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Tethered Cord as a Cause of Scoliosis in Children with a Myelomeningocele

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Abstract. Scoliosis interferes significantly with the functional ability of most children with a myelomeningocele. While it is recognized that tethered cord at the repair site causes neurological deterioration, it has been controversial whether tethered cord causes scoliosis. The spinal cord was untethered in 30 children with progressive loss of function and scoliosis. Of 6 children with curves greater than 50° only 1 improved. Of the other 24 children their curves were stable or improved at 1 year follow-up. At late follow-up, 2–7 years, 63% were stable or improved while 38% began to progress. Tethered cord causes scoliosis and stability or improvement can be anticipated following untethering. Close long-term follow-up is essential to identify those individuals with retethering of their cord.

Introduction

Scoliosis occurs in greater than 80% [5] of children with myelodysplasia by the 10th year of life. This significantly reduces the possibility of functional independence. The curvature is typically proximal to the level of the myelomeningocele and not associated with vertebral anomalies. They differ from 'idiopathic' curves because they occur equally in boys and girls, left and right curves are equally frequent and it occurs in preadolescence. While hydromyelia and the Chiari II malformation have been implicated as causes of the scoliosis the role of tethering at the repair site has remained speculative. It is well recognized that deterioration in gait, weakness, spasticity, neuro-orthopedic deformities, loss of urinary control and pain are common signs of a tethered cord in children with a myelomeningocele [6]. It is less clear whether scoliosis can be a sign and even possibly the first sign of a tethered cord. In this paper we present a group of children born with a myelomeningocele, who developed scoliosis in the absence of a shunt malfunction, hydromyelia, or hindbrain compression. All of the curves were above the repair site and not associated with vertebral anomalies. They were treated with untethering alone at the repair site.

Clinical Materials and Methods

Ninety-one children had surgical release of their tethered spinal cord performed at the site of their initial myelomeningocele repair. Forty-three had scoliosis as one of their signs of deterioration and of these 30 had a tethered cord alone without hindbrain compression or hydromyelia. All of the children had lumbar level myelomeningoceles ranging from thoracolumbar to lumbosacral. The curves were proximal to the repair site and in the absence of vertebral anomalies. The age range of the 30 children at operation was from 2 to 18 years with a mean of 6.4 years. The sex ratio was 49 females and 42 males in the 91 children untethered. Of the 30 children with scoliosis, 13 were male and 17 were female. The diagnosis of a tethered spinal cord at the repair site was made on a clinical basis after a shunt malfunction was ruled out. MRI and/or CT myelography were used to rule out hydromyelia or other causes of deterioration. The untethering procedure was carried out with the CO_2 laser by a technique similar to the one published previously [4]. Follow-up ranged from 1 to 7 years.

Results

Of the total 91 children operated upon for tethered cord, two had an increased motor deficit in their lower extremities as a direct result of the surgery. One child recovered the motor loss partially and also had a significant improvement in his scoliosis. The other child did

not have scoliosis and remains stable. Neither were expected to be community ambulators prior to the untethering procedure. There was no mortality. Thirty children had scoliosis without hydromyelia. We initially looked at the degress of curvature of each child at 1 year postuntethering of the cord. The curve was classified as improved if decreased by 10° or more and progressive if it increased by 10° or more. Curves between these limits were considered stable. The prediced rate of progression of the curve in a child with a myelomeningocele would be a 1° per month or about 12° per year. All but 1 of the 6 children with curves greater than 50° progressed and required fusion. This single child droped below 50° and is stable at 45° in a body jacket. The remainder of the evaluation will consider only the 24 children below 50° at the time of untethering. All but 1 of the 24 children (96%) improved or were stable at 1 year follow-up. Eight (33%) improved and 15 or 63% were stable. We were surprised and gratified by the results at 1 year follow-up. However, at subsequent follow-up, 2–7 years, progression began to occur in some of the children. At last follow-up 5 (21%) remain improved, and 10 (42%) are stable (63% are improved/stable) and 37% began to progress beyond 1 year of follow-up.

