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ORIGINAL PAPER

Diastematomyelia in children: treatment outcome and natural history of associated syringomyelia

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Abstract
Objective To quantify the long-term outcome of children with diastematomyelia and the implication of syringomyelia in the natural history.
Materials Retrospective study of 17 children (nine girls and eight boys) with diastematomyelia operated during 1989–2004. Mean age at diagnosis was 3.4 years (range 5 days–12 years), mean follow-up was 5 years; 14 had excision of spur and cord untethering, 3 had excision of spur alone.
Results The bony spur was in the lumbar region in 12 and thoracic in 5 patients. Syringomyelia was present in eight (47%), associated with bony spur between L1 and 4 (p=0.088), spina bifida occulta in ten (58.8%), and spinal lipoma in one (6%). Preoperative Necker Enfants Malades (NEM) scores were 17 in eight patients, 16 in four, 15 in three, 13 in one, and 12 in one patient. Presenting neurological deficits were motor in eight, sensory deficits in three, and anal incontinence in one patient. There was improvement of NEM scores postoperatively in five patients only, but still with residual deficit. Repeat spur excision and cord untethering was performed in three patients for neurological deterioration 1–4 years after first operation. On postoperative MRI scans syringomyelia remained unchanged in all eight patients. There were one skin infection, one transient motor deterioration, and one patient with sensory deficit after surgery.
Conclusion Prophylactic operations were associated with the best clinical outcome. Despite improvement, all patients with established preoperative deficit still had residual neurological deficits at their last follow-up. The associated syringomyelia remained unchanged after surgery, indicating that it does not contribute to the neurological syndrome.

Keywords Diastematomyelia · Syringomyelia · Spinal dysraphism · Cord untethering

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Introduction

Diastematomyelia is a rare form of occult spinal dysraphism mainly in the pediatric population but occasionally in adults [27, 32], with the published incidence of 2–4 per 1,000 live births [18]. It is first described in 1837 by Ollivier and the term is derived from the Greek words diastema, meaning cleft, and myelos, meaning cord. It is as its name implies, a malformation where a portion of the spinal cord is split into two half cords with each half cords displaying a single set of anterior and posterior nerve roots [14]. It is a part of split cord malformations (SCMs). SCMs are commonly divided into two types: type 1 is where the neural tube is split into two separate entities along with the dural sheath with an intervening fibrocartilaginous or bony septum and type 2 where there is a single dural sheath enclosing a split spinal cord caused by fibrous elements. The type 2 malformation is known as diplomyelia whereas the type 1 malformation is called diastematomyelia. The term diastematomyelia is used commonly to imply both types of SCM. Most if not all patients have tethered cords requiring surgical attention. The incidence of low-lying cord is also said to be as high as 83% of the patients and is also often associated with a thickened or fatty filum.

Syringomyelia is often seen to be associated with diastematomyelia on imaging but the incidence and the natural history of the associated syringomyelia is unclear. To attempt to answer this question, we reviewed a group of patients diagnosed with and operated for diastematomyelia (type 1 SCM) in our practice, record the clinical and radiological outcome of surgical management for this group of patients, and review the incidence and natural history of the associated syringomyelia after surgical treatment.

Materials and methods

This is a retrospective study of all the patients diagnosed and operated for diastematomyelia (type 1 SCM) from May 1989 to November 2004. During the same time period no patients with type 2 SCM were operated. All the case notes and pre- and postoperative radiographs were reviewed. There were 17 children, 9 girls and 8 boys. The mean age at presentation was 3.4 years (range 5 days to 12 years). The mean follow-up time for these patients was 5 years (range 1 to 13 years). The presenting symptoms and clinical signs were recorded and quantified objectively by using the Necker Enfants Malades (NEM) functional score scale developed by Pierre-Kahn et al. [26] for spinal lipoma. Each preoperative radiograph was reviewed with particular attention to the level of the diastematomyelia, the level of the spur, any associated syringomyelia with its extent and level, the level of the conus, and the presence of tonsillar herniation and any other associated abnormalities.

After surgery, the clinical details of each patient were then recorded at a minimum of 1 year of follow-up postprocedure. The information is recorded objectively via the NEM scores and also subjectively according to whether the patient or family perceived any difference in symptoms postsurgery. Any postoperative complications and the need for a second operation to reexcise the spur with the timeframe in reference to the first operation were also recorded. The results of postoperative radiographs for each patient were also reviewed to see whether there were any changes in the size of the associated syringomyelia, any residual spur, and any changes in the position of the cerebellar tonsils.

