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The Currarino triad: What pediatric surgeons need to know☆

Amr Abdelhamid AbouZeid [a,*], Shaimaa Abdelsattar Mohammad [b], Mohammad Abolfotoh [c], Ahmed Bassiouny Radwan [a], Mohamed Mohamed ElSayed Ismail [c], Tarek Ahmed Hassan [a]

[a] Pediatric Surgery Department, Faculty of medicine, Ain-Shams University
[b] Radiodiagnosis Department, Faculty of medicine, Ain-Shams University
[c] Department of Neurosurgery, Faculty of medicine, Ain-Shams University

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ABSTRACT

Purpose: We report our experience in managing a group of patients with Currarino syndrome, highlighting diagnostic challenges, surgical techniques, in addition to a review of current neurosurgical options.
Patients and methods: The study included patients with Currarino syndrome who presented to our pediatric surgery department during the period 2010 through 2016. The 'sacral scimitar' in plain X-ray provided the clue for the diagnosis; while MRI examination was essential to define the nature of the presacral mass and associated spinal anomalies.
Results: The study included 17 patients (13 girls and 4 boys). Their age at presentation ranged from 7 months to 10 years. We used posterior sagittal approach to correct anorectal anomalies, and excise presacral cysts that were subjected to histopathological examination. Two cases presented with a pelvic abscess (infected presacral dermoid cyst), which were initially drained followed by excision.
The presacral mass consisted of either lipomyelocele (6), lipomyelomeningocele (3), or a developmental (dermoid) cyst (8). Tethering of the spinal cord was a common association (70%)
Conclusion: Apart from diagnostic challenges, the management of Currarino syndrome is similar to the usual management of ARM regarding the surgical approach and probably the prognosis that mainly depends on degree of associated sacral dysplasia.
Level of evidence: This is a case series with no comparison group (level IV).

The Currarino syndrome is a rare congenital disorder that refers to the association of three components: an anorectal malformation (ARM), a sacral vertebral bony defect, and a presacral mass [1]. Constipation is the presenting symptom in the majority of cases [2–5]. The commonly reported presacral masses are either some sort of developmental cyst (dermoid, epidermoid, or tailgut), or an anterior sacral lipomyelomeningocele [3,6]. Cases may present with the complete form, or may have one of the three components missing [5]. Spinal cord anomalies (tethered cord, thickened filum, syrinx) are common associations, and duplication of the urogenital tract has also been described with the triad [5].

Early descriptions of the triad were reported in the literature by Kennedy in 1926 [7] and Aschraft in 1968 [8]; however, in 1981, Currarino, et al., reported this association as a unique syndrome with autosomal dominant inheritance, and suggested an embryological explanation for its occurrence [9]. Although early reported cases had been shown to run in families, more sporadic cases have recently been described.

The Currarino triad is believed to increase the challenges of managing cases of ARM [10,11]. Commonly, the integrated efforts of pediatric surgeons, radiologists, neurosurgeons, and pathologists are required. Here, we report our experience in managing a group of patients with Currarino syndrome, highlighting the diagnostic challenges, surgical techniques, and current neurosurgical options.

1. Patients and methods

The study included patients with the diagnosis of Currarino syndrome who presented to our pediatric surgery department from 2010 through 2016. The diagnosis of Currarino syndrome was established by identifying the following three components: 1) an anorectal anomaly; 2) sacral vertebral anomalies; and 3) a presacral mass (dermoid cyst and/or anterior lipomyelomeningocele).

Patients presented with either an imperforate anus (ARM), constipation, or pelvic abscess (suppuration on top of a presacral dermoid cyst, Fig. 1). Some patients presented with fecal incontinence after the repair of an ARM elsewhere. We performed operations to correct anorectal anomalies and excise presacral cysts, which were

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* Corresponding author at: Lotefy El-Sayed Street, 9 Ain-Shams University buildings, Abbassia, Cairo 11657, Eygpt. Tel.: +20 1116560566; fax: +20 224830833.
E-mail address: amrabdelhamid@hotmail.com (A.A. AbouZeid).

