Congenital Anal Fistula With Normal Anus

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● **Three patients with a congenital anovestibular/perineal fistula were treated at the Montreal Children's Hospital. Two females (one of East Indian and the other of Japanese origin) had anovestibular fistulae that became symptomatic in the first few months of life. The third patient, a boy of Korean descent, presented at 9 months of age with a chronically draining perineal fistula. During surgery, a small fistula tract was easily dissected out and excised. Microscopic examination showed a well-preserved rectal mucosa throughout the tract. Most male patients described to date had anourethral fistulae, often accompanied by other major anomalies such as esophageal atresia or renal malformations. We believe our patient is the first male to be described with a congenital perineal fistula; this suggests that some fistula-in-ano in male infants may be due to a congenital sinus that secondarily becomes infected and drains to the skin.**

INDEX WORDS: Anorectal malformations; congenital anal/rectal fistula.

ANAL AND RECTAL AGENESIS are generally accompanied by perineal, vaginal, or urethral fistulae. However, such fistulae are rare in the presence of a normal anus. In recent publications, they have been termed H-type urethroanal[1] or anourethral fistula,[2] N-type anorectal malformations,[3] or rectourethral fistula[4] for males. For females, the terms "rectovestibular fistula"[5,6] and "double termination of the alimentary tract"[7,8] are commonly used.

We describe one male with an anoperineal fistula and two females with anovestibular fistulae. None of the patients had any other abnormality of the anorectum.

CASE REPORTS

Patient 1

A 2-month-old East Indian girl presented when her parents noted the presence of stool at the vulva. There was no history of abscess or inflammation. She was admitted at 6 months of age for surgery. The anus was in normal position and had good sphincter tone. A 1.5-mm opening just inside the posterior fourchette, distal to the hymen, was seen. Vaginal swabs grew *Klebsiella, Escherichia coli,* and *Clostridium perfringens.* A barium enema documented the fistula. An abdominal ultrasound demonstrated normal kidneys.

Under anesthesia, a lacrimal probe was inserted from the anal side into the fistulous opening, 0.5 cm above the dentate line (Fig 1). The probe was used as a stent in the tract, enabling it to be cored out. The tract was excised using the technique described by Tsuchida et al.[8] This consisted of an elliptical excision of the anterior wall of the anal canal around the fistula opening, followed by dissection of the tract from both the anal and the vulvar sides, closure of the vulvar wound, and pull-through of the anterior wall of the anal canal (and rectum). She had an uneventful postoperative course. Histology of the fistulous tract showed stratified squamous and columnar epithelium in continuity. There was no inflammation. Two and one-half years postoperatively, there has been no recurrence of the fistula and the patient remains asymptomatic.

Patient 2

A Japanese girl presented at four years of age. Her parents had noted passage of feces per vaginam since the girl was 6 months of age. Presentation to our department had been delayed because of the advice of the family physician, who felt this would heal spontaneously. A 2-mm opening at the fourchette was seen. The anus was normal in position, size, and sphincter tone. A spina bifida occulta of S1 was seen on an x-ray. There were no other abnormalities.

Under anesthesia, a feeding tube was inserted into the perineal opening and passed readily into the anus. The fistula was excised and the rectum and vagina reconstructed as in the previous patient. Histology of the tract again documented colonic mucosa in continuity with vaginal mucosa (Fig 2). There was minimal inflammation. The child was readmitted 1 year postoperatively with a diagnosis of mucocutaneous lymphadenitis (Kawasaki's disease). There have been no sequelae. The patient is currently asymptomatic 3 years postoperatively, with normal bowel movements and no recurrence of the fistula.

Patient 3

A Korean boy presented at 9 months of age with a perineal fistula that intermittently drained purulent material. The duration of the symptoms was unknown because the patient was adopted. A fistula that opened 3 cm anterior to a normal anus was found. The remainder of the physical examination results were normal. During surgery, a thin well-defined fistulous tract, which was not adherent to surrounding tissue, could be easily dissected out. This fistula led from a fibrous subcutaneous nodule at the site of the perineal opening, through fibers of the external sphincter, to the anal canal. A microscopic examination showed a well-preserved rectal mucosa throughout the tract. There was no recurrence 2 years postoperatively.