Results

Eight patients had no neurological symptoms and signs at presentation and nine patients (47.1%) had symptoms and neurological deficits: five patients had purely motor deficit, two patients had both motor and sensory deficits, one patient had sensory deficit, and one patient had both motor and anal deficit. On preoperative MRI scans the bony spur was in the lumbar region in 12 children (70%) of the patients and in the thoracic region in five patient. All but one of the patients had associated tethered cord with low-lying clonus. With the exception of three patients who were born with open myelomeningocele and Chiari II malformation, none of the other patients had a hindbrain hernia. There was spina bifida occulta in ten patients (58.8%) and spinal lipoma in one (6%). Syringomyelia was present in 8 of the 17 patients (47.1%), seven located in the thoracolumbar junction and one in the high thoracic area (Fig. 1). There was a tendency to statistical association between syringomyelia and presence of bony spur between L1 and 4, although it did not reach significant levels (p=0.088, chi-square). In seven patients syringomyelia was just above the level of the bony spur, in one it was six levels below, and in five patients it was located in the middle of the diastematomyelia, involving both halves of the spinal cord and the rest above the area of the diastematomyelia. The extent of the syringomyelia ranged for a length involving one to four vertebrae. The syringomyelia was long (more than three vertebrae in length) in three patients and short (less than three vertebrae in length) in five patients. All were judged radiologically to avoid tension and not causing pressure to the surrounding cord. Assessing the presenting symptoms and signs of each patient, none had symptoms and signs that could be reasonably attributed to the presence of the syringomyelia.

Table 1 shows the NEM scores before and after surgery. Preoperatively, 9 of the 17 patients had neurological deficits as assessed by the NEM scores with the rest having no neurological deficit. All the patients underwent surgery to excise the bony spur; 14 of the 17 patients also had untethering of the spinal cord by sectioning the filum terminale. Three patients did not undergo untethering of the cord as they had previous surgery to close a lumbar myelomeningocele. None of those three patients developed recurrent cord tethering in the region of the previously excised diastematomyelia spur. None had surgery for the syringomyelia. Reviewing the NEM scores postoperatively (Table 1), only five patients improved their score objectively, two in motor function, two in sensory function, and one in anal function, although that patient's motor function deteriorated after the operation. The rest showed no change on postoperative NEM scores. None of the patients deteriorated postoperatively. Subjectively, in all nine patients there was perceived improvement of symptoms after surgery, as the patients and/or the families reported; one patient reported decrease in motor function as mentioned above. Complications were superficial wound infection in one patient, which was treated with intravenous antibiotics, and transient worsening of sensory function in another patient.

