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ORIGINAL ARTICLE

C. Festen · R. S. V. M. Severijnen
F. H. J. van der Staak · P. N. M. A. Rieu

Rectal atresia: pathogenesis and operative treatment

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Abstract Rectal atresia (RA) with a normal anus is a rare anomaly mostly described as part of a series of anorectal malformations. Most authors believe it to be an acquired lesion with a vascular genesis. One of the arguments quoted is the lack of other congenital anomalies. Several operative procedures are recommended for this lesion. We describe four patients with RA who had other significant congenital anomalies; two other cases were found in the literature. A lack of other congenital anomalies in patients with RA does not seem to be a strong argument for an acquired lesion. All four patients were treated by a posterior sagittal approach with good functional results.

Key words Rectal atresia · Anorectal malformation · Posterior sagittal anorectoplasty

Introduction

Rectal atresia (RA) with a normal anus is a rare anomaly, mostly described as part of a series of anorectal malformations (ARM) [9, 12, 13, 15, 17–19, 22]. Most authors believe it to be an acquired lesion rather than a developmental error. Besides experimental work that supports a vascular genesis, one argument used is the lack of associated congenital anomalies, in contrast to other ARM. Several operative procedures are described in the literature with varying results, most of them originally designed for Hirschsprung's disease or anorectal atresia. From January 1970 through December 1995, in a series of 311 patients with ARM treated in the Pediatric Surgical Center, Nijmegen, we observed four patients with RA.

C. Festen (✉)
Department of Pediatric Surgery, P. O. Box 9101,
NL-6500 HB Nijmegen, The Netherlands

Case reports

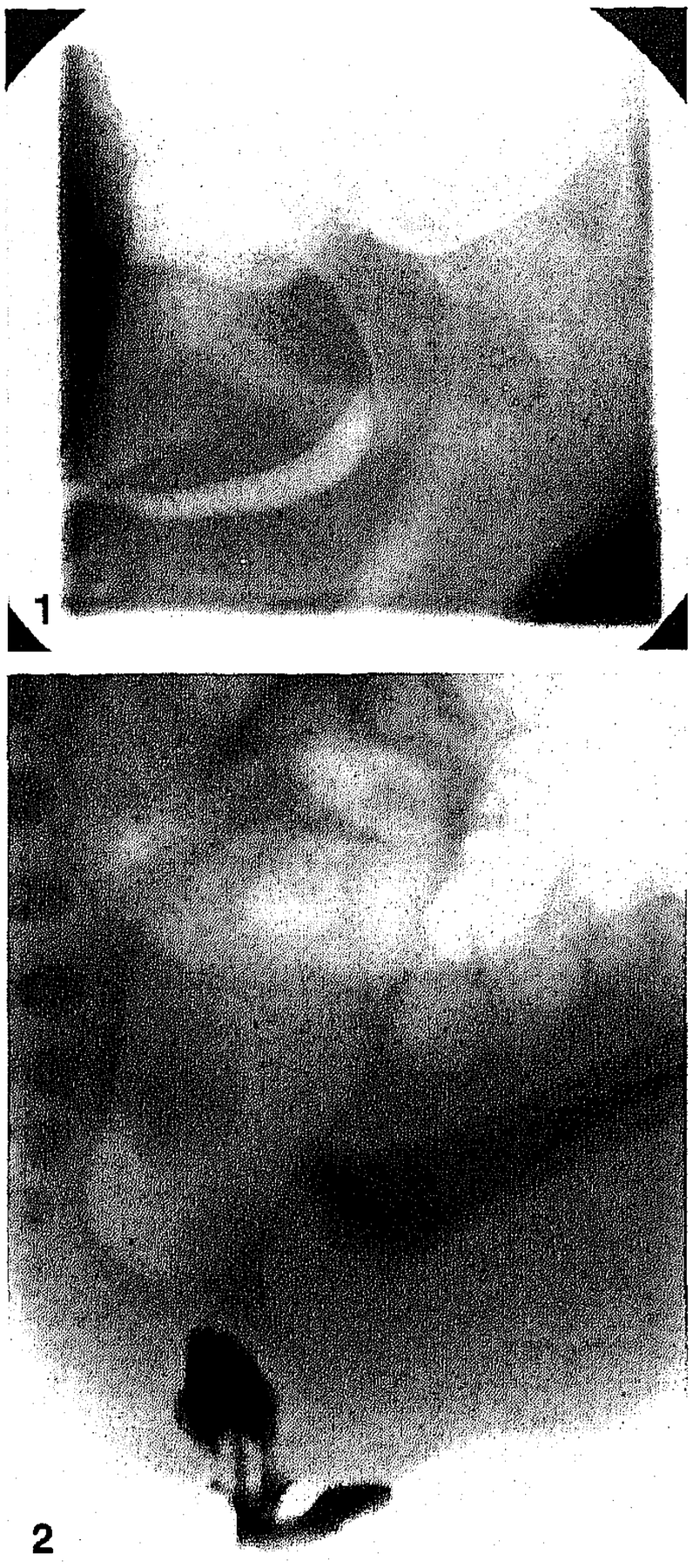
Case 1
A 3-day-old male was referred because of abdominal distension and failure to pass meconium. He was born at full term after an uncomplicated pregnancy with a birth weight of 2,540 g. On examination, there was massive abdominal distension with a normal perineum, genitalia, and anus. A tube would not pass the rectum, and during rectography RA was diagnosed. During this investigation there was massive extravasation of barium, preventing adequate X-ray pictures. At operation the same day, after laparotomy and rectotomy, a diaphragm remained between the anal canal and the rectum that could be perforated from below, guided by an index finger in the rectum. The opening in the diaphragm was dilated with bougies and an "endless thread", as described by Rehbein [16], was guided through the anus, the opening in the diaphragm, and a low colostomy on the sigmoid colon. With the aid of this thread, the opening in the diaphragm could be further dilated.

