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Magnetic resonance detection of myelodysplasia in children with Currarino triad

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Abstract *Purpose*. To evaluate the role of MRI in the detection of myelodysplasia in children with Currarino triad.
Materials and methods. Six patients (two girls, four boys, aged 7 months–14 years, mean age 6 years) were studied with MRI, voiding cystourethrogram and barium enema or fistulography. CT and ultrasonography were also performed in two patients.
Results. All patients presented with partial agenesis of the sacrum. Three patients suffered from an intermediate form of anorectal malformation (ARM) and three had a high form of ARM. The presacral masses consistent with Currarino triad included anterior meningocoele in three patients, lipoma in two patients and anterior lipomeningocoele in one patient. MRI diagnosed tethering of the spinal cord in four of six patients. The tethering of the spinal cord was due to a lipomeningocoele in one patient, an intradural lipoma in one patient and a lipoma of the filum in two patients.
Conclusion. The association of Currarino triad with tethered spinal cord seems more common than generally reported in the literature. Preoperative MRI of the lumbosacral spine is essential to detect significant myelodysplasia in all patients with Currarino triad.

Introduction

Currarino triad is a rare association of congenital anal stenosis, sacral defect and a presacral mass described by Currarino et al. in 1981 [1]. The association of anorectal malformation (ARM) and tethered cord syndrome has been more frequently reported [2]. MRI has been recognised as the procedure of choice to study congenital anomalies of the paediatric spine [3]. The purpose of the present study is to discuss the role of MRI in the diagnosis of the various expressions of the Currarino triad and to study the association of tethered spinal cord in patients presenting with this triad.

Materials and methods

Among 55 patients seen at our institution for ARM between 1985 and 1995, six patients, four boys and two girls, with a mean age of 6 years (range 7 months–14 years) presented with ARM, sacral agenesis and a presacral mass and were included in the study. All patients were imaged using 0.35-T or 1.5-T MRI. The scan sequences obtained were T1-weighted (600/20), T2-weighted (2500/80) and proton density (2500/20) spin-echo sequences in the coronal, sagittal and axial planes. In selected cases, T1 W (600/20) images after i. v. injection of Gd-DTPA were also obtained. Conventional radiographs of the lumbar spine and sacrum were obtained and standard voiding cystourethrogram (VCUG), barium enema and/or fistulography were performed in all patients. Ultrasound and CT were also performed in two patients. Anomalies seen on conventional radiographs of the sacrum in all patients included anterior or anterolateral sacral hypoplasia (the so-called scimitar sacrum) (Fig. 1). One patient had been operated on at birth for a tracheo-oesophageal fistula. No patient had an associated cardiac or urinary tract malformation.

The MR imaging results regarding the level of atresia and the development of the levator muscles agreed with the operative find-

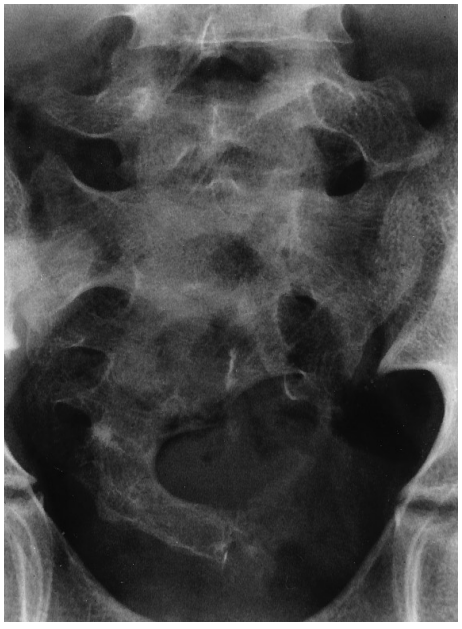


Fig. 1 AP view of sacrum of a 14-year-old girl. Note the left sacral dysplasia with a curved deviation to the right, so-called scimitar sacrum

