducted using Fisher's Exact test or χ^2 analysis where appropriate, with a P value less than .05 considered significant. SPSS version 11.0 was used.

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Table 1. Patient Demographics

Types of ARM	Male to Female Ratio	Tethered Cord (%)	Neuro/motor Symptoms (%)
High (n = 161)	99:62	15 (9.3)	2 (1.2)
Low (n = 62)	44:18	7 (11.3)	2 (3.2)
Total (n = 223)	143:80	22 (9.8)	4 (1.8)

RESULTS

Patient Demographics

One hundred sixty-one patients of 223 ARM patients diagnosed during the 11-year study period had high lesions (72%), whereas 62 of 223 had low lesions (28%; Table 1). The distinction of high versus low type of ARM was made on the basis of clinical assessment and type of surgical or medical treatment required. There was higher preponderance of boys in both high (61% *v* 38%) and low (71% *v* 29%) types of anorectal malformation. Tethered cord, however, was detected only in 15 (9.3%) of high lesions and 7 (11.3%) in low lesions. Less than 2% of ARM patients (n = 4) had associated neuro/motor symptoms with 1.2% in high lesions and 3.2% in low lesions, respectively.

In our series, 22 patients of 223 ARM patients (9.86%) had a concomitant radiologic diagnosis of tethered cord. Fourteen of these patients (64%) were boys, whereas (36%) were girls. Four (18%) patients showed signs of neuromotor deterioration requiring surgery (1.1 to 4.94 years; mean, 2.5 ± 1.7 years), whereas 18 (82%) remained asymptomatic. Three patients (14%) had prophylactic surgery (2.6 months to 9.8 years; mean, 3.7 ± 5.3 years), whereas the remaining 15 patients (68%; 10.4 months to 5.3 years; mean, 2.5 ± 1.5 years) remain asymptomatic and continue to be followed up closely in clinic. In the prophylactic surgery group, all 3 patients (2 boys and 1 girl) had a high anorectal malformation. In the symptomatic surgery group (4 boys), 2 patients had a high malformation, whereas the remaining 2 had a low malformation. In the group being followed up, 10 had a high malformation, and 5 had a low malformation.

Effect of Practice Pattern on Management of Tethered Cord in ARM Patients

Although the study institution provided health care coverage to a relatively stable pediatric population base (1.3 million children) in a geographically well-defined area, the study period represented a significant turnover (80%) in the make up of pediatric surgical practice pattern. Between 1992 and 1997, only 6 patients of 143 ARM patients had tethered cord diagnosed, and 3 (50%) of these 6 patients required corrective surgery for symptoms (Fig 1A). The use of screening sacral ultrasound scan or MRI was not a routine practice during this period; hence, the overall detection rate of tethered cord was only 4%. The relative incidence of tethered cord in high versus low type of ARM was not significantly different (Fisher's Exact test; $P = .625$).

In contrast, between 1998 and 2002, with consensus and accordant change in practice pattern at the institution, screening sacral ultrasound scan and MRI were used routinely. Of 80 patients with ARM diagnosed during this period, 16 patients (20%) had concomitant tethered cord detected, and only 1 patient required surgery for symptoms (Fig 1B). The relative incidence of tethered cord in high versus low type of ARM was comparable and similar to the previous period. This change in practice pattern resulted in a dramatic 5-fold increase in the detection rate of tethered cord between the periods 1992 through 1997 (4.19%) and 1998 through 2002 (20%; $P \leq .0001$). Despite this difference between the 2 time periods, little change can be seen in