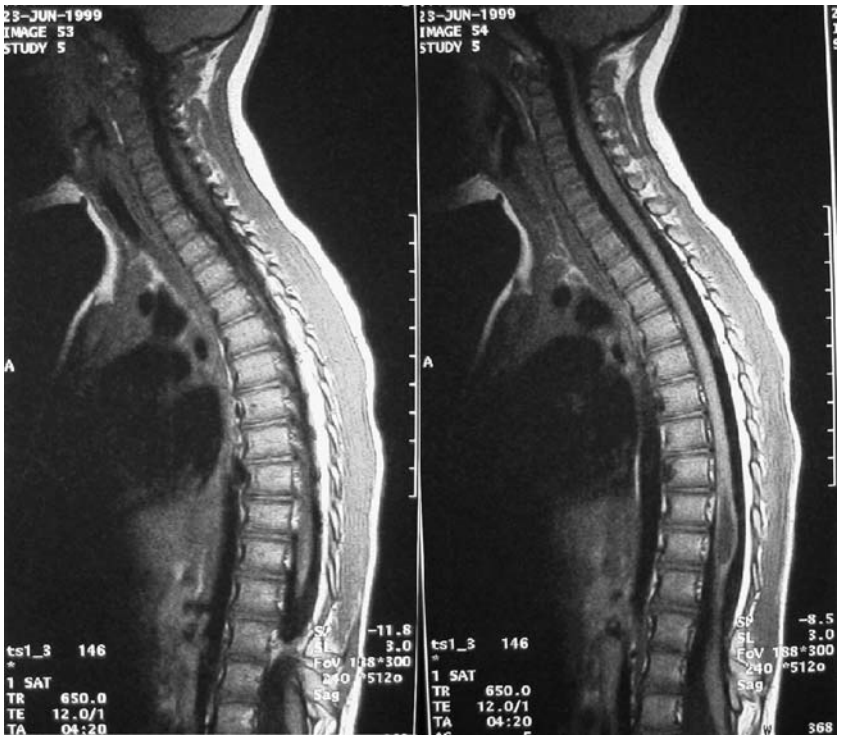

Fig. 1 T1-weighted sagittal MR scan of the spine of a 2.5-year-old girl who presented with a history of limping from the right leg while running. On examination she had significant distal weakness and foot drop. There is diastematomyelia with the bony peg at the level of T12 vertebral body. There is syringomyelia extending from T9 to T10, above the level of the diastematomyelia peg

Postoperative MRI scans at 1 year were available for 12 patients, including all those with syringomyelia, which showed no change in the size and extent of the syringomyelia in all 8 patients (Fig. 2). Two patients had residual bony spurs and one patient developed retethering with development of hindbrain hernia. All three patients subsequently deteriorated 15 months, 7 and 8 years, respectively, after the first operation due to retethering of the cord, requiring another operation to reexcise and untether the spinal cord. Two of these three patients who required repeat surgery had associated syringomyelia, which did not change in size and extent before or after the second operation. All three patients improved neurologically after the second operation. Fourteen patients had repeat MRI scans at last follow-up, which showed no residual bony spurs, no change in the size and extent of the associated syringomyelia, and no retethering of the spinal cord. All patients remained unchanged neurologically at last follow-up.

Discussion

It was initially thought that diastematomyelia had a different pathogenesis than diplomyelia and that they were two different entities. Diplomyelia was thought to be true duplication of the spinal cord at certain segments [5, 13, 21, 31] and diastematomyelia was thought to be caused by the bony spur, which suggests that it resulted from mesodermal invasion of the neural tube [1–3]. This was disproved effectively by Pang et al. [24] and now both conditions are thought to have similar embryogenesis. They are thought to be caused by the formation of an endomesenchymal tract by an accessory neurenteric canal between the yolk sac and amnion, which divides the neural canal and notochord. Depending on the timing of the formation of the endomesenchymal tract and subsequent mesenchymal infiltration, the neural tube may split into two separate components with an intervening fibrocartilaginous or bony septum (diastematomyelia) or remain a single dural tube with a split cord by fibrous tissues (diplomyelia). This would also

Table 1 Pre- and postoperative NEM scores

NEM score	Preoperatively (no. of patients)	Postoperatively (no. of patients)
17	8	11
16	4	2
15	3	2
13	1	1
12	1	1

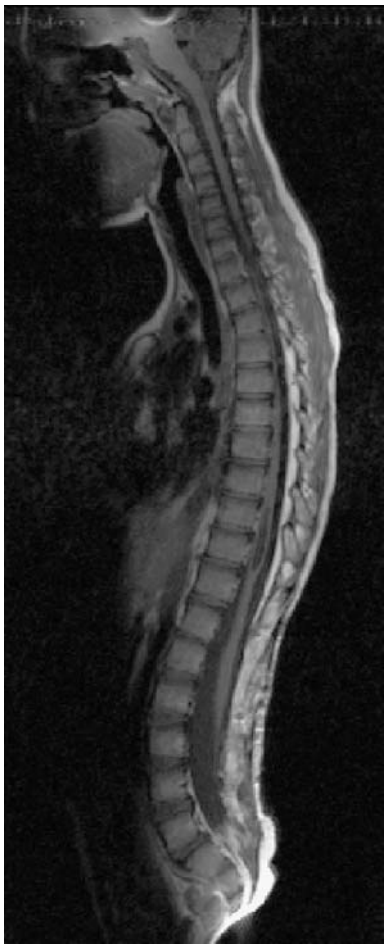

Fig. 2 T1-weighted sagittal MR scan of the patient of Fig. 1, 6 years after removal of the diastematomyelia peg, dural reconstruction, and division of the filum terminale. The syringomyelia cavity has changed little is size in comparison to the preoperative appearance in Fig. 1 (allowing for different magnification factor of the images). The right leg weakness improved after operation but she was left with a permanent foot drop for which she is using an ankle splint

explain the presence of commonly associated spinal abnormalities such as dermal sinus tracts, spinal lipomas, dermoids, neurenteric cysts, and even a meningocele or myelomeningocele as these abnormalities can arise from various ectodermal or endodermal remnants. Diastematomyelia usually presents in childhood [6, 23, 31] but adult cases are not unknown [27, 32]. It is also three times more common in female than male patients [11, 15, 19, 30].