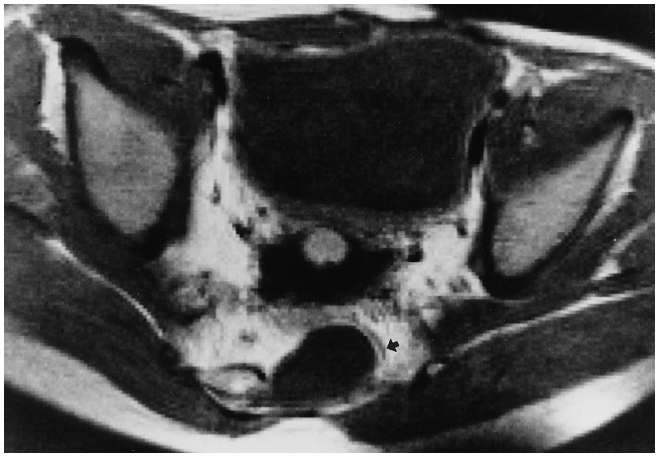


Fig. 2 Axial T1-weighted MRI at the level of the tip of the sacrum showing left sacral agenesis and a left anterolateral meningocoele (*arrow*)

ings in all patients. Three patients had an intermediate form and three patients presented with a high form of ARM.

All patients were operated on by pull-through procedures according to Peña or Cywes for ARM and three out of six patients underwent a posterior laminectomy to untether the spinal cord. One patient had a combined pelvic and posterior sagittal approach for a complex form of anterior lipomeningocoele. The clinical follow-up included anorectal manometry and urodynamic studies in four patients.

Results

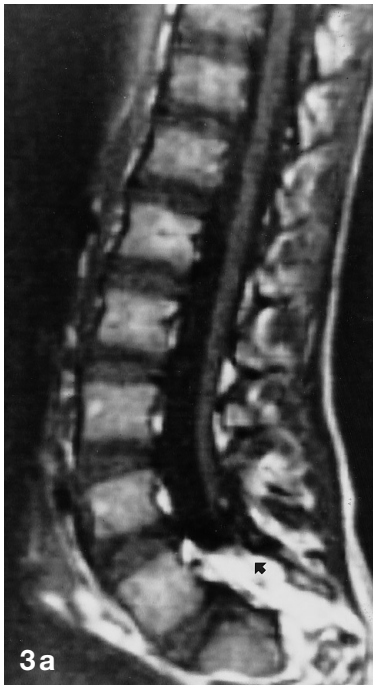
The clinical, MRI and follow-up findings in our patients are summarised in Table 1. One patient had a family history of intermediate ARM in a sibling. An anterior meningocoele was present in three patients (Fig. 2) and the characteristic hyperintense signal of a presacral lipoma was noted in two patients (Fig. 3). The T2-weighted image of the spinal cord in a 7 month-old girl showed a presacral mass of mixed signal intensity and a low-lying conus with its inferior end tethered by a lipomeningocoele which included hypointense and hyperintense elements communicating with the subarachnoid space (Fig. 4). MRI disclosed the presacral mass consistent with the diagnosis of Currarino triad in all patients. Tethering of the spinal cord was diagnosed by MRI in four out of six patients, and the intraoperative findings of the lesions causing tethering of the spinal cord did not differ from the MRI findings. Lesions causing tethering of the spinal cord included lipomeningocoele (one patient), intradural lipoma in one patient (Fig. 3 a) and lipoma of the filum (two patients) (Fig. 5).

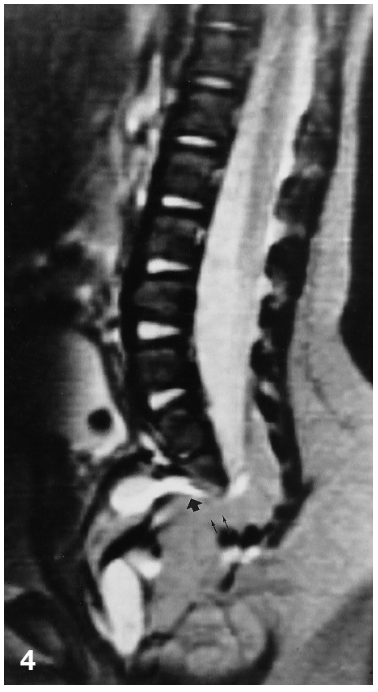
Motor dysfunction of the lower limbs was present in one patient, but improved following anorectal surgery, posterior laminectomy and orthopaedic correction of hip dysplasia. Following surgery, faecal continency was improved in five out of six patients. Urodynamic studies were available in four patients and showed improvement in two patients. Persisting signs of a neurogenic bladder were present in one patient and a second intervention was performed to untether the filum after worsening of the urodynamic studies in another patient.