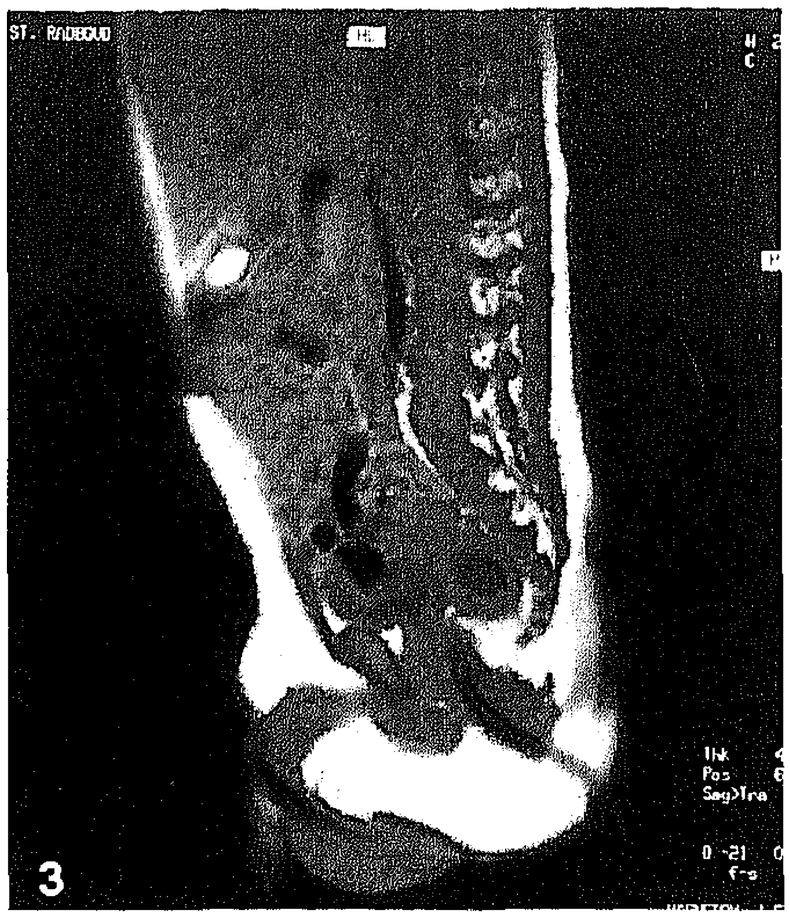
After the operation, feces emerging from the penis indicated a rectourethral fistula that could be demonstrated by an urethrogram (Fig. 1) and urethroscopy. Further examinations showed a hemivertebra at T_{12} but no other abnormalities. After good passage through the rectum was established, at the age of 6 months, we tried to close the rectourethral fistula by a transsphincteric approach, but this failed. Because the boy developed marked scoliosis that required a bandage and Boston brace, the colostomy was closed temporarily and a second attempt to close the fistula postponed. During that time no feces were passing through the penis, but urine was passed via the anus.