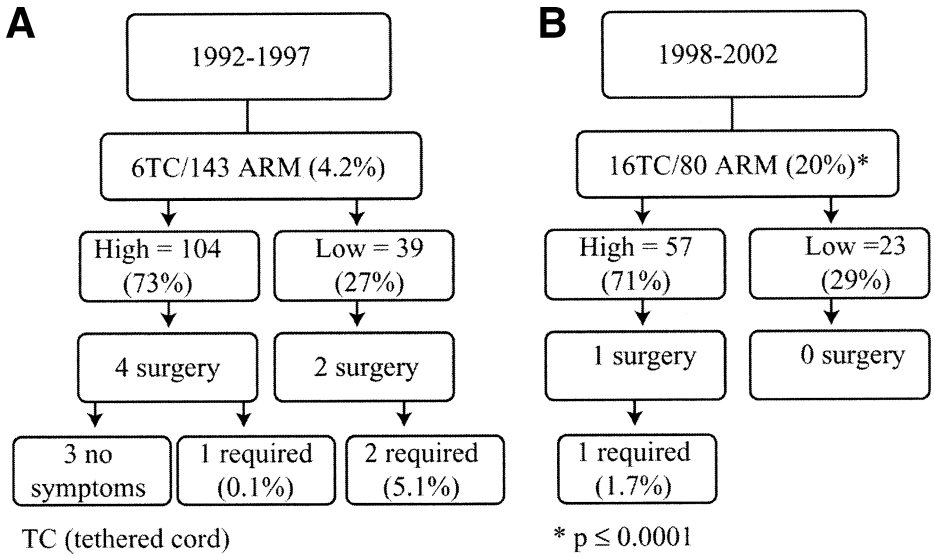

Fig 1. ARM was diagnosed in 223 patients with between 1992 and 2002. These patients were categorized into 2 time periods, (A) 1992 through 1997, and (B) 1998 through 2002. The relative distribution of high versus low type anorectal malformation is shown. In addition, relative proportions of ARM patients with diagnosis of tethered cord and their indications for surgery are shown. The difference in detection rates between the 2 periods, 6 of 143 (4.19%) in 1992 through 1997 and 16 of 80 (20%) in 1998 through 2002, was found to be statistically significant. ($\chi^2 = 12.68$; $P \leq .0001$).

Table 2. Radiographic Description and Associated Anomalies

	High ARM	Low ARM	Radiologic Findings	Skeletal Anomalies	Other Anorectal Malformations
Symptomatic (n = 4)	2	2	Low conus + thickened filum (2)	VACTERL (2)	Bifid scrotum (2)
			Low conus + thickened filum + lipoma (1)	Scoliosis (1)	Hypospadias (1)
			Lipoma only (1)		Bladder-rectal fistula (1)
Asymptomatic (n = 18)	13	5	Low conus + lipoma (4)	VACTERL (6)	Recto-vaginal fistula (3)
			Low conus + thickened filum (7)	Scoliosis (1)	Cloacal malformation (2)
			Low conus + thickened filum + lipoma (1)	Skeletal anomalies (4)	Cloacal extrophy (1)
			Low conus only (6)		Vaginal duplication (1)

the rate of onset of symptoms, which were 2.1% (1992 through 1997), and 1.2% (1998 through 2002).

Radiologic Findings and Associated Anomalies in Tethered Cord Patients

Given the fact that the definition of tethered cord is descriptive and radiographically morphologic, it is important to characterize tethered cord clearly in detail. Radiologic findings in all 22 patients included tethered cord (n = 11), thick and fatty filum (n = 13), low conus (n = 18), lipoma (n = 8), and hydromyelia (n = 2). Other findings (n = 4) included syrinx, syringomyelia, spinal dysraphism, and caudal regression syndrome (Table 2).

Associated anomalies included VACTERL, skeletal anomalies, dysmorphic features, kidney and chromosomal abnormalities, and patent ductus arteriosis. A number of patients also presented with additional anorectal malformations besides imperforate anus. These included cloacal extrophy, other cloacal malformations, rectovaginal fistula, double vaginal opening, bifid scrotum, hypospadias, bladder-rectal fistula, and undescended testis. When comparing the groups, no significant difference was observed ($\chi^2 = 17.5$; $P = .23$).

Clinical Outcomes in Symptomatic Versus Asymptomatic Tethered Cord Patients

Symptoms improved significantly in all patients who had surgery for neuro/motor deterioration (Table 3). Specifically, the surgery was beneficial in reversing the damage that caused gait disturbances, muscle tone (weakness or rigidity), and delayed developmental milestones. Urinary and bowel incontinence present preoperatively did not improve with the mean follow-up of approximately 5 years. There was no death associated with surgery; however, one patient's recovery was prolonged by a ceredrospinal-fluid (CSF) leak post operatively. No other surgical complications were noted in this group.

Similarly, 3 patients underwent prophylactic untethering of the cord (Table 3). The decision to have prophylactic surgery was because of patient/surgeon preference. All 3 patients had urinary and bowel symptoms, and these did not improve after the surgery with mean follow-up of 8.4 years. These patients, however, did not have neuro/motor symptoms. There were no complications.

Fifteen ARM patients with the diagnosis of tethered cord are being followed up with the mean follow-up of only 2.5 years. Eleven patients in this group were not currently toilet trained and, therefore, could not be assessed for bowel and urinary function. However, no deterioration in clinical status is noted to date with respect to neuro/motor, urinary and bowel functions.

DISCUSSION

From 1986 to 2002, a total of 435 children also underwent surgery for untethering of the spinal cord at the same institution. Of these, only 7 patients (4 patients for neuron/motor symptoms and 3 for prophylactic reasons), had concomitant diagnosis of anorectal malformation. These represent 7 patients of 22 patients with ARM and tethered cord diagnosed (32%) or 7 of 223 patients with ARM diagnosed between 1992 and 2002 (3%). It is interesting to note that only 4 patients of 223 in the ARM patient population (<2%) had symptoms related to tethered cord during the period.

