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

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 dia-

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 showed 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
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].