J Korean Radiol Soc 1997;37:1127—1133

MR Imaging of the Currarino Triad[1]

Ji Hye Kim, M.D., Ji Eun Kim, M.D., In-One Kim, M.D.[2], Hee Jung Lee, M.D.[3]
Young Seok Lee, M.D., Tae Hoon Lee, M.D.[4], Hyung Sik Kim, M.D.

Purpose : The purpose of this study was to describe the MR findings of the spectrum of the Currarino triad and to discuss the potential role of MR imaging in evaluating these anomalies.

Materials and Methods : Seven children (age range : 2—12 months) with Currarino triad were evaluated using MR imaging, plain radiography, and barium study. In addition, CT scans (n=3) and sonography (n=2) were performed. We retrospectively analyzed MR imaging findings and correlated these with the findings of other imaging modalities.

Results : Anorectal anomalies included anorectal stenosis in five patients and an imperforate anus in two. MR imaging findings of anorectal stenosis included an elongated thick-walled anorectal canal and dilatation of the proximal segment of the rectum. In the patients with an imperforate anus, the location of the blind rectal pouch and sphincteric musculature was delineated. In one case, a transcolostomy enema revealed a fistula not evident on MR images. Presacral masses included four teratomas and three lipomas associated with various spinal anomalies. On MR imaging, which gave better results than CT or sonography, a detailed evaluation of presacral masses and associated anomalies was possible. Sacral anomalies included a typical scimitar-shaped sacral defect in five patients, abnormal curvature in one, and malsegmentation in one. In all cases, MR imaging showed the abnormal sacrum, but plain radiography more clearly demonstrated its anomalous shape.

Conclusion : Various anorectal anomalies, presacral masses, and other associated anomalies were demonstrated by MR imaging. When the Currarino triad is suspected, MR imaging should therefore follow plain radiographs.

Index Words : Anus
Sacrum
Magnetic resonance(MR), in infants and children
Children, gastrointestinal tract

The Currarino triad is a very rare complex of congenital anomalies explained by a common embryogenesis, and first described by Currarino et al. in 1981 (1). The three main components are congenital anorectal stenosis, sacral defect, and presacral mass including meningocele, teratoma, enteric cyst, or a combination of these. Autosomal dominant inheritance has been found in about 50% of cases (2).

Imaging examinations play a key role in establishing and defining the multiple aspects of these complicated anomalies. In most reported cases in the past, barium enema examination, CT scanning, and myelography have been the main diagnostic modalities (1—3). Recently, MR imaging of the Currarino triad was reported in a small series, as the next step for detection of

[1]Department of Diagnostic Radiology, Chung-Ang Gil Medical Center
[2]Department of Radiology, Seoul National University, College of Medicine
[3]3Department of Radiology, Keimyung University School of Medicine, Dongsan Medical Center
[4]Department of General Surgery, Chung-Ang Gil Medical Center
Received March 31, 1997 ; Accepted July 29, 1997
Address reprint requests to : Ji Hye Kim, M.D., Department of Diagnostic Radiology, Chung-Ang Gil Medical Center # 1198 Kuwal-Dong Namdong-Gu, Incheon 405-220, Korea. Tel. 82-32-460-3058/3061 Fax. 82-32-460-3055

presacral mass after the diagnosis of sacral defect and anorectal anomaly (4). MR imaging has also been used for preoperative evaluation of anorectal anomalies (5, 6).

We retrospectively reviewed MR imaging and other radiological findings in seven cases of Currarino triad. The purpose of this study was to describe the spectrum of anomalies demonstrated on these MR images and to discuss the potential role of MR imaging in the diagnosis of the various anomalies.

Materials and Methods

Seven children, six boys and a girl, were shown to have congenital anorectal anomalies, abnormal sacrum, and presacral mass. Their ages ranged from two to 12 (mean, nine) months and two were sisters. Clinical presentation and individual anomalies are summarized in Table 1.