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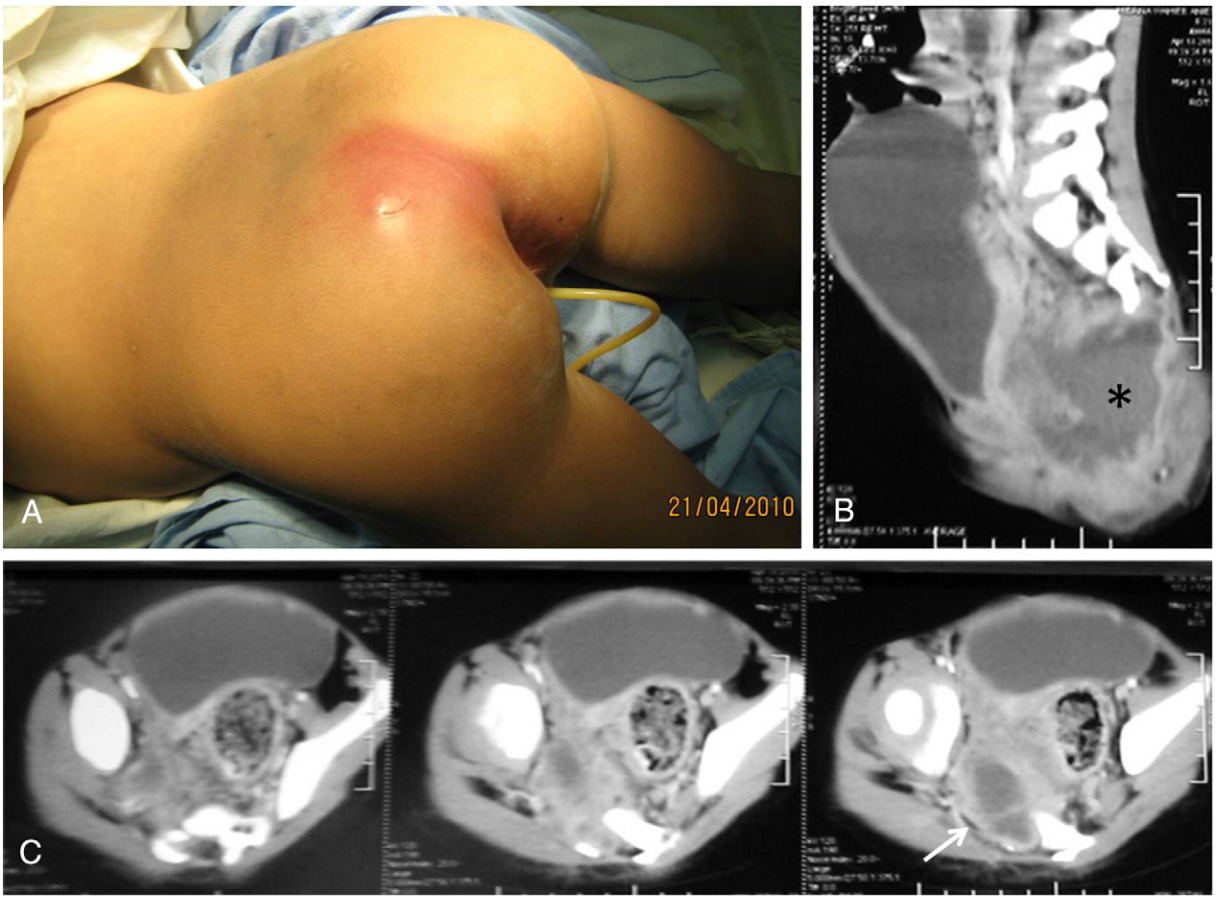

Fig. 1. A 34-month-old female presenting with pelvic abscess proved to be super-added infection on top of a presacral dermoid cyst as a part of Currarino Syndrome: A) signs of suppuration in the gluteal region; B) midsagittal CT scan showing large pelvic abscess cavity (asterisk); C) serial axial sections showing the abscess cavity extending into the sacral bony defect (white arrow).

sent for histopathological examination. In some cases, the diagnosis of Currarino syndrome was initially missed, only to be discovered later during or after the operation (Fig. 2). Usually, the 'sacral scimitar' seen on plain X-ray films was the clue to the diagnosis (Fig. 3), while magnetic resonance imaging (MRI) was essential in the workup of these patients to define the nature of the presacral mass and the associated spinal cord anomalies (Fig. 4). Unfortunately, genetic counseling was not available for this study group, although a familial background was not apparent in any of these cases. Owing to the retrospective nature of the study, an IRB number was not required, and the study was approved through expedited review by the scientific committee.

1.1. MRI technique

MRI of the pelvis and spine was done with a 1.5-T magnet (Achieva, Philips Healthcare, Eindhoven, The Netherlands). For better image resolution, we used a dedicated phased-array surface coil (cardiac or torso coil according to the patient's size). For infants and uncooperative children, sedation with chloral hydrate was usually sufficient; otherwise, the examination was done under general anesthesia.

Multiple pulse sequences were acquired in different planes. For pelvic examination, the following sequences were obtained: axial T1WI, axial T2WI (with and without fat suppression), coronal T2WI, and sagittal T2WI (with and without fat suppression). Complementary sagittal images (T1WI and T2WI) of the lumbosacral spine were also necessary to detect possible associated spinal anomalies.

1.2. Surgical technique

In this study, the principle surgical procedure was concerned with correcting the ARM, in addition to excising co-existing presacral cysts. The posterior sagittal anorectoplasty procedure (PSARP), first introduced by DeVries and Pena in 1982 [12], was used to correct a high-type ARM (rectourethral fistula), while a limited PSARP was the technique of choice for the more common low types (rectoperineal/vestibular fistula).

In three female patients who presented with a low-type ARM, we extended the limited sagittal incision more posteriorly (similar to a standard PSARP) to allow concomitant excision of the coexisting presacral masses. In two cases, the procedure was done primarily without a covering colostomy, while a colostomy was done initially at birth for one patient with a neonatal intestinal obstruction (case 1, Table 1). Those without a covering colostomy were admitted two days before the operation for bowel preparation.

First, a circumferential incision is made around the abnormal anus (whether an anteriorly displaced anus, or a narrow 'funnel' anus). Another posterior sagittal incision is made in the midline extending from the anus to the coccyx. This second incision is deepened to expose the posterior rectal wall, which is identified by its characteristic covering fascia. Dissection continues all around the anorectum, separating it from the surrounding attachments so that it can be repositioned backwards to the normal site of the anus. Care should be taken to proceed in the correct plane of dissection, keeping strictly outside but close to the rectal wall to prevent injury to other pelvic