Figures 1 through 9 are examples of children that had some improvement in their curves following untethering of the spinal cord at their repair site. Improvement less than 10° was considered stable. A CT myelogram or MRI image is included to show the absence of hydromyelia. The child in figure 6 had a shunt revision several months after the release of the tethered cord. The improvement in the curve predated the shunt revision.

Discussion

Hendrick et al. [2] reported 3 children with painful scoliosis and a tight filum terminale that did not require spinal fusion following release of the tethered cord. In the series of Hoffman et al. [3] 7 of 9 had a fusion. In the reports of Hoffman et al. [3] and Hendrick et al. [2] the children had tethered spinal cords but did not have a myelomeningocele. It is not clear in Reigel's series whether all the children had a repaired myelomeningocele as the cause of tethered cord. Reigel [6] stated 'Our data does not permit a conclusion regarding the role of tethered cord in the etiology of scoliosis'. In Reigel's series of 14 children no patient improved, 10 of 14 had arrest of their progression and 6 of these had a subsequent fusion. In the series of Venes and Stevens [7] 1 of 6 children improved after untethering; however, this case also had hydromelia.

Aggressive management of shunt function and the belief that deterioration in a child born with a myelomeningocele has a treatable cause has led us to search for the cause and aggressively intervene. We feel strongly that the initial search for a cause of deterioration in a child a with a myelomeningocele should be directed at the shunt. Once it is determined that the shunt is functioning optimally other causes are sought. These include hydromyelia, compression of the Chiari II malformation and tethered cord. Evidence that tethered cord causes scoliosis is indirect. From this study it is clear that something associated with the untethering of the spinal cord at the repair site stabilized or improved the curve in most children with curves less than 50°. This stability lasts beyond 1 year in 63% of the children. One can only speculate as to the mechanism producing scoliosis in these children.

We have noted compression of the cord held against the normal thoracic kyphobic curve on flexion in children with tethered cords at the repair site. This produces flattening of the spinal cord as seen on ultrasound. Possibly this repeated flattening causes a reversible focal myelopathy. Fujita et al. [1] have shown that the tethered cord is much more sensitive to compression.

Storrs [pers. commun.] has postulated a mechanism involving abnormal function in ascending intersegmental pathways resulting from ischemic injury at the repair site. This would produce asymmetric tone or imbalance in parathoracic muscles leading to scoliosis. Untethering would release local ischemia effects of tethering and return balance to the ascending system. A phenomenon similar to this has been noted in patients with cerebral palsy where a selective dorsal root rhizotomy at lumbar levels improves tone and function in the upper extremities.

Whatever the cause, untethering of the spinal cord at the repair site has a significant effect on the scoliosis. One can initially anticipate at least stabilization. Progression beyond 1 year of follow-up probably represents retethering in most cases. Obviously a shunt malfunction, hydromyelia, or Chiari II compression must be considered. Previously scoliosis alone was not an indication for untethering. We have reconsidered our position and feel these cases represent retethering and reoperation is indicated. Because our numbers are small and the follow-up too short we cannot make a conclusion about the effectiveness of this approach.