Table 1. Summary of Seven Cases of Currarino Triad

Case	1	2#	3#	4	5	6	7
Age/sex	11months/girl	8months/girl	5months/girl	2months/boy	9months/girl	7months/girl	6months/girl
Clinical presentation	Constipation Pull through op.*	Constipation	Frequent defecation	Constipation	Absent anal opening	Absent anal opening	Constipation
Triad anomalies	Anorectal stenosis, Sacral defect, Lipomyelo-schisis	Anorectal stenosis Sacral defect, Anterior MMC** with lipoma	Anal stenosis Abnormaly curved sacrum Teratoma	Anorectal stenosis Sacral defect Teratoma	Imperforated anus Malsegmen-tation of the sacrum Teratoma	Imperforated anus Sacral defect Teratoma	Anorectal stenosis Sacral defect Lipoma and thick filum
Associated anomalies	None	None	None	None	None	None	Multicystic dysplastic right kidney, Syringohy-dromyelia
Plain radiography	Scimitar sacrum	Scimitar sacrum	Accentuated sacrococcy-geal curvature	Scimitar sacrum	Irregular segmentation of the sacrum	Scimitar sacrum	Scimitar sacrum
Barium enema examination	Stenotic rectum with reversed recto-sigmoid ratio	Stenotic distal rectum	Stenotic distal rectum with posterior indentation	Stenotic distal rectum with posterior indentation	Low type malforma-tion with recto-vestibular fistula♣	Low type anorectal malforma-tion♣	Stenotic distal rectum
MR imaging	Pulled through intestine within the sphincter musculature Sacral defect Lipomye-loschisis	Thick, elongated anal canal Sacral defect Anterior MMC** with a presacral lipoma containing epidermoid cyst	Thick, elongated anal canal Accentuated sacrococcy-geal curvature Presacral fatty mass and cyst	Thick, elongated anal canal Sacral defect Fat containing presacral mass	Distal rectal pouch traced down to the ischial rami Abnormal sacral shape Presacral soft tissue mass attached to the rectal wall	Distal rectal pouch at the level of the ischial rami Sacral defect Presacral mass with fat component	Thick and elongated anal canal Sacral defect Presacral lipoma with thick filum
CT	Sacral defect Lipoma	Not performed	Not performed	Not performed	Not performed	Sacral defect Presacral mass	Sacral defect Lipoma
USG	Tethered cord to echogenic mass	Not performed	Not performed	Not performed	Presacral mass was not evident	Not performed	Not performed

Patient 2 and 3 are sisters.
♣ Instead of the barium enema, trans-colostomy water soluble enema was performed
* op. : operation ** MMC : myelemeningocele

In all patients, MR imaging, plain radiography, and barium or water soluble contrast enema were performed and a total of eight sets of MR images were obtained. Six of the eight MR scans were obtained for an initial evaluation of the anomalies. One was performed after abdominoperineal pull-through procedure, and the remaining one as a follow-up study after excision of the presacral mass. In addition, CT scanning (n=3) and sonography (n=2) were also performed for the evaluation of the presacral masses.

For MR imaging, a 0.38-T (Resonex 4000, USA), 0.5-T (Spectro Goldstar, Seoul, Korea), and 2.0-T superconducting units (Supertech, Goldstar, Seoul, Korea) were used. Contiguous sections, 3.5—6mm wide, were obtained in transaxial, coronal, and sagittal planes and the scanning of the abdomen and spinal canal was included. Patients were studied in the supine position with a 20—24 cm field of view using a 6×60 cm belt or surface coil. Spin-echo T1 (500—600/25), T2 (2000—3000/80—100), and contrast enhanced T1-weighted images were obtained after the injection of gadopentetate dimeglumine (0.1mmole/kg : Magnevist, Schering, Germany). All patients were sedated during scanning.

We reviewed the MR images of these patients and assessed the configuration of the rectum and anorectal canal, location of the blind rectal pouch, demonstration of sphincter musculature, fistula, abnormal sacral shape, extent and tissue characteristics of the presacral masses, and associated spinal and other anomalies. These findings were correlated with those of other imaging modalities.