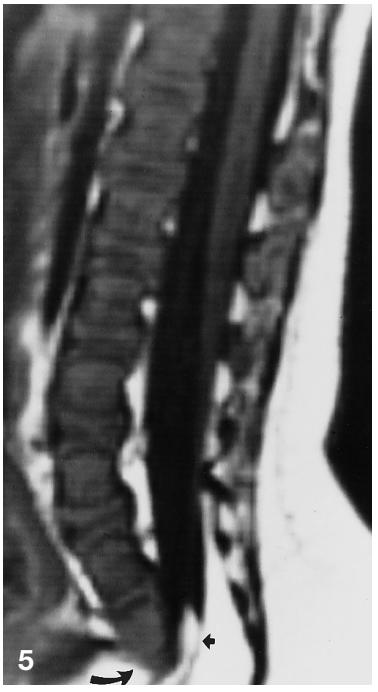
Discussion

Anorectal malformations are a common form of congenital malformation in children. The estimated incidence of this condition is 1 in 5000. In addition, many malformations of the gastrointestinal, urogenital and cardiovascular system are associated with ARM. Although an association between ARM, sacral agenesis and anterior meningocoele was reported as early as 1926 [4], Currarino et al. [1] recognised these disorders as a new entity explained by a common embryogenesis. The three main features of Currarino triad are an anterior sacral defect, congenital anorectal stenosis and a presacral mass, which may be a meningocoele, a teratoma, an enteric cyst or a combination of these [1]. In our series, the presacral mass consisted of anterior meningocoele in three patients, lipoma in two patients and anterior lipomeningocoele in one patient.

The Currarino triad has been recognised as an autosomal dominant inherited disease in about 50 % of the patients [1], and this diagnosis should prompt evaluation of family members. Surprisingly, only one of our pa-







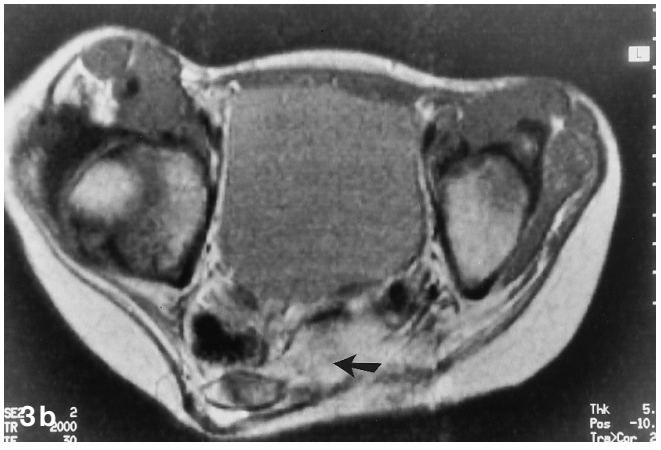


Fig. 3 **a** Midsagittal T1-weighted MRI of the spinal cord in a 9-year-old girl showing a low-lying conus with posterior and inferior tethering by an intradural lipoma (*arrow*). **b** Transverse T1 image at the level of the tip of the sacrum showing a persisting left presacral lipoma (*arrow*) following partial resection

Fig. 4 Midsagittal T2-weighted MRI of the spinal cord in a 7 month-old girl showing partial agenesis of the sacrum and a low-lying conus with inferior tethering by a lipomeningocoele which includes fatty elements (*small arrows*) and communicates with the subarachnoid space (*large arrow*)

Fig. 5 Midsagittal T1-weighted MRI image of the spinal cord in a 1-year-old girl showing partial agenesis of the sacrum (*curved arrow*) and a low-lying conus which is tethered by a lipoma of the filum (*arrow*)

tients showed a positive familial history of intermediate ARM in a sibling. Several explanations have been reported for the embryogenesis of this triad. Currarino et al. [1] proposed that abnormal endoectodermal adhesions and notochordal defects in early fetal life result in a fistula between the gut and the spinal canal with enteric elements ventrally and neural elements dorsally. This entity would be a variant of the split notochord syndrome. The presence of both enteric, neuroectodermal and mesodermal elements is, however, required to explain the formation of presacral lipomas and teratomas [1–5]. The relatively high incidence of sacral abnormalities in patients with anorectal anomalies has only recently been recognised. Tunell observed, only in 1987, that there was a much higher incidence of sacral and neural tube abnormalities in patients with high anorectal malformations and caudal regression than in patients with a low imperforate anus [6]. The origin of occult spinal dysraphism of the lumbosacral region could be the same as that of ARM, because they arise at the same time of fetal development [2].