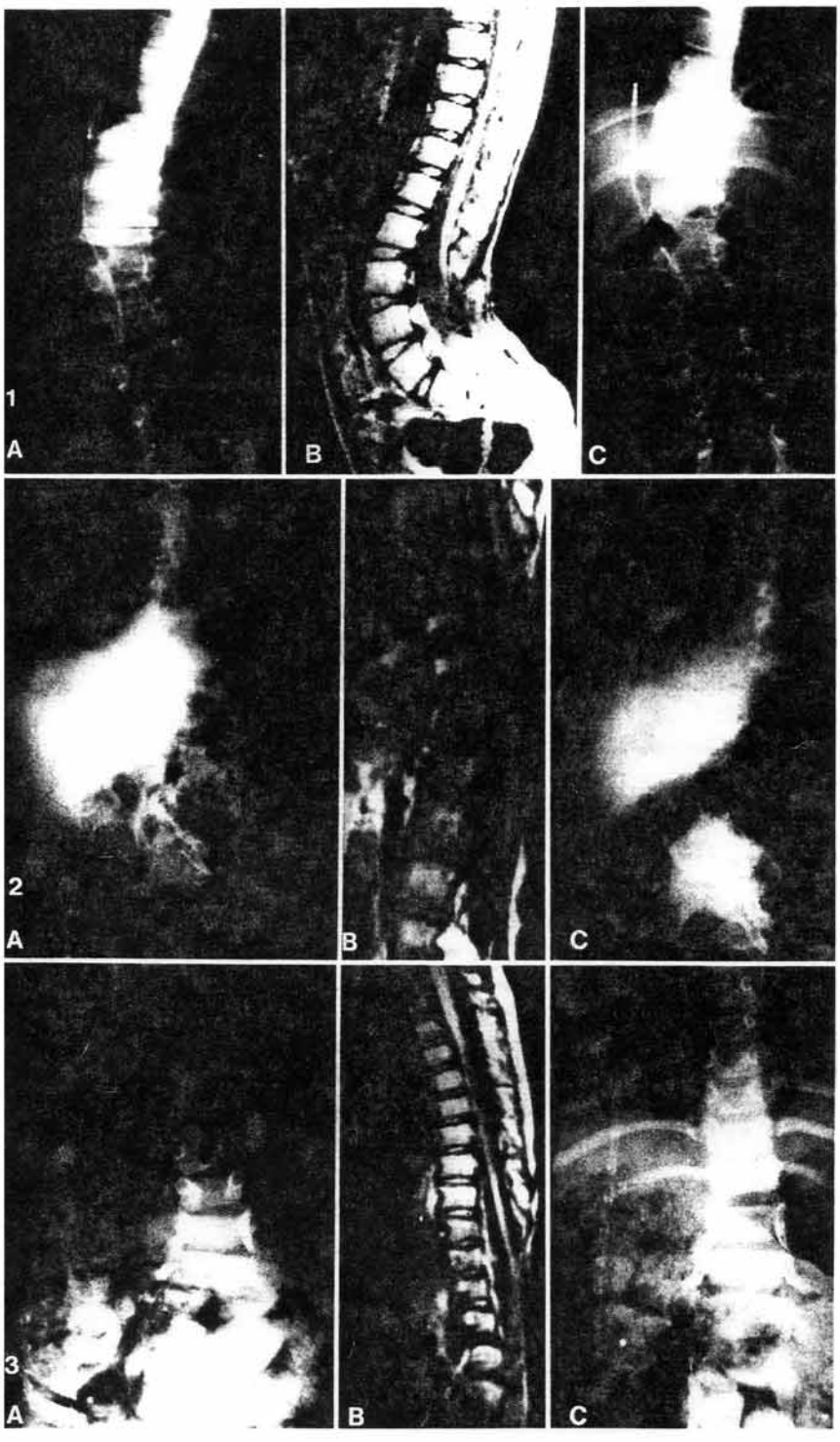

Fig. 1–3. Three children are shown with a preoperative spinal film **(A)**, an MRI which shows the spinal cord without hydromyelia **(B)** and the postoperative curve **(C)**. In this group of children the curve improved between 5 and 20°.