Results

Anorectal anomalies

Anorectal anomalies included anorectal stenosis in five patients and an imperforate anus in two. In patients with anorectal stenosis, sagittal MR images directly visualized the anorectal canal itself, which had an elongated lumen with a thick wall. The rectum proximal to the stenotic segment was distended (Fig. 1). These findings were seen in all patients with anorectal stenosis except one, whose MR scan was performed after a Duhamel abdominoperineal pull through procedure. These MR findings of anorectal stenosis were comparable with the findings of barium enema examination. Because barium enema examination visualized only the mucosal surface of the rectum, it did not easily differentiate congenital megacolon involving a short segment. In this respect, MR findings of a thick and elongated anorectal canal

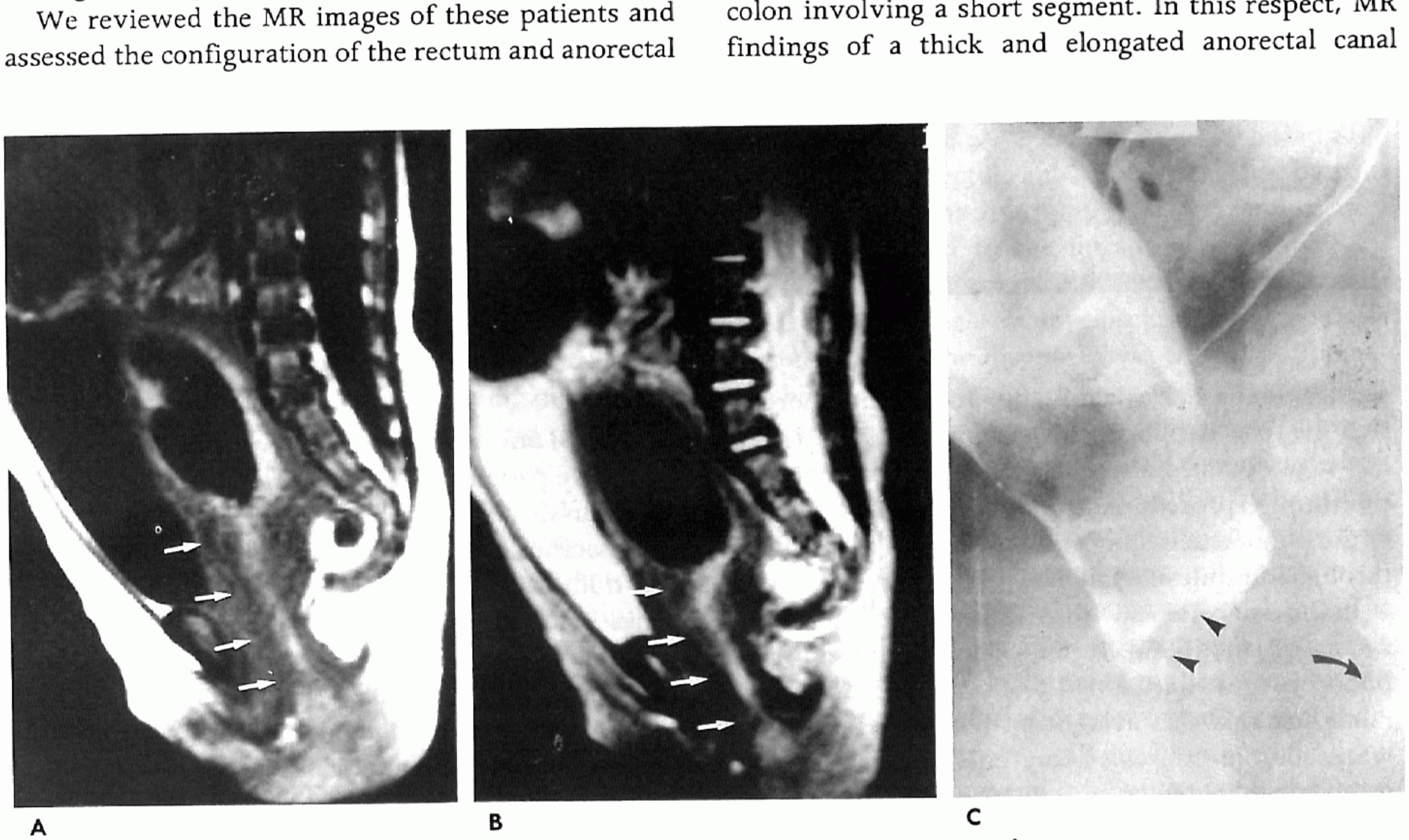

Fig. 1. A 5-month-old girl with anorectal stenosis, abnormal sacral curvature, and presacral teratoma.
T1 weighted (**A**) sagittal MR image shows presacral mass of high signal intensity with an internal hypo-intensity area, which is bright on T2 weighted image (**B**). Findings of distended rectum, elongated anorectal canal with a thick wall (arrows) can indicate anorectal stenosis.
C. Barium enema examination shows stenotic distal rectum comparing with proximal segment. The inferoposterior indentation of the rectal wall (arrowheads) and the abnormal sacrococcygeal curvature (arrow) suggest presacral mass.

