Original Study

Treatment of Adults with Unrecognized or Inadequately Repaired Anorectal Malformations: 17 Cases of Rectovestibular and Rectoperineal Fistulas

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A B S T R A C T

Study Objective: To analyze all cases of congenital rectovestibular and rectoperineal fistulas diagnosed and treated later in life, and to describe presenting complaints, treatment, and outcome.
Design: Retrospective cohort study.
Setting: Pediatric surgery departments of 3 major referral centers in the US and Europe.
Participants: Seventeen women with untreated or inadequately treated rectovestibular or rectoperineal fistulas.
Interventions: Analyses of all eligible patients: charts were analyzed for the classification of the malformation, main complaints, continence, sexual function, indications for surgery, associated anomalies, surgical procedure, complications, and outcome.
Main Outcome Measures: Patients' complaints, continence, constipation, and sexual function.
Results: Major complaints at time of diagnosis were fecal incontinence, and concerns for hygiene and cosmesis. All patients were repaired by a posterior sagittal approach. In all but 1 patient the complaints disappeared or improved after surgery.
Conclusions: Anorectal malformations in females are congenital malformations mostly seen and treated in early childhood. If unrepaired or inadequately repaired the patient, when reaching adulthood, can suffer from significant morbidity. Surgical treatment is similar as in childhood and has an excellent clinical outcome.
Key Words: Rectovestibular fistula, Rectoperineal fistula, Delayed diagnosis, Imperforate anus, Anorectal malformation

Introduction

Anorectal malformations (ARM) in females comprise a spectrum of malformations ranging from rectoperineal and rectovestibular fistulas to complex malformations such as cloacas.[1] The incidence is 1 in 3500-5000 births.[1] The most severe and complex cases are often recognized early after birth. Less complex malformations, such as rectovestibular and rectoperineal fistulas, may not be diagnosed in a newborn as the child can successfully stool through the fistula. They may then be recognized in early infancy, usually with the presentation of severe constipation.[2] Some are repaired inadequately leaving the patient with an anteriorly mislocated anus and an inadequate perineal body.[3] Seldom, rectovestibular and rectoperineal fistulas remain undiagnosed until the patient is in early adulthood. If not diagnosed early or treated properly, patients may present with complaints such as severe constipation, fecal incontinence, overflow of stool into the introitus or into the vagina and sexual dysfunction. These late presentations have only been reported in case studies.[4,5] Late surgery makes the surgical repair more difficult and can influence the outcome.[6] Furthermore, experience with these congenital anomalies for adult specialists, especially gynecologists, may be very limited, which makes recognition of these malformations difficult and treatment options often unknown. Therefore, our aim was to describe our cases with anorectal malformations such as rectovestibular and rectoperineal fistulas treated in adulthood. The surgical procedure to repair the congenital fistula and associated anomalies and outcome are described.

Methods

By querying our database of 2500 patients with anorectal malformations, 17 female patients above the age of 15 years were identified with rectovestibular or rectoperineal fistulas (Fig. 1, *A*, *B*) that were unrepaired or inadequately reconstructed. Charts were analyzed for the classification of the malformation, main complaints, continence, sexual function, indications for surgery, associated anomalies, surgical procedure, complications, and outcome.

As an adolescent or young adult (Fig. 2), patients underwent a posterior sagittal anorectoplasty (PSARP) primarily to repair the malformation or to redo the previous

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The protocol of the study was approved by the ethics committee of the Erasmus Medical Centre on March 27, 2012 (EC# MEC-2012-171). The study was exempted from individual informed consent, because of the study's retrospective and anonymous design.

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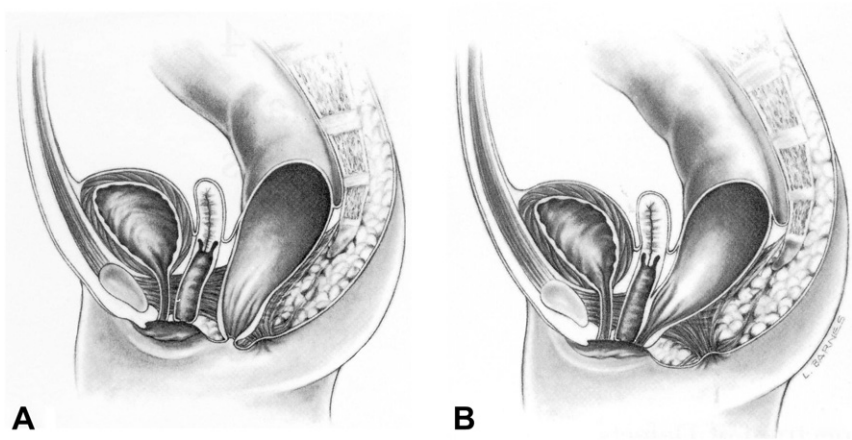