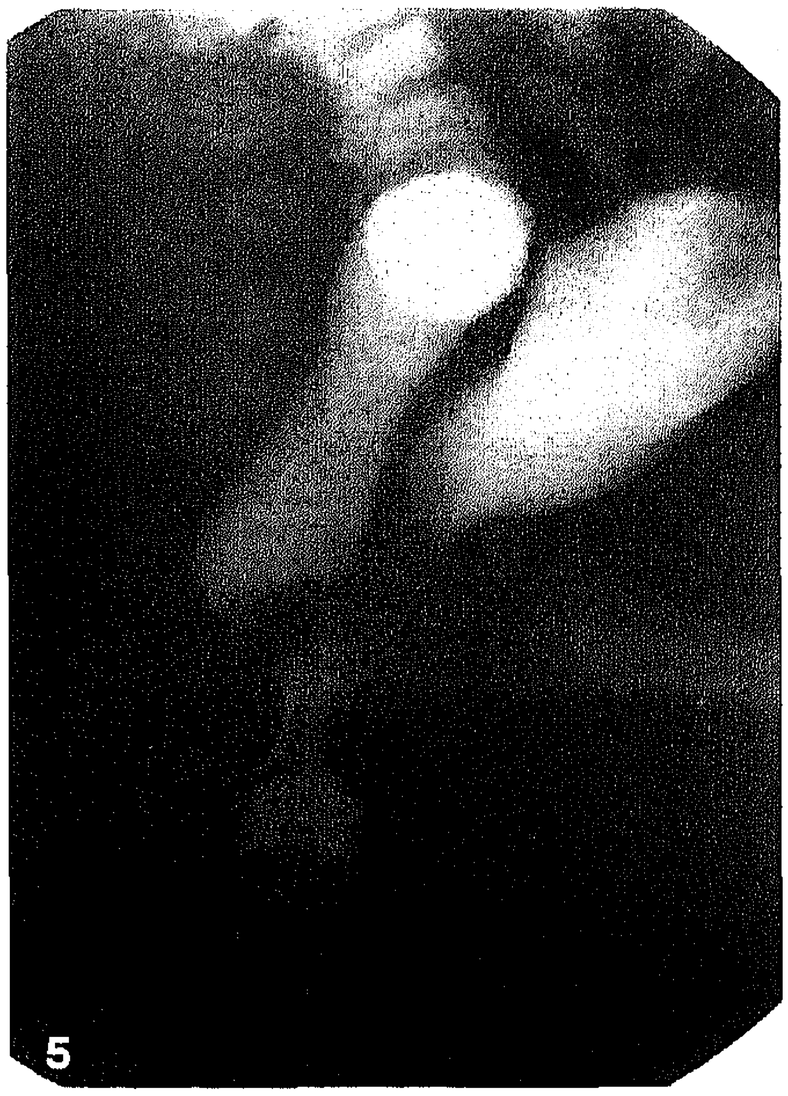
At the age of 10 years the urine loss through the anus and borderline fecal continence became socially unacceptable, so that another attempt was planned to close the fistula. After a new colostomy, a posterior sagittal approach (PSAPR) was used. The fistula was closed and the rectum reconstructed; 3 months later the colostomy was closed. The fistula is presently closed; the boy has normal daily stools and only occasional soiling.

Case 2 A male baby born after an uncomplicated pregnancy with a birth weight of 2,055 g was admitted 2 days later because of failure to pass meconium and abdominal distension. There was a normal-looking anus, but on rectography (Fig. 2) and MRI (Fig. 3) RA was diagnosed. A colostomy was performed the next day. Further investigations disclosed a patent ductus Botalli (PDB), which after 3 weeks had been closed operatively. At the age of 6 months we performed a PSARP and found a rectal diaphragm that could be excised. One month later the colostomy was closed. Since that time he has had normal defecation without soiling.

Case 3 A male baby was born spontaneously after an uncomplicated pregnancy with a birth weight of 2,960 g. He was admitted the same