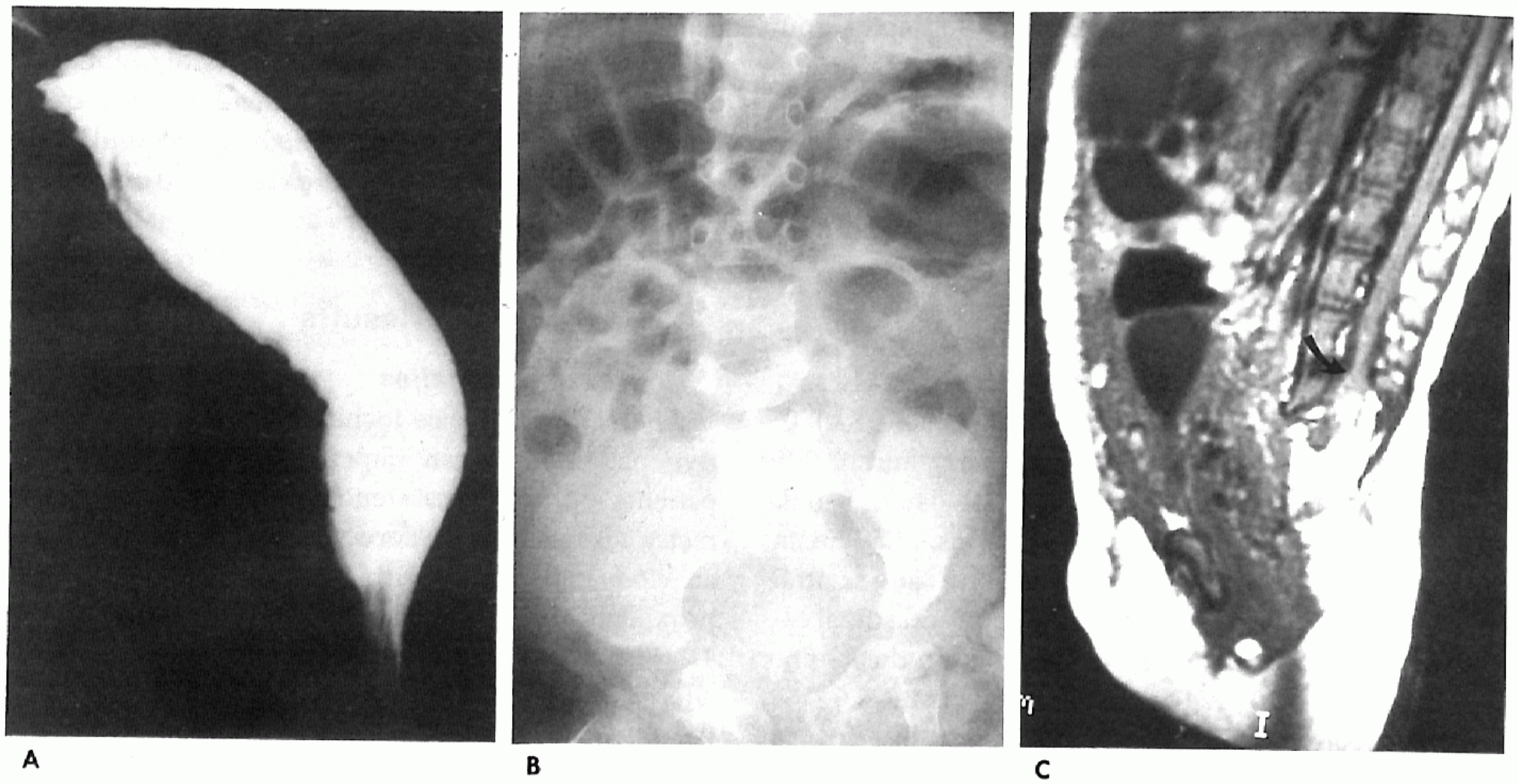

Fig. 2. An 11-month-old girl with anal stenosis, sacral defect, and lipomyeloschisis.
A. Preoperative barium enema shows reversed recto-sigmoid index. Under the initial impression of congenital megacolon, pull through procedure was done.
B. Typical scimitar shaped sacral defect and surgical staples are seen on plain radiograph.
C. Sagittal T1 weighted MR image shows the spinal cord (arrow) attached to the lipoma extending both extra- and intraspinal canal through the sacral defect.
D. Axial T1 weighted image shows pulled through intestine located within the puborectalis sling (arrows).