Fig. 1. (A) Sagittal view of a rectoperineal fistula. (B) Sagittal view of a rectovestibular fistula. *With kind permission from Springer Science+Business Media: Pena A, Atlas of Surgical Management of Anorectal Malformations, Springer Verlag, New York, 1989, pp 50-55.*

inadequate repair. The indications for surgery were to restore normal anatomy, with creation of an adequately sized and located anal orifice, as well as the development of an adequate perineal body with the hope to allow for normal function. After opening of the skin and subcutaneous tissue the sphincter complex is incised in the posterior midline, and nerves are thus spared (Fig. 3, *A-F*). The rectal fistula and vagina in all cases share a common wall that needs to be separated into 2 parts, with no discrete anatomical plane between the 2 structures. Once separated, the rectum is placed within the sphincter complex. The perineal body is built from the fourchette to the anterior boundary of the muscle complex. Then the posterior part of the muscle is approximated to the posterior rectal wall. After restoration of the muscle complex, an anoplasty is performed. Postoperatively, patients are kept with nil per os for 7 to 10 days and given intravenous nutrition. At least 14 days after surgery daily anal dilatations with Hegar dilators are started and continue for approximately 3 to 6 months.

Results

Patient Description

One patient had a rectoperineal fistula; sixteen patients had a rectovestibular fistula. The median age at time of surgery was 25 years (range 15-55 years, Table 1). Twelve patients (72%) were initially evaluated by a gynecologist who then referred the patient to one of our centers. Thirteen patients (76%) had previously undergone repair of the anorectal malformation but required a redo procedure to establish normal perineal anatomy. Of these 13 patients, 6 were primarily operated as newborns with a cutback or anoplasty only; the other 7 patients had had a previous PSARP (Table 1). The key technical deficiency in all of these cases was the inadequate surgical separation of rectum from posterior vagina.[3] In 5 patients other genital malformations were found (Table 1). In 2 patients there was associated vaginal agenesis and a replacement with sigmoid was performed. Two other patients had duplication of the Müllerian system, the most common genital tract anomaly found in ARM patients.[7,8]

Indications, Surgery, and Outcome

All patients underwent a posterior sagittal approach to separate the congenital fistula from vagina or perineum. There were no major complications after surgery. One patient had a wound dehiscence and needed an early resuturing of the anoplasty.

The major complaints that patients presented with at time of diagnosis were fecal incontinence (41%), concern for personal hygiene and cosmesis (24%), rectal prolapse (24%),

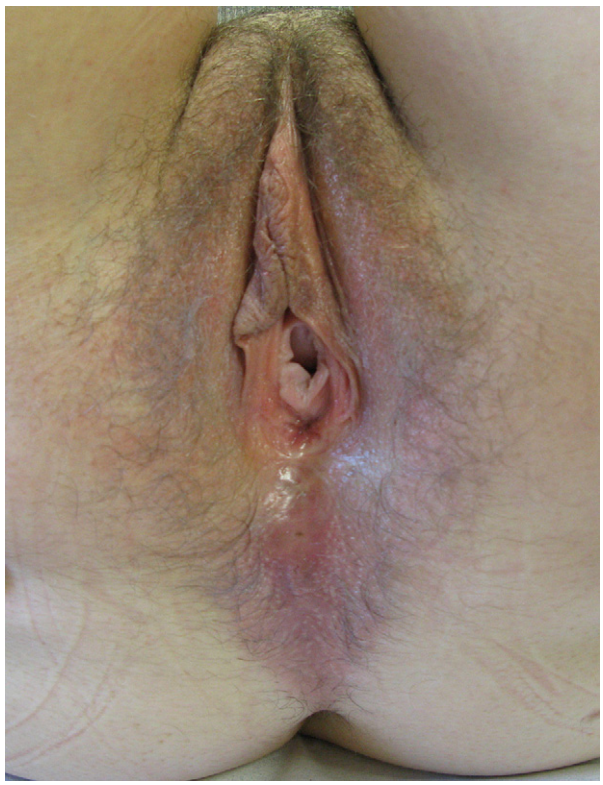

Fig. 2. Photograph of an adult patient with a rectovestibular fistula at the fourchette "treated" as an infant. She was fully continent until her first sexual contact after which daily soiling started. After a posterior sagittal repair she became fully continent again with normal sexual activity.