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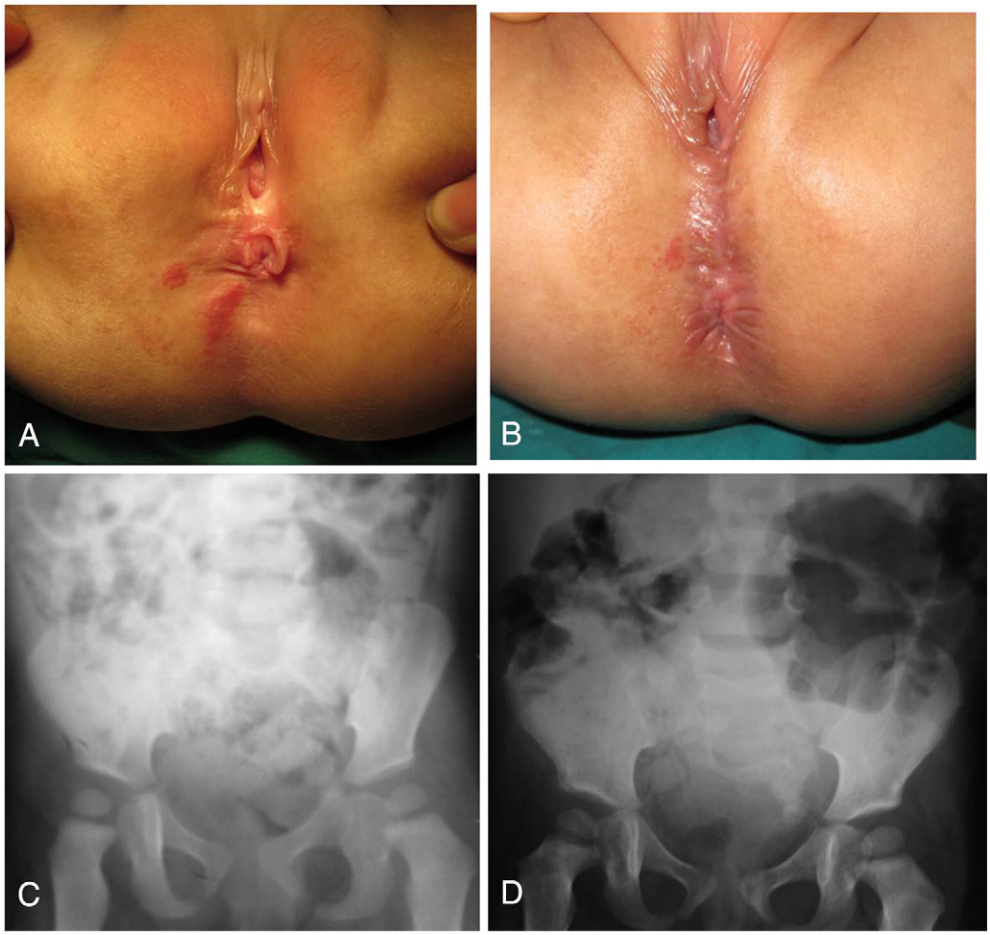

Fig. 2. A 24-month-old female presenting with constipation and ventrally displaced anus (A). B) Postoperative picture of the same patient after limited PSARP to correct the position of the anorectum. C) Preoperative plain X-ray anteroposterior view showing the stool load in the pelvis masking the sacral defect. D) The 'scimitar' sacral deformity in the postoperative plain X-ray has become apparent after correction of constipation, providing the clue for the diagnosis of Currarino syndrome.

structures [13]. Finding this plane is easier by starting from the back of the rectum with an incision through the perirectal fascia. Dissection then continues on the lateral walls of the rectum and, finally, the rectum is separated from the more adherent anterior structures (the vagina).

Effective mobilization of the anorectum is important to prevent postoperative anal retraction and wound dehiscence, especially when operating without a covering colostomy [14]. Moreover, mobilizing the anorectum facilitates exposure of the presacral space. Excision of the coexisting presacral cyst is now feasible. Care should be taken during excision of the cyst to prevent injury to the posterior rectal wall (inserting a metal dilator into the rectum at that time may be of help, Fig. 5C). Release of the deep attachments to the anterior of the sacrum is facilitated by applying outward traction on the cyst. Not uncommonly, these deep attachments contain an anterior tongue-like projection of the thecal sac. This projection must be identified and carefully closed during the operation; otherwise, the condition may be complicated by postoperative leakage of cerebrospinal fluid (CSF) from the surgical wound. Intraoperatively, the presence of pulsating clear CSF deep in the sacral bony defect can help define the opening in the meninges. Closure of the meningeal defect may be challenging because of its deep location, which requires generous exposure (Fig. 5D).

For an anteriorly displaced anus and after the excision of the presacral dermoid cyst, the anorectum is repositioned backwards to its normal position in the center of the sphincter-muscle complex. In patients with anorectal stenosis, the terminal stenosed bowel segment is excised, and normal-caliber bowel is pulled down and anastomosed to the skin at the normal site of the anus.

2. Results

Our study included 17 patients (13 females and 4 males) with the diagnosis of Currarino syndrome. Their age at presentation ranged from 7 months to 10 years (mean 39.7 months; median 24 months). A limited PSARP was performed for three patients with a rectoperineal/vestibular fistula, while a formal PSARP was needed in four patients: one male with a rectourethral fistula and three females to facilitate the excision of coexisting presacral dermoid cysts. We did not see postoperative wound complications except in the male patient, who had a postoperative CSF leak from the posterior sagittal wound (from an unnoticed iatrogenic injury to an associating anterior meningocele). This complication required reexploration of the wound to close the meningeal defect (case 17, Table 1). Two patients presented with a pelvic abscess (infected presacral dermoid cyst), which was initially drained and then excised. Six patients were referred to us with problems of constipation or fecal incontinence after undergoing surgery elsewhere for ARM. The remaining two patients are still being prepared for their operations. The postoperative follow-up period ranged from 5 months to 6 years (mean 34 months; median 37 months), with four patients lost to follow-up. The clinical data of the 17 patients are summarized in Table 1.