seemed to be more helpful in diagnosing anorectal stenosis than barium enema examination (Fig. 1). The MR scan obtained after a Duhamel abdominoperineal pullthrough procedure demonstrated the development of the puborectalis sling and the location of the pulled through intestine in relation to this (Fig. 2).

In the two patients with an imperforate anus, the location of the blind rectal pouch was below the pubococcygeal line, down to the level of the ischial rami, and this was evident on both MR images and water-soluble transcolostomy enema (Fig. 3). In addition, anal sphincter musculature was demonstrated on MR images in one patient, and in the other, the pubococcygeal sling was not separated from the teratoma, which was firmly attached to the rectum. A rectovestibular fistula was demonstrated on transcolostomy enema in this patient, but could not be demonstrated by MR imaging (Fig. 3).

Sacral anomalies

In five patients, plain radiograph revealed a typical scimitar-shaped sacral defect (Fig. 2B) and in another, the sacrum and coccyx were abnormally curved ventrally to make a wider presacral space (Fig. 1C). This latter patient was the sister of the Currarino patient with typical sacral defect. A poorly-developed sacrum with an irregular segmentation (Fig. 3A) was seen in the remaining one patient. Although MR images also showed abnormal curvature and sacral defect, the exact shape of the sacrum was easily assessed by plain radiograph.

Presacral masses and spinal anomalies

In four patients, surgical findings and pathological examination demonstrated the existence of teratoma, which in all except one case, was easily diagnosed on