Syringomyelia is often associated with diastematomyelia on MRI imaging. It is thought to be caused by tethering of the spinal cord [9, 22] from a tight filum terminale in the presence of a myelomeningocele [25], spinal lipoma [8, 10, 29], spina bifida occulta [28], and diastematomyelia [16, 29, 32], although rarely, no cause can be found [20]. The mechanism is thought to be deranged intracordal circulation of CSF from the perivascular and interstitial space to the subarachnoid space causing increased accumulation of CSF in the central canal as the result of tethering of the nerve roots of the spinal cord [29]. The syringomyelia is reported to disappear after untethering surgery unless there is associated arachnoiditis disrupting the CSF mechanism causing worsening of the syringomyelia after surgery [29]. Terminal syringomyelia with occult spinal dysraphic lesions is most often associated with tethered spinal cord from a tight filum terminale in the presence of an anorectal anomaly (67% of cases), meningocele manqué (54%), and diastematomyelia (38%).

The incidence of syringomyelia associated with diastematomyelia published in medical literature ranges from 29 to 55% [4, 7, 12, 16, 17, 29, 30, 33]. There is a difference of opinion on whether the syringomyelia contributes to neurological symptoms of the patient and whether surgical treatment, direct or indirect, alters the size of the syringomyelia and improves symptoms. A study done by Iskandar et al. [16] where there were ten syringomyelia cavities in 26 patients with diastematomyelia, eight of them large (>2 cm in length, >50% cross-sectional area of cord) and two small. It should be noted that all the small cavities were found to be asymptomatic and the size did not change throughout the study after untethering surgery, whereas the seven of the eight large syringes were symptomatic after untethering with five having back or leg pain and seven neurological deficit. The large syringomyelia cavities were treated with shunts with improvement of neurological symptoms in two patients with the rest unchanged, improvement in pain in all five patients, and reduction in size of the syringes in six patients. Hence, the study concluded that large syringomyelia should be treated surgically. In contrast, Scatliff et al. [29] found 11 syringomyelia cavities in 20 patients with diastematomyelia (55%) and the majority (8 of the 11) remained unchanged after untethering surgery with 3 patients showing improvement. None presented with symptoms attributed to the syringomyelia and none had it treated directly.

In this study, there were 8 of the 17 patients (47.1%) with syringomyelia, three with large and five small cavities. This incidence of syringomyelia is similar to the other published studies. Most patients had surgery to excise the bony spur and untether the cord. Preoperatively, no patients had symptoms attributable to the syringomyelia, and postoperatively, the size of the syringomyelia in all eight patients remained unchanged. In two of the patients with syringomyelia subsequent deterioration after the surgery was due to retethering of the cord and not due to increase in the size of their syringomyelia. From these results, we conclude that syringomyelia in patients with diastematomyelia very rarely, if at all, contributes to the neurological symptoms. The management of the syringomyelia should therefore be conservative at the first instance with regular monitoring. The presence of syringomyelia should not influence the decision for first surgery.

In this study, throughout the whole group of patients with diastematomyelia, surgery improved symptoms objectively (as assessed by the NEM scores) only in a small group of patients (four out of nine patients) once fixed neurological deficits have developed and even then the neurological deficits rarely resolved completely. However, subjective symptom improvement was felt by all the patients and their parents in some way, which was encouraging although this was not reflected in the objective NEM scores. This may indicate that prophylactic cord untethering and repair of diastematomyelia before neurological deficits were established may lead to better clinical results.

Conclusion

This study indicates that in patients with diastematomyelia prophylactic surgical repair before development of neurological deficits achieves the best outcome. Syringomyelia associated with diastematomyelia does not contribute to the clinical syndrome and its presence should not influence the decision for surgical treatment.

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