2.1. Anorectal anomalies

Most patients had minor, 'low' types of ARM. Nine (8 females, 1 male) had a rectoperineal fistula (Fig. 2A); two females had the characteristic 'funnel' anus (congenital anorectal stenosis); three

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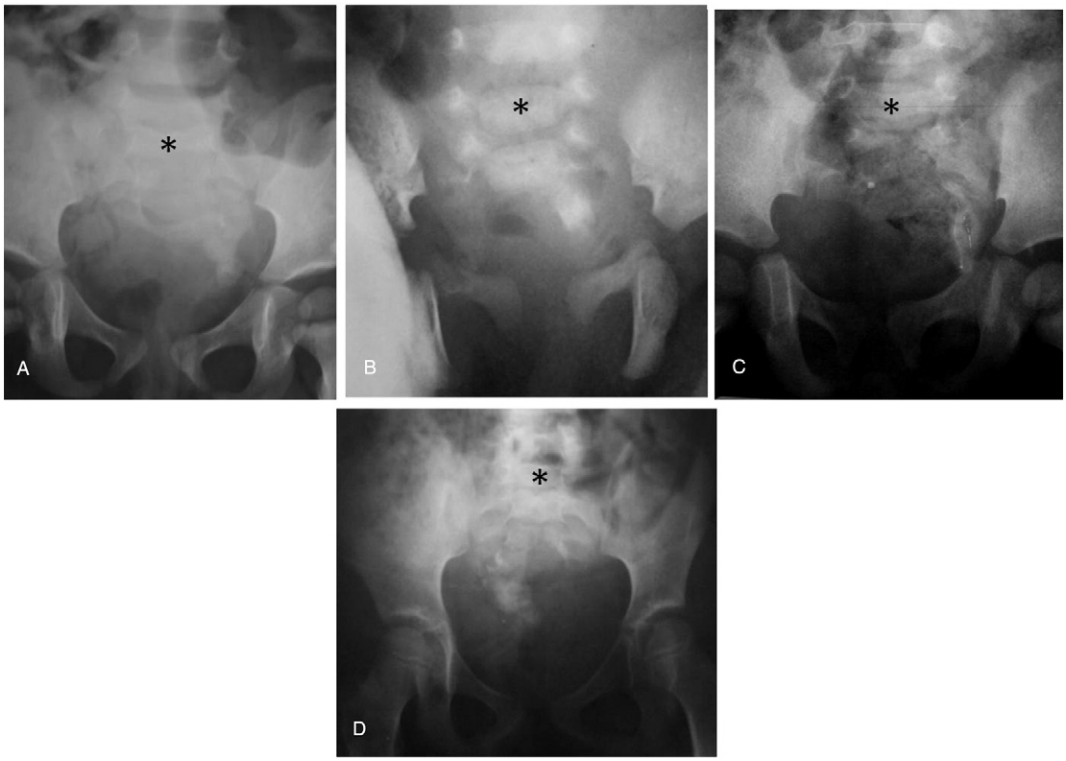

Fig. 3. Variable degrees of the sacral defect (sacral scimitar) characteristic of the Currarino syndrome; the asterisk (*) is marking the first sacral vertebra: A) small right defect sparing the upper three sacral vertebrae; B) right defect sparing the upper two sacral vertebrae; C) large sacral defect sparing only the first sacral vertebra; D) a case of caudal duplication syndrome with the sacral defect being more or less central.

females had a rectovestibular fistula; and three of the four males had a rectourethral fistula.

2.2. Sacral bony vertebral abnormality

In most patients (15/17 cases; 88%), the hemisacral vertebral bony defect resulted in the typical notched appearance of the sacrum (sacral scimitar on the plain X-ray anteroposterior view), which presented the key to the diagnosis of Currarino syndrome (Fig. 3). However, the diagnosis could be easily missed initially because of constipation, which may obscure the sacral deformity because of the shadow of loaded stools in the rectum (Fig. 2).

In the 15 patients with the characteristic sacral scimitar, the size of the sacral defect ranged from a small defect affecting only the last sacral vertebra (one case), a medium-sized defect involving the last two or three sacral vertebrae (11 cases), to a larger defect affecting the last four sacral vertebrae (3 cases). The sacral notch was on the right side in 9 cases, the left side in 5, and, in one patient with caudal duplication, the defect appeared more or less central (Fig. 3).

2.3. The presacral mass

The presacral mass was composed of a lipoma as part of an anterior lipomyelocele in six cases, lipomyelomeningocele in three, and a developmental cyst in eight (six of which were excised and had a pathological

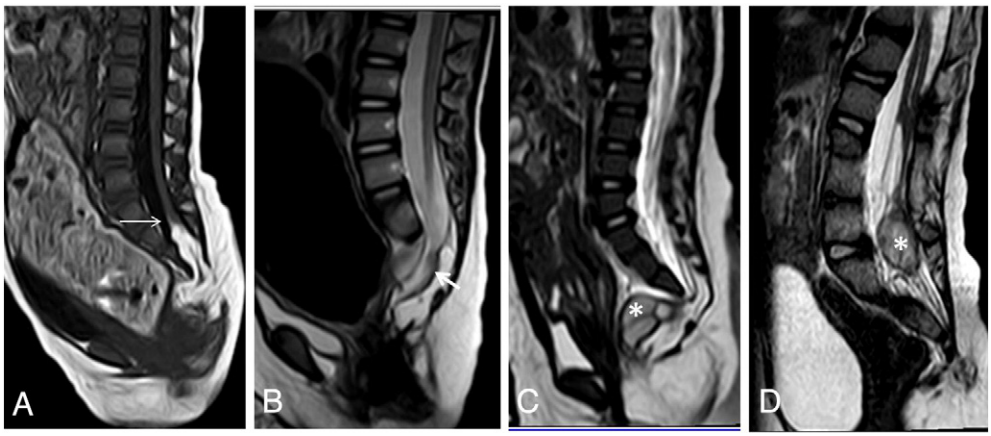

Fig. 4. Varieties of sacral masses seen with Currarino syndrome: A) anterior lipomyelocele with tethering of spinal cord characterized by the location of the placode lipoma interface (long white arrow) inside the spinal canal; B) anterior lipomyelomeningocele with the characteristic anterior bulging of the meningeal sac (short white arrow); C) presacral multiloculated dermoid cyst (asterisk); D) intraspinal dermoid cyst (asterisk) associated with cord tethering and syrinx. Note the hugely distended rectum in (A) and (B), as constipation is a common presenting symptom for the Currarino syndrome.