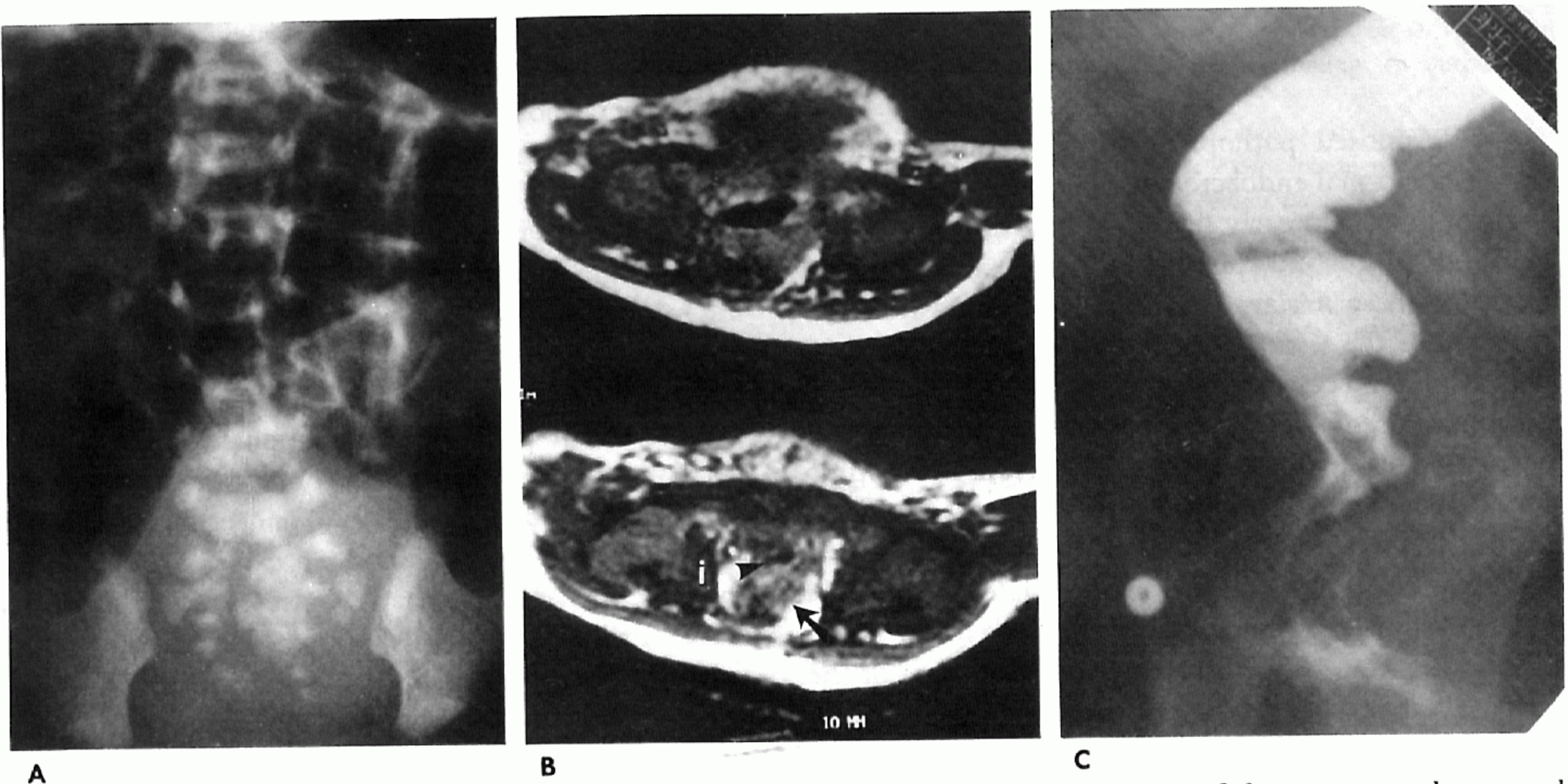

Fig. 3. A 9-month-old girl with imperforated anus, rectovestibular fistula, malsegmentation of the sacrum, and presacral teratoma.
A. Plain radiograph reveals multiple abnormal segmentation in the sacrum.
B. Fluid filled distal rectal pouch (arrowhead) is traced to the level of the ischial rami (i) on T1 weighted serial axial MR images. Presacral teratoma (arrow) is attached to posterior wall of the distal rectal pouch and the sphincter musculature can not be delineated from the mass.
C. Water soluble trans-colostomy enema demonstrates leakage of the contrast agent through the vestibule suggesting rectovestibular fistula. A marking is seen which was attached on the anal dimple.

MR images. Because in this one case, the mass was encircling the posterior rectal wall and on MR images, delineation of the mass from the rectum was difficult and perirectal teratoma was found postoperatively (Fig. 3B). Preoperatively, sonography could not also detect the mass. In the remaining three patients, MR clearly defined the size, location, and extent of presacral masses in relation to the rectum, sacrum, and coccyx (Figs. 1A, B), and moreover, demonstrated the tissue characteristics of the mass. CT scanning also visualized a presacral teratoma in another patient, but for the evaluation of anatomical detail, MRI was better.

The other three presacral masses were an anterior meningocele with lipoma, a lipomyeloschisis (Fig. 2), and a lipoma with thick filum. Correct diagnosis was made on the basis of MR images and closely correlated with surgical findings. Anterior meningocele was seen as a dural sac ventrally through the sacral defect, and a neural element within the dural sac. The lipoma was seen as a bright signal intensity on T1 weighted images and located outside the dural sac in the presacral space. A small round lesion with prologation of T1 and T2 relaxation time was seen in one case with the presacral lipoma and it was proved to be an epidermoid cyst. Postoperative MR images visualized an abscess cavity in the presacral space and barium enema revealed a fistula between the rectum and the cavity. In one patient, the spinal cord was attached to the presacral lipoma, and this extended to the intradural space without bulging of the dural sac and neural plaque. On the basis of these MR findings, lipomyeloschisis was diagnosed. On sonography, the diagnosis was the same; lipomyeloschisis was seen as lower lying cord attached to an echogenic mass. Real time observation revealed loss of normal motion of the cord, which was recorded on M-mode ultrasound scan, and on the basis of this finding, we assumed the cord was tethered. In the same patient, CT scanning revealed presacral lipoma extending into the spinal canal through a sacral defect. The last presacral mass was also a lipoma and a thick filum (〉 2mm) was seen to be tethered to the lipoma on MR images. CT scanning also demonstrated the sacral defect and presacral lipoma. MR imaging showed a dural sac, anomalous shape and level of the cord, the intra- and extra- dural extent of the presacral lipoma, and the thick filum by multiplanar images. MR also demonstrated associated anomalies including syringohydromyelia in two patients and unilateral multicystic dysplastic kidney in one.