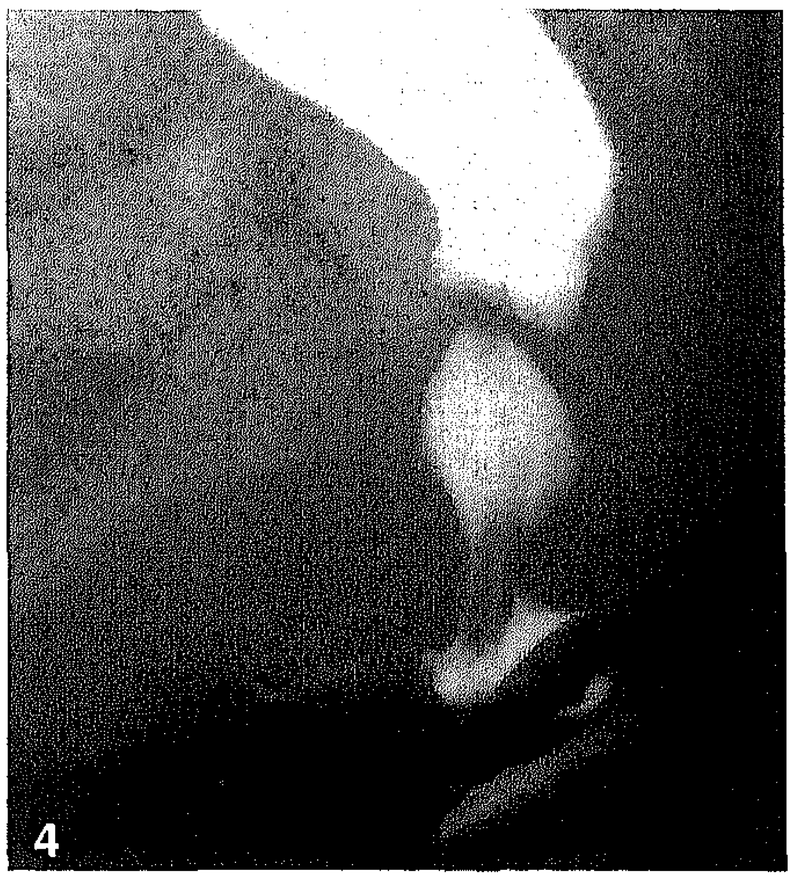


Fig. 1 Urethrogram with overflow of contrast into rectum. Contrast in pelvis is the result of previous extravasation

Fig. 2 Rectography combined with invertogram

Fig. 3 MRI scan with tube in rectum showing anatomic configuration

Fig. 4 Distal loopogram with contrast in rectum showing diaphragm

Fig. 5 Distal loopogram and rectography showing diaphragm

day with a low intestinal obstruction and a normal-looking anus. There was also glanular hypospadias and a small ventricular septal defect (VSD). Rectography (Fig. 4) demonstrated RA and a colostomy was performed the next day. At the age of 5 months a rectal diaphragm has corrected by PSARP; 2 months later the colostomy was closed. He is now completely continent.

Case 4 A female baby was born at full term with a birth weight of 3,500 g. Shortly after birth she started vomiting and did not pass meconium. She was referred the next day, and clinical examination revealed abdominal distension with normal-looking genitalia and anus. Rectography (Fig. 5) showed RA. There appeared to be no further abnormalities. The next day a colostomy was performed. At the age of 3 months a rectal diaphragm was corrected through by PSARP at which time a vaginal atresia with a normal-looking vulva was diagnosed. During the sagittal exploration, no vagina was identified. Histologic examination of the diaphragm showed smooth-muscle structures. The karyotype was 46,XX. Three months later the colostomy was closed and the presence of normal internal genitalia was confirmed. Postoperatively there was normal defecation; vaginal reconstruction will be done in the future.

Discussion

The incidence of RA in most series of ARM is 1% or 2% [13, 15, 17, 20], with the exception of certain districts in India [2], where it is as high as 14% without obvious reasons. We found an incidence of 1.3%. RA is more common in males and has been classified into four types [2]. 1: RA with a short gap; 2: RA with a long gap; 3: septal type; and 4: rectal stenosis. Our patients had a male/female ratio of 3:1 and all had type 3 RA. The clinical picture is mostly one of low intestinal obstruction, and because of the normal anus, there may be a delay in diagnosis.

Most authors believe RA to be an acquired lesion. The etiology is more likely to be a vascular accident than a developmental anomaly. In 1961 Partridge and Gough [13] speculated that RA may be the result of an obstruction of the arterial blood supply through the superior rectal branch of the inferior mesenteric artery. Freeman [3] suggested classifying RA as a colonic rather than a rectal atresia. Magnus [11] dissected a female child who died shortly after birth and found strong histologic evidence of a vascular cause. Stone and Wilkinson [21] demonstrated the experimental production of rectal stenosis and atresia in rabbits by partial or complete interruption of the blood supply to the rectosigmoid. Investigations by Lambrecht et al. [10] in pigs also support this hypothesis. Dorairajan and Durham Smith found an absence of the middle rectal vessels [2].

As a strong argument for an acquired genesis of RA, many authors quote the lack of other congenital anomalies in contrast to other ARM. In most small series of RA described there were no other anomalies [2–8].

In our series one of the patients had a rectourethral fistula, and of the four described by Peña and DeVries [15] there was one with a rectovaginal fistula and one with a double rectourethral fistula. These fistulae cannot be explained by the vascular theory, and better match the usual ARM. The other anomalies were scoliosis, PDB, glanular hypospadias combined with a VSD, and vaginal atresia. In all four of our patients, therefore, other significant anomalies were found, which indicates that associated anomalies may not be as uncommon in patients with RA as previously suggested, and thus this is not a strong argument for an acquired lesion.

Several operative procedures have been described for RA, and are collected in the monograph of Stephens and Smith [20]. Techniques designed for Hirschsprung's disease (Swenson [18, 19], Duhamel [6]), for high anal atresia (abdominal [9], abdominoperineal [22], abdominosacral pull-through [12], PSARP [5, 14], and transanal approaches are used [1, 4, 16, 23–25]. The results of these techniques are not well-described.

In our experience, a PSARP as designed by Peña and De Vries [15] for anorectal atresia seems to be the most rational, and an anastomosis can usually be established without cutting the external sphincter complex. The functional results are excellent [7], as was also our experience.

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