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diagnosis of mature cystic teratoma 'dermoid cyst'). In one case, the developmental cyst was intraspinal (Fig. 4D). The lipoma exhibited hyperintense signal on T1WI and T2WI (Fig. 4A), with the characteristic drop of signal on fat-suppressed T2WI. The anterior lipomyelocele was characterized by the location of the placode lipoma interface inside the spinal canal (Fig. 4A), while in cases with lipomyelomeningocele, the placode lipoma interface was seen outside the spinal canal, in addition to the characteristic anterior bulging of the meningeal sac (Fig. 4B).

The developmental 'dermoid' cysts had variable imaging characteristics. Commonly, they appeared as a multilocular cystic lesion with mixed signal intensity on T2WI (Fig. 5A), and with a hypointense signal on T1WI. An associated fatty component (which was hyperintense on T1WI) was seen in one patient. A small accompanying anterior tongue-like projection of the thecal sac exhibited hypointense signal on T1WI and hyperintense signal on T2WI. This structure could be differentiated from the loculi of the dermoid cyst by its uninterrupted continuity with the thecal sac (Fig. 5B). An infected dermoid cyst presented initially with a pelvic abscess in two patients; it appeared as a multilocular cystic lesion with a thick enhancing wall. Despite its close relation to the spinal canal through the sacral bony defect (Fig. 1), the infection was walled off and there were no manifestations of neurologic impact.

2.4. Spinal cord anomalies

Tethering of the spinal cord was common in our patients with Currarino syndrome (12 cases; 70.5%). Nine of these patients had a lipomyelocele or lipomyelomeningocele, while another three had a presacral dermoid cyst. High termination of the spinal cord was seen in one patient (caudal regression type 1), while normal conus termination with normal cord signal was seen in the remaining four patients with a presacral dermoid cyst. Spinal cord syrinx was seen in three patients (Fig. 4D).

Urinary incontinence was used rather than defecatory disorders as an indicator of the presence of associated neurological problems, as disorders of bowel control (constipation, fecal soiling/incontinence) may be more related to the repair of anorectal anomalies [6]. In our study group, five patients had disturbed bladder function in the form of urinary incontinence; all of them had significant sacral dysplasia (more than two dysplastic sacral vertebrae) (Table 2). The presence of disturbed bladder function appears to correlate with the degree of sacral dysplasia rather than spinal cord tethering. Cord tethering alone did not cause urinary dysfunction in the patients in our series; only four of the 12 patients with a tethered spinal cord (33.3%) complained of urinary incontinence, and all had significant sacral dysplasia.

3. Discussion

Currarino syndrome is a rare congenital disorder which a pediatric surgeon might face occasionally in his career. The available literature focuses mainly on case reports or 'retrospective' case series of relatively small numbers of patients collected over long periods of time. Some misconceptions can result from inaccurate observations, and may persist in the literature by being quoted and requoted by other authors, leading to several points of confusion in the management of these cases [11]. In our opinion, the introduction of MRI has revolutionized studying the anatomy of congenital anomalies by providing detailed images of serial sections of the body in multiple planes, with high soft-tissue resolution and without the risk of exposure to radiation. This modality can be applied in almost all cases in a safe and reproducible way, and presents a more recent and superior alternative to the traditional ways of studying the anatomy of congenital anomalies (autopsy studies on dead fetuses with congenital anomalies, and studies on animal models).

In this report, we tried to clear up some of the confusion that pediatric surgeons might face in the management of Currarino syndrome by discussing the debatable issues point by point, based on the best evidence from the literature and from studying our cases.

Constipation is the most common presenting symptom among patients with Currarino syndrome, and the most common postoperative gastrointestinal sequela [15]. The differential diagnosis includes idiopathic constipation and Hirschsprung disease [5]; however, clinical examination usually yields the clue to diagnosis by disclosing an abnormal anus (either anteriorly displaced, or stenotic). A plain X-ray of the sacrum (anteroposterior view) is the best way to screen for Currarino syndrome as it delineates the characteristic notched sacrum (scimitar deformity) [16]. The diagnosis can be easily missed, however, when a small sacral defect is masked by a rectum loaded with stools. The sacral notch indicates the presence of a presacral mass, which is best defined by MRI. Detecting the characteristic genetic defect of the disease is another more sophisticated way to reach the diagnosis [5]. Although prompt evaluation of other family members has been recommended [4], we believe that screening patients presenting with constipation or ARM might be more relevant.

The severity of constipation varies among patients with Currarino syndrome. It may lead to intestinal obstruction in some patients with anorectal stenosis, which may necessitate a colostomy at birth. Several etiological factors have been proposed to explain constipation in these patients: a narrow anal canal, anterior anal displacement, a pressure effect from a large presacral mass, and neurological factors [3,15]. Although we do agree on the multifactorial etiology of constipation in patients with Currarino syndrome, the effective repair of the anorectal anomaly may be considered one important and correctable factor that can cure constipation in these patients or at least make it more manageable [17].

Currarino syndrome was predominant among females with low-type ARM [15]. More than 50% of our patients had a rectoperineal fistula (9/17 cases), another 3 (17.6%) had a rectovestibular fistula, and only 2 (11.7%) had the characteristic 'funnel' anus (anorectal stenosis). The 'funnel' anus is a rare condition; however, whenever encountered, it is frequently seen in association with the Currarino triad (sacral scimitar and presacral dermoid) [16]. Four of our patients (23.5%) were males; three of them had a rectourethral fistula.