Discussion

The proposed pathophysiology of the Currarino triad is abnormal endoectodermal adhesions and notochordal defects in early fetal life, resulting in a fistula between the gut and spinal canal. These abnormalities appear to be a variant of split notochord. Partial resorption of the fistula would give rise to an anterior sacral meningocele or retrorectal enteric cyst. The combination of these enteric and neuroectodermal elements with mesodermal elements from developing somites could explain the formation of a presacral teratoma (3).

The sacral bony defect varies from lateral deviation of the coccyx to unilateral absence of the lower sacral segments (4). In two of our patients, the shape of the sacrum was atypical ; it did not had the typical scimitar shape, with abnormal curvature or segmentation anomaly. Because the Currarino triad is a hereditary disorder, other family members should be evaluated promptly, but the complete constellation of defects is present in only a minority of cases (3). Among the triad of anomalies, one or two basic features of the syndrome may be lacking in members of the same family, suggesting an incomplete form of the syndrome (1, 7).

To evaluate the anorectal anomalies, conventional imaging techniques such as invertography, barium study, cystourethrography, sonography, and CT scans have been widely used preoperatively. Our results indicate, however, that for the evaluation of anorectal anomalies, MR imaging is very useful ; the MR findings of anorectal stenosis appeared to be a thick and elongated anorectal canal and ectatic proximal rectum (Figs. 1A, B). On barium study, however, because only the lumen of the rectum is seen, anorectal stenosis can mimic congenital megacolon involving a short segment (Fig. 1C, 2A). Although inspection or finger examination of the anus can diagnose anorectal stricture, misdiagnosis is possible. In one of our patient, in fact, the Duhamel abdominoperineal pull-through procedure was performed under the initial impression of megacolon. Fortunately, this procedure did not seem to result in a poor outcome since it has been reported that anorectal strictures are uniformly resistant to dilatation, requiring the abdominoperineal pull-through procedure for management (8).

It is known that in cases of imperforate anus, initial MR imaging is helpful (6). By delineating the distal rectal pouch and the sphincteric muscles, it demonstrates the nature of atresia correctly. Furthermore, for determining the level of the rectal pouch, MR imaging during sedation may be more accurate than conventional radiographic techniques, in which the position of the rectal pouch may be erroneously estimated because of crying or straining by the patient (5, 6). Preoperative information regarding the anal sphincter is essential for guiding the pull-through procedure and for functional prognosis. In addition, by demonstrating hypoplastic sphincteric muscles and inappropriate placement of neorectum, MR imaging also provides information regarding the possibility of repeating the procedure, in postoperative patients suffering from persistent incontinence. The only drawback of MR imaging in the demonstration of anorectal anomalies is delineation of the fistula, which can be improved by the injection of oily contrast agent (5) or insertion of a catheter filled with oily contrast media through the fistula. Our study was, however, a retrospective analysis of cases from multiple hospitals, and so we were unable to use this method.

In our cases, presacral mass was mainly teratoma or lipoma associated with various anterior spinal dysraphia. After detecting such a mass, preoperative information concerning its nature and extent, and anatomical detail concerning the relations of the cord, dural sac, and lipoma is essential ; because of its excellent soft tissue contrast and multiplanar images, MR imaging demonstrates this information more clearly than other imaging modalities and without radiation or invasive procedures. Presacral masses may be firmly adherent to the rectum or dura making surgical removal difficult. Postoperative abscess or meningitis are not uncommon and may be due to injury to adjacent structures during surgery, and preoperative infection due to a preexisting rectal fistula or infection of an enteric cyst can also occur (1). By contrast enhancement, MR can also demonstrate the presence and extent of such complications.

Congenital anorectal malformation is frequently associated with other anomalies with a reported incidence of 28—72% (5). An additional advantage of MR imaging is the ability to detect clinically unsuspected anomalies including spinal cord and renal dysplasias, which are potentially correctable or influence prognosis.