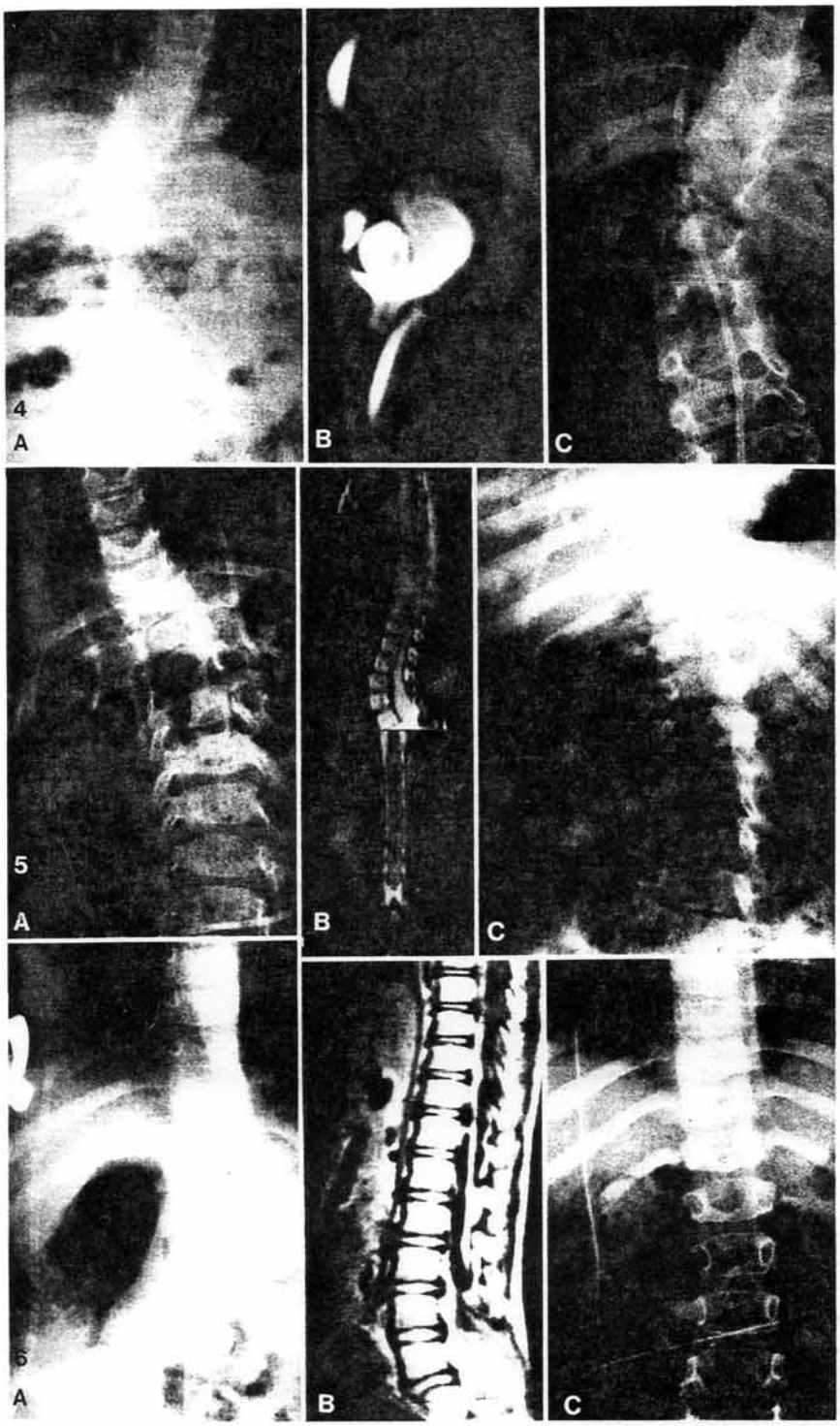

Fig. 4–6. Three children are shown with preoperative curves **(A)**, a study to show the absence of hydromyelia **(B)** and the postoperative study **(C)**. Curves improved between 15 and 20°.