The surgical techniques are the standard techniques (PSARP/limited PSARP) used to repair an ARM. When a presacral mass is to be excised, it can usually be done in the same sitting of repairing the ARM, and through the same posterior sagittal incision. A preliminary colostomy is performed at birth for a high-type ARM, while a primary operation (without covering colostomy) is usually feasible for the more common low types, even when the concomitant excision of a presacral dermoid cyst is planned. In the latter situation, the necessity of a covering colostomy will depend on the previous operative experience of the pediatric surgeon in dealing with cases of ARM [14]. In our series, we achieved a successful outcome with both primary and staged procedures, with no postoperative wound complications except for one patient who had CSF leakage from the surgical wound owing to iatrogenic injury of an anterior meningocele, which passed unnoticed during the operation. This was managed by reexploring the wound and closing the meningeal defect.

The most common pathological diagnosis of the excised presacral masses in patients with Currarino syndrome is a dermoid cyst (also known as a mature cystic teratoma). Unlike other teratomas, presacral dermoids associated with Currarino syndrome are known for their low potential for malignant transformation [4,15,18]. However, excision is recommended to prevent other possible complications (mainly super-added suppuration 'pelvic abscess'). During excision, presacral dermoid cysts (unlike rectal duplications) are always separable from the posterior rectal wall [19]. On the other hand, care should be taken during dissection of the posterior attachments of the cyst, when an anterior projection of the thecal sac may be encountered. This projection must be identified and secured during the operation to prevent a postoperative CSF leak. A well-performed MRI of the pelvis can be very

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Table 1
Summary of the clinical, radiological, and operative data of the 17 cases with Currarino Syndrome.

No.	Age	Sex	Presentation	Currarino triad			Associated anomalies	Operative data
				Anorectal anomaly	Presacral mass + spinal anomalies	Sacral vertebral bony abnormality		
1	12 m	female	Neonatal low intestinal obstruction underwent colostomy	Anorectal stenosis (Funnel anus)	Dermoid cyst + anterior tongue-like protrusion of thecal sac; low lying conus with thickened filum + syrinx	Right scimitar (Dysplastic lower 3 sacral vertebrae, Fig. 3b)		Repair of the anorectal anomaly (PSARP) + Excision of the dermoid + successful intraoperative identification and closure of meningeal defect
2*	8 y	female	Fecal incontinence after repair of anorectal anomaly	Rectovestibular fistula	Lipomyelocele + tethered cord	left scimitar (Dysplastic lower 3 sacral vertebrae)		Operated elsewhere for vestibular anus
3	7 m	female	Constipation	Rectoperineal fistula	Lipomyelocele + tethered cord	left scimitar (Dysplastic lower 3 sacral vertebrae)		Repair of the anorectal anomaly (limited PSARP)
4*	4 y	female	Fecal incontinence after repair of anorectal anomaly	Rectoperineal fistula	Lipomyelocele + tethered cord + Syrinx	left scimitar (Dysplastic lower 3 sacral vertebrae)	***Urinary incontinence***	Repair of anorectal anomaly elsewhere
5	34 m	female	Pelvic abscess (infected dermoid cyst, Fig. 1) + postanal sinus	Mild anterior anal displacement (Rectoperineal fistula)	Dermoid cyst Normal spinal cord	Right scimitar (Dysplastic lower 2 sacral vertebrae)		Abscess drainage followed by delayed excision of residual sinus
6	10 y	female	Constipation	Mild anterior anal displacement (Rectoperineal fistula)	Dermoid cyst + tethered cord	left scimitar (Dysplastic lower 2 sacral vertebrae)		Excision of presacral dermoid cyst elsewhere during the neonatal period
7	24 m	female	Constipation	Rectoperineal fistula (Fig. 2)	Lipomyelocele + tethered cord (Fig. 4a)	Right scimitar (Dysplastic lower 2 sacral vertebrae, Fig. 3a)	Right vesicoureteric reflux	Repair of the anorectal anomaly (limited PSARP). Cystoscopic injection for right vesicoureteric reflux.
8	8 m	female	Constipation	Anorectal stenosis (Funnel anus)	Dermoid cyst (Fig.4c) + anterior tongue-like protrusion of thecal sac (Fig. 5a,b) + normal spinal cord	Right scimitar (Dysplastic lower 2 sacral vertebrae)		Repair of the anorectal anomaly (PSARP) + Excision of the dermoid cyst + successful intraoperative identification and closure of meningeal defect
9	24 m	female	Constipation + post anal sinus	Rectoperineal fistula	Dermoid cyst + Post anal inflammatory sinus + normal spinal cord	Right scimitar (Dysplastic lower sacral vertebra)		Repair of the anorectal anomaly (PSARP) + Excision of the dermoid cyst and post anal sinus