These findings raise 2 critical clinical issues relating to diagnosis of tethered cord in ARM patients. First, the incidence of tethered cord in the setting of ARM is significantly influenced by practice pattern. This is shown clearly by our practice pattern where with introduction of screening ultrasound scan or MRI, our detection rate for tethered cord in ARM patients increased by 500%. The controversy surrounding the incidence of

Table 3. Clinical Outcomes for Tethered Cord in ARM Patients

	Age at Surgery	Age at Follow-Up	Preoperative Incontinence	Postoperative Incontinence	Preoperative Motor Symptoms	Postoperative Motor Symptoms
Symptomatic (n = 4)	2.5 yr	4.9 yr	3	3	4	0
Prophylactic (n = 3)	3.7 yr	8.0 yr	3	3	0	0
Followed up (n = 15)	—	2.5 yr	—	—	—	—

tethered cord in ARM patients has been raised previously with fairly disparate data.[4,5] Our results are consistent with the report from Peña's group with overall incidence in the range of 20% of patients with ARM.[4] Although our data represent an institution-based reporting, the health coverage of the institution represents a geographically defined, relatively stable population base, in keeping with a population data. We therefore feel that the overall incidence of approximately 20% may be an accurate estimation of the "true" incidence of tethered cord in the setting of ARM.

The second and more important issue relates to the natural history of tethered cord in patients with ARM. Practice difference, referral pattern, and introduction of new imaging modalities can significantly influence and skew the incidence of tethered cord in any population. But what does this mean? Our data indicate that less than 2% of patients with ARM have symptoms attributable to tethered cord, and only approximately 18% of patients with both tethered cord and ARM have symptoms. Even with development of symptoms, neuron/motor symptoms attributable to tethered cord can be effectively corrected. A number of investigators have suggested previously that the natural history of tethered cord is one of progression.[5-8] This has been suggested on the basis of development of symptoms in adult patients.[8-11] Although it was assumed that because adults had completed their growth curve, they were less likely to show any neurologic deterioration because the spinal column had already been subject to rapid growth.[10] However, it has been reported that a sudden onset of symptoms including pain, motor and sensory deficits, and incontinence may occur after sudden movements that cause traction in the spinal cord.[10,11] In a study of 56 adult patients with tethered cord reporting symptoms, 26 of them (46%) had been previously asymptomatic in childhood.[11] Because only 50% of sensory and motor deficits showed improvement if surgery was done within the first 5 years of symptoms, these investigators extrapolated their observation that the natural history of tethered cord is one of potential progression and pitfalls. However, none of these patients had concomitant ARM. We feel that it may be important to make a distinction and put *tethered cord* as a subgroup in the setting of ARM separately, because they may represent a different spectrum of spinal dysraphism the natural history of which may be different from those tethered cord patients without ARM. This view is supported by the long-term follow-up in our study. Considering the relative stability of patient populations in different regions in Canada, if the natural history of tethered cord in the setting of ARM is one of progression, these patients would have been detected at the study institution over the follow-up of 18 years reported here.

Now, having shown that the natural history of tethered cord in ARM patients is not one of progression, what do we do with all these ARM patients with tethered cord? Routine prophylactic correction of tethered cord has been advocated previously with the supporting evidence that these patients remain asymptomatic, and the surgery has minimal complications.[5] If the natural history of these patients is to remain asymptomatic for most part, it is not clear how "true" benefit of correction of asymptomatic lesion can be shown. Three patients in our study group who had prophylactic surgery for tethered cord did not show any significant improvement, especially with respect to bowel and urinary symptoms. These outcomes are consistent with the current literature.[4,9] In a study conducted by Morimoto et al,[3] all 10 patients continued to have bladder dysfunction. Furthermore, prophylatic surgery is associated with a number of risks. First, surgical release of the tethered cord always carries the inherent risk of injuring neighbouring motor and sensory nerve roots.[8] Patients with a lipoma present an even greater risk of injury to the nerve roots during surgery.[8] Second, there is risk of retethering after surgery, because postoperative scar tissue causes the cord to retether to the dura and subcutaneous tissues.[1] If markedly diminished movement of the cord is seen while the patient is in prone position, retethering is highly suspect.[1] In addition, there is no way to substantiate if "there was an ischemic cord or that the cord would develop ischemia"[9] despite the radiologic diagnosis of a tethered cord. The lack of improvement in bowel and urinary function after untethering for symptoms and prophylactic reasons is not uncommon. None of our 4 patients operated on for symptoms and 3 operated on for prophylactic reasons had any improvement of bowel and urinary symptoms. These outcomes are in keeping with the experience from Peña's group and those in the neurosurgical literature.[4-8] In contrast, surgical correction halted the progression of neuromotor symptoms and, in fact, reversed them. The most dramatic illustration of this was in a 4-year-old patient with concomitant cerebral palsy CP who had never walked. Three months after surgery, he was able to take his first steps.

We advocate a conservative approach for ARM patients with asymptomatic tethered cord. We believe that our study is reflective of the true natural history of tethered cord in the ARM population. Despite the changes in practice with the introduction of routine screening and the subsequent 5-fold increase of tethered cord in these patients, ultimately, the outcome appears to be the same. In patients that did proceed to neuro/motor deterioration, neuromotor functions clearly improved with surgery. Bowel and urinary functions remained unchanged in both the symptomatic and prophylatic surgery groups. Prophylactic surgery appears to have had minimal benefit in this population.

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