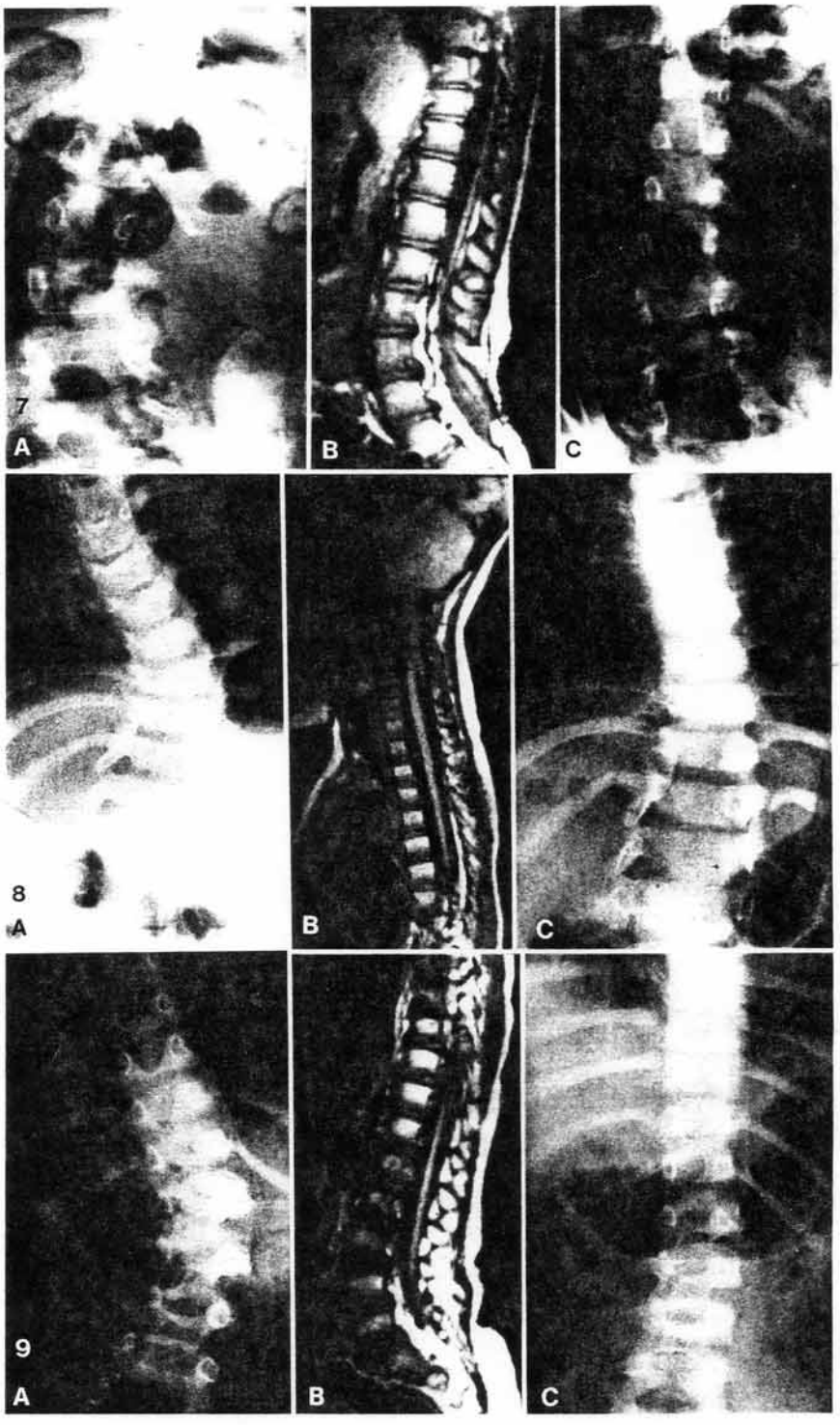

Fig. 7–9. Three children with pre- **(A)** and postoperative **(C)** curves with an MRI **(B)** to show the absence of hydromyelia. Curves in t group improved by 20–30°.

Is it reasonable to reoperate upon a child at 2–4 years for a retethered cord? As high as a third may show progression within 4 years. One must weigh reoperation versus functional limitations imposed by progressive scoliosis and early spinal fusion. Reoperation seems preferable even if it only delays fusion till maturity. Obviously early permanent untethering is optimal but further refinement of our technique is needed to achieve this goal.

In the past, we simply braced the slowly progressive curve until the children were older and then fused their spines. It is our hope that with untethering and body jackets we can hold the curves to less than 40° and often to less than 20° until the child is mature. While it is too early to state with certainty, we anticipate that many of these children will not require a fusion.

Conclusions

Findings in this study were as follows. (1) Tethered cord alone was the most common cause of scoliosis proximal to the repair site (70%). (2) Untethering improved the curve in 21% and stabilized the curve in 42% of the children with curves less than 50° at 2–4 years of follow-up. (3) Only 1 in 6 (17%) curves greater than 50° improved. The other 5 progressed. Early untethering prior to 50° is important. (4) All children, except for 1 with curves less than 50° were either stabilized or improved by untethering of the spinal cord at 1 year. While nearly 40% again show progression beyond 1 year. This suggests a high retethering rate for this group. (5) Of the total 91 children operated upon for tethered cord 2 were made worse (2%), both lost some motor function. One had scoliosis which improved following surgery.

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