10	3 y	female	Vestibular anus	Rectovestibular fistula	Anterior lipomeningomyelocele (Fig. 3b) + tethered cord	Right scimitar (Dysplastic lower 4 sacral vertebrae)		Limited PSARP for ARM.
11	3 y	female	Pelvic abscess (infected presacral dermoid cyst)	Rectoperineal fistula	Dermoid cyst + normal spinal cord	Right scimitar (Dysplastic lower 2 sacral vertebrae)		Staged procedure: Colostomy + drainage of abscess. Delayed excision of residual sinus + correction of anal position. Closure of colostomy.
12	8 y	female	Caudal duplication (double anus + double external genitalia)	Rectoperineal fistula	Lipomyelocele + tethered cord	More or less a central sacral defect (Fig. 3d) (Dysplastic lower 3 sacral vertebrae)	Caudal duplication syndrome; ***Urinary incontinence;*** Bilateral vesicoureteric reflux with renal dysplasia (more on the left).	A staged surgical correction is planned for the complex urological anomalies
13	6y	Female	Fecal incontinence after repair of anorectal anomaly	Rectovestibular fistula	Intraspinal dermoid cyst + tethered cord + syrinx (Fig. 4d)	Right scimitar (Dysplastic lower 4 sacral vertebrae)	Esophageal atresia; Tetralogy of Fallot; ***Urinary incontinence***	Repair of anorectal anomaly elsewhere
14	2y	male	Urinary and fecal incontinence	Rectoperineal fistula	Developmental cyst + high cord termination D12 (caudal regression type 1)	Sacral dysgenesis (absent last 4 sacral vertebrae)	***Urinary incontinence***	Preparing for operation
15*	12 m	Male	Fecal incontinence after repair of anorectal anomaly	Rectourethral fistula	Lipomyelomeningocele + tethered cord	Left scimitar (Dysplastic lower 3 sacral vertebrae)	Absent left kidney	Operated elsewhere for imperforate anus
16*	17 m	Male	Fecal and urinary incontinence	Rectourethral fistula	Lipomyelocele + tethered cord	Right scimitar (Dysplastic lower 4 sacral vertebrae, Fig. 3c)	***Urinary incontinence***	Operated elsewhere for imperforate anus
17	9 m	Male	Imperforate anus	Rectourethral fistula	Lipomyelomeningocele + tethered cord. Sacral dermal sinus	Sacral dysplasia (S3 hemivertebra; absent S5)		Repair of the anorectal anomaly (PSARP), complicated by postoperative CSF leakage from the operative wound owing to iatrogenic injury of an anterior meningocele which passed unnoticed during the operation. This was managed by reexploring the wound and closure of the meningeal defect.

The asterisk (*) marks patients who were lost to follow up.

helpful in identifying such a condition before the operation. The pediatric surgeon should be prepared to close the meningeal defect; otherwise, attendance by a neurosurgeon during the operation is necessary. Ideally, dural closure should be done with nonabsorbable monofilament sutures, which are used to decrease the risk of infection. The extra knotting needed with such suture material, however, may cause some inconvenience during repair through an anterior approach. Instead, we used polyglactin to close the dural defects in these settings. An intraspinal dermoid cyst is a quite rare and difficult situation, and its excision will always cause some degree of neurological deficit (especially if located in the conus medullaris). However, patients with an intraspinal dermoid cyst will most likely need surgery to prevent acute paraplegia that can happen if the condition becomes complicated by a superimposed infection of the contents of the cyst [20].

The decision for surgery on anterior sacral meningoceles associated with Currarino syndrome depends on several factors: the size of the sac, the position and width of its neck, whether it is associated with other presacral masses, and the sex of the patient [21–24]. The classic practice has been to repair the meningocele before correcting the anorectal anomalies to avoid the risk of meningitis [3,19,25–28]. However, many surgeons nowadays describe ligating and excising the sac in the same session when using the posterior sagittal approach. The ideal situation for such primary excision is a large sac compressing the rectum with a low, accessible, narrow neck [11,22,28–30]. Asymptomatic small or incidentally discovered sacs should be managed conservatively (no intervention). In such cases, we recommend obtaining a baseline MRI of the spine that can be repeated any time the patient develops any symptoms of concern (headache with a low intracranial tension pattern, manifestations of meningeal irritation, or progressive unexplained urinary or defecatory dysfunction) [25].

Anterior sacral meningoceles occurring in the female is an old, well-known obstetric/gynecological dilemma [7]. Although Currarino syndrome occurs with an autosomal-dominant inheritance pattern, it shows the female predominance trend (perhaps owing to associated gynecological and urinary-tract problems seen in women) [31]. Nevertheless, some huge anterior sacral meningoceles may pass unnoticed in males [25]. Medium-sized sacs in females can be managed conservatively [27,30]; however, if the patient plans to become pregnant, one would consider offering excision of these medium-sized sacs to avoid the increased risk of obstructed labor or rupture of the meningocele. If a patient with a medium-sized or large anterior sacral meningocele becomes pregnant, delivery through cesarean section should be advised with subsequent elective excision of the meningocele before the next pregnancy [24,27,31,32]. Based on our review of the literature, we recommend that every female with a moderate or large anterior sacral meningocele should be offered repair of that lesion before any attempt to conceive [6].

Although a tethered spinal cord is commonly associated with Currarino syndrome, its contribution to the symptomatology of these cases is not clear and has never been confirmed in any study. Lee et al.