In conclusion, MR imaging clearly delineated various anorectal anomalies, presacral masses, associated spinal and other anomalies, and postoperative complications. When the Currarino triad is suspected, MR imaging should therefore follow the diagnosis of abnormal sacrum using plain radiograph, and a trans-colostomy enema will help to demonstrate the fistula.

J Korean Radiol Soc 1997;37:1127-1133

References

1. Currarino G, Coln D, Votteler T. Triad of anorectal, sacral, and presacral anomalies. *AJR* 1981;137:395-398
2. Kirks DR, Merten DF, Fisto HC, Oaks WJ. The Currarino Triad: complex of anorectal malformation, sacral bony abnormality and presacral mass. *Pediatr Radiol* 1984;14:220-225
3. O'Riodain DS, O'Connell PR, Kirwan WO. Hereditary sacral agenesis with presacral mass and anorectal stenosis: the Currarino triad. *Br J Surg* 1991;78:536-538
4. Pfluger T, Czekalla R, Koletzko S, Munsterer O, Willemsen UF, Hahn K. MRI and radiographic findings in Currarino triad. *Pediatr Radiol* 1996;26:524-527
5. Taccone A, Martucciello G, Dodero P, Delliacqua A, Marzoli A, Salomone G, Jasonni V. New concepts in preoperative imaging of anorectal malformation. *Pediatr Radiol* 1992;22:196-199
6. Sato Y, Pringle KC, Bergman RA, et al. Congenital anorectal anomalies: MR imaging. *Radiology* 1988;168:157-162
7. Norum J. Incomplete Currarino syndrome with a presacral leiomyosarcoma. *Acta Oncol* 1990;30(8):987-988
8. Moazam F, Talbert JL. Congenital anorectal malformations: harbingers of sacrococcygeal teratomas. *Arch Surg* 1985;120:856-859

대한방사선의학회지 1997;37:1127-1133

Currarino Triad의 자기공명영상 소견[1]

[1]중앙 길병원 진단방사선과
[2]서울대학교 의과대학 방사선과
[3]계명대학교 의과대학 동산병원 방사선과
[4]중앙 길병원 일반외과

김지혜 · 김지은 · 김인원[2] · 이희정[3] · 이영석 · 이태훈[4] · 김형식

목 적 : Currarino triad의 다양한 자기 공명 영상 소견을 알아보고 여러 가지 기형을 진단하는데 자기 공명 영상의 역할을 논하고자 하였다.

대상 및 방법 : 7예의 Currarino triad 환아 (연령 분포 : 2-12개월) 에서 MRI, 단순 촬영, 대장 검사를 시행하였고 일부에서 CT (3예)와 초음파 (2예)를 시행하였다. 저자들은 MRI 소견을 다른 영상 소견과 비교하여 후향적으로 분석하였다.

결 과 : 항문 직장 기형은 항문 직장 협착 5예, 항문 직장 폐쇄 2예가 있었다. 항문 직장 협착은 MRI에서 길고 두꺼워진 항문 직장과 그 근위부 확장으로 진단할 수 있었다. 항문 직장 폐쇄에서는 원위부 직장의 위치와 괄약근을 MRI로 평가할 수 있었다. 인공 항문을 통해 시행한 대장 검사에서 확인된 직장 누공 (1예)은 MRI로 발견할 수 없었다. 천추 앞 종괴는 기형종 4예, 다양한 척추 기형을 동반한 지방종이 3예 있었다. MRI로 천추 앞 종괴와 동반된 다른 기형을 자세히 평가할 수 있었으며 함께 시행한 CT나 초음파 검사보다 우월하였다. 천추 기형은 전형적인 검 모양의 천추가 5예, 비정상적인 굴곡과 분절을 보인 예가 각각 1예씩 있었다. MRI로도 비정상적인 천추를 모든 예에서 확인할 수 있었으나 천추 기형의 정확한 모양은 단순 촬영으로 보다 쉽게 진단할 수 있었다.

결 론 : Currarino triad 의 다양한 항문 직장 기형, 천추 전방의 종괴, 그리고 동반된 다른 기형을 MRI로 진단할 수 있었으며 단순 촬영 후 Currarino triad가 의심될 때 다음 검사로 MRI가 시행되어야 할 것으로 생각된다.