Urinary disturbance in patients with ARM have often been considered to be caused by anorectal or urogenital anomalies, sometimes obscuring the diagnosis of neurogenic bladder caused by the tethered spinal cord. However, recent reports have emphasised the association between anorectal malformations and tethered cord syndrome [7]. Among the reported causes of tethered cord are spinal lipoma, tight filum terminale, anterior sacral meningocoele and dermal sinus tract. In our series, lesions leading to tethering of the spinal cord included lipomeningocoele (one patient), intradu-

Table 1. Profile of six children with Currarino triad

Patient	Age (years)	Sex	MR findings		Surgical treatment	Follow-up (urodynamic studies and anorectal manometry)
			Presacral mass	Tethered cord		
1	14	M	Lipoma	Fibrolipoma of the filum	Peña/L5 laminectomy	Improved
2	9	F	Lipoma	Posterior intradural lipoma	Peña/L4–L5 laminectomy	Persistent neurogenic bladder
3	5	M	Meningocele	–	Cywes	Not available
4	7 m.	F	Lipomeningocele	Lipomeningocele	Combined pelvic and posterior approach	Worsening of urodynamic studies until second untethering of the cord
5	10	M	Meningocele	–	Peña/L5 laminectomy	Persisting fecal incontinence
6	1	M	Meningocele	Fibrolipoma of the filum	Peña	Not available

ral lipoma (one patient) and lipoma of the filum (two patients). Although the precise prevalence of tethered spinal cord in patients with Currarino triad is not known, several authors report a frequency of this association varying from one patient out of three [5] to three out of three [8]. In our series, MRI diagnosed tethering of the spinal cord in four out of six patients.

The lumbar spine, sacrum and pelvis may be imaged using a variety of imaging modalities. Spinal sonography of the neonate has been recently accepted to screen neonates for occult dysraphic lesions [9], and M-mode sonography has become an accepted study to help identify pathological motion patterns of the cord related to tethering or re-tethering [10]. Abdominal ultrasonography has been used mainly to screen for possible associated renal malformations, but the usefulness of conventional pelvic ultrasound in the preoperative diagnosis and staging of ARM has not been proven. Therefore, we believe that the use of spinal sonography should be restricted to the screening for tethered spinal cord in patients with Currarino triad, when normal skeletal maturation does not limit its visualisation. MR imaging is mandatory when the sonographic findings are equivocal or abnormal.

Myelography and CT have been used in the diagnosis and presurgical planning of Currarino triad [5]. However, multiple sacral and pelvic anomalies are often present in individual patients, and associated anomalies of the urinary and digestive tract may also occur. Therefore, it is important to image the entire lumbosacral spine and the pelvic region at the same time.

The ability to image the spinal cord and the abdomino-pelvic region in the sagittal and the coronal planes in a non-ionising and non-invasive way, and the ability to perform three dimensional display of the pelvic structures are definite advantages of MRI, which has been recognised as the modality of choice in the diagnosis and preoperative planning of Currarino triad [11–16]. Recent developments of phase contrast MR imaging show promising results [17] for the pre and postoperative demonstration of tethering or re-tethering of the spinal cord. These sequences were not available in our institution at the time of the study and the postoperative follow-up, when feasible, was done in our series by means of anorectal manometry associated with urodynamic studies [18, 19], which showed improvements in three patients.

The main goal after surgical treatment of children with ARM is to achieve as much faecal and urinary continence as possible. Since 1982, posterior sagittal anorectoplasty has emerged as a new approach for the treatment of ARM and allows treatment of all types of defects with the best prognosis in terms of continence [20]. When tethering of the spinal cord is associated with ARM, we believe in early surgery to untether the cord, although there is no consensus about the timing for such surgery [2]. In our series, all patients were operated on using procedures according to Peña or Cywes, four out of six patients underwent a posterior laminectomy to untether the spinal cord, and one patient underwent a combined pelvic and posterior sagittal approach for a complex form of anterior lipomeningocoele.

In conclusion, the association of tethered cord with Currarino triad seems to be under-recognised. Awareness of this association by the paediatrician and neurosurgeon should allow early untethering of the cord when necessary. Correction of the neurosurgical problems may lead to better management of the gastrointestinal and urinary lesions. We believe that preoperative MRI of the lumbosacral spine is essential to detect significant myelodysplasia in all patients with Currarino triad.

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