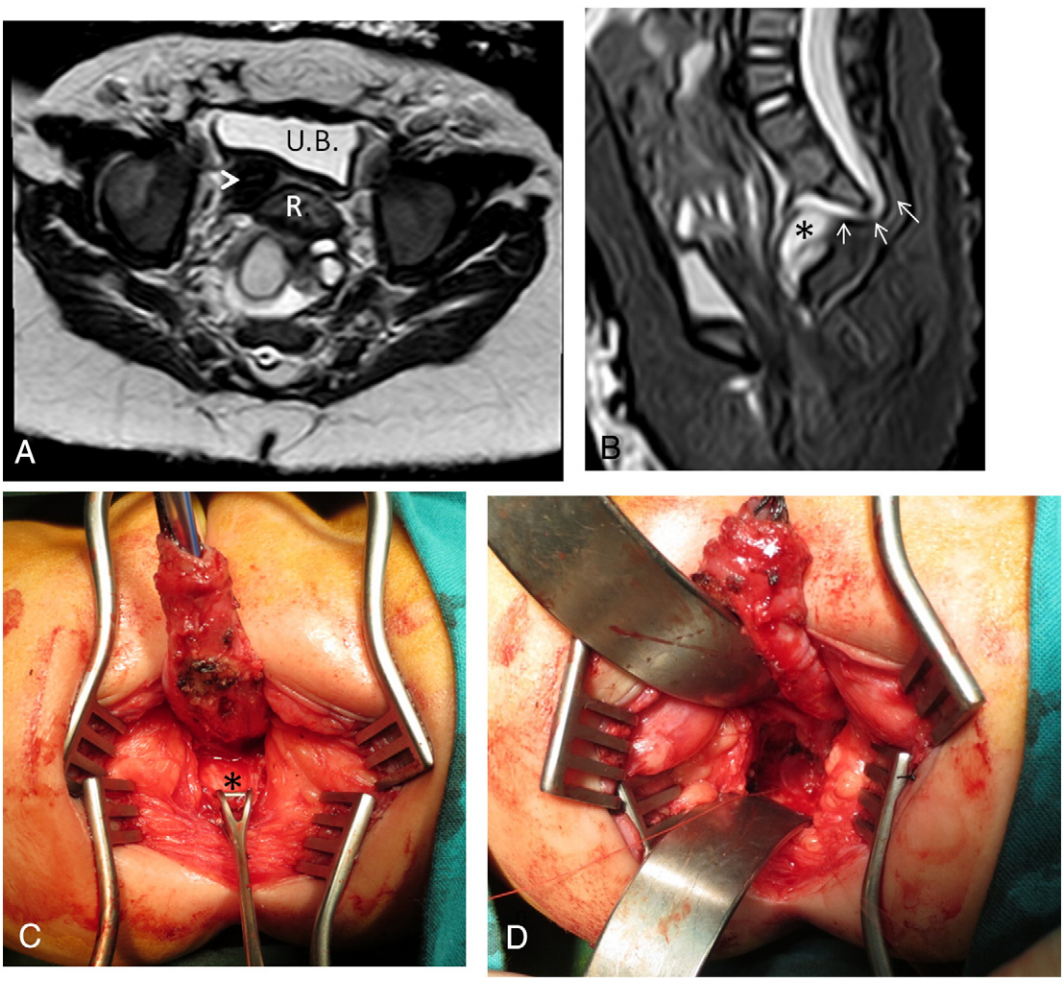

Fig. 5. An 8-month-old female patient with Currarino triad. A) Axial T2WI showing the urinary bladder (U.B.), uterus (arrow head), rectum (R), and the postrectal (presacral) multiloculated dermoid cyst with mixed signal intensity on T2WI. B) Midsagittal T2WI (with fat suppression) showing a small anterior tongue-like projection of the thecal sac exhibiting hyperintense signal on T2WI (white arrows), which could be differentiated from loculi of the dermoid cyst (asterisk) by its uninterrupted continuity with the thecal sac. C, D) Operative photos of the posterior sagittal approach to repair ARM and excise presacral mass: C) mobilization of the anorectum, a Bapcock forceps is used to apply traction on the presacral cyst (asterisk); D) closure of the meningeal defect after excision of presacral cyst.

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Table 2
Correlation between the degree of sacral dysplasia and the presence of urinary bladder dysfunction (indicator for neurological disability).

Number of dysplastic hemi/absent sacral vertebrae	Number of patients	Presence of bladder dysfunction (urinary incontinence)
1 vertebra	one case	–
2 vertebrae	6 cases	–
3 vertebrae	6 cases	2/6 (33.3%)
4 vertebrae	4 cases	3/4 (75%)

reported on the urodynamic findings in 12 patients with Currarino syndrome who underwent spinal cord detethering. While 10 patients had persistent voiding difficulties postoperatively, only three of five patients who underwent urodynamic testing before and after surgery demonstrated improvement [15]. Moreover, the authors referred to similarities in the urodynamic findings among patients with Currarino syndrome and those with isolated sacral agenesis [15]. Hence, it appears that there is no great indication for prophylactic spinal cord detethering [33]. One could follow these patients closely for worsening gastrointestinal/urinary symptoms or any deterioration in the form of new lower-limb complaints (weakness, or paresthesia) [6]. If there is an indication to operate on the anterior sacral meningocele, cord detethering should always be considered in the same sitting [34]. In our series, there was no case with a clear indication to remove the associated anterior sacral meningocele or to detether the spinal cord.

Although Currarino syndrome appears to be a complex disease with a variety of presacral masses (dermoid cyst, lipoma, meningocele) in addition to associated spinal-cord anomalies, it is the dermoid cyst (mature cystic teratoma) that is likely to cause complications and should be excised. Other conditions, like presacral lipomas, meningoceles, and a tethered cord, can usually be managed conservatively (no intervention). The surgical approach is more or less similar to that used for the repair of an ARM, which can be done through a single or staged procedure (according to the clinical situation and the surgeon's previous experience with ARM). During excision of the presacral dermoid, however, the pediatric surgeon should be prepared to deal with a possible communication with the thecal sac.

Our study is limited by its retrospective nature and small number of patients, factors that should be expected with such a rare anomaly. The study also lacks comprehensive urodynamic studies to precisely distinguish the denervation potentials in these patients [15], who will still need long-term follow-up for any possible neurosurgical consequences [6].

4. Conclusion

Apart from diagnostic challenges, the management of Currarino syndrome is more or less similar to the usual management of ARM regarding the surgical approach, and probably the prognosis, which seems to depend mainly on the degree of associated sacral dysplasia. A presacral dermoid cyst is likely to lead to complications and is better excised. The excision can be done concomitantly during the repair of the ARM through the same posterior sagittal incision.

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