DISCUSSION

Rectourethral fistulae in males with a normal anus were described as early as 1908[9] and 1915.[10] These rare cases, up to 1970, were summarized by Stephens and Smith.[11] DeVries and Friedland coined the term H-type fistula[2] in 1974, and more cases of similar malfor-

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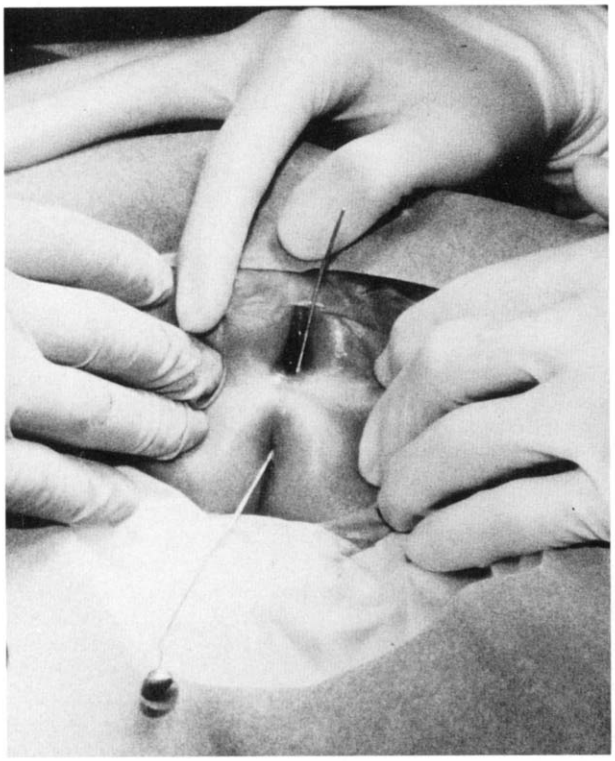

Fig 1. Intraoperative picture of patient 1 with probe in place.

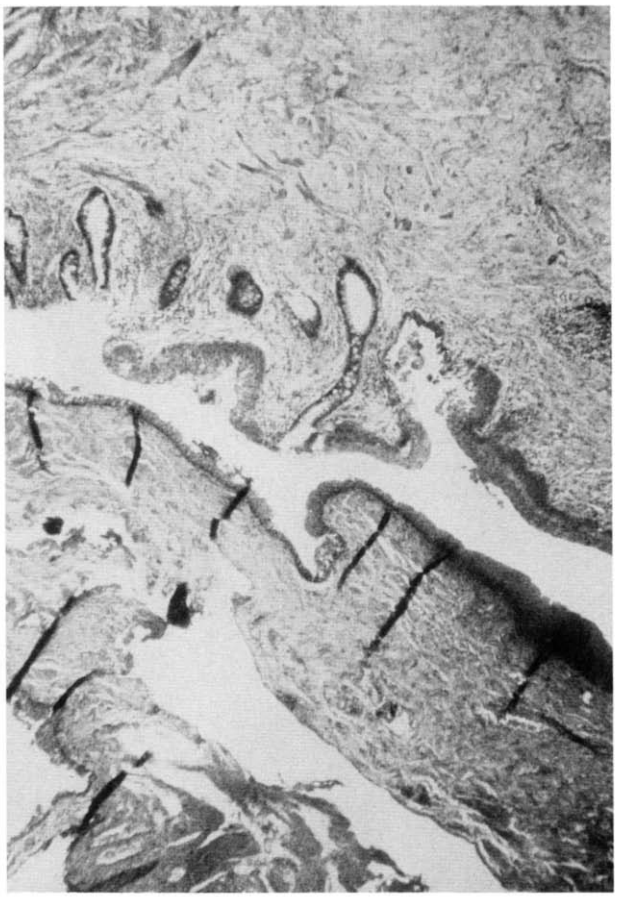

Fig 2. Microphotograph of the histology of the fistula in patient 2 (original magnification ×10). The colonic mucosa with crypts and goblet cells is visible on the left side, and becomes stratified squamous epithelium on the vaginal side.

mations were described by Pellerin et al,[4] Stephens and Donnellan,[1] and White et al.[3] Combining the more recent review of Spitz and Jung[12] with that of Stephens and Smith,[11] and adding other case reports missed by the former authors[3,4,13] or appearing since,[14] we found a total of 14 cases of ano- or rectourethral fistula (rectovesical in one case) in males with a normal anus, and seven more with mild anal anomalies such as anal stenosis, anterior perineal anus, or membranous anal atresia. This does not include other cases mentioned as "personal communications." Patients can present with passage of urine per rectum, passage of meconium in the urine, or recurrent urinary tract infections. The majority of the males reported with anourethral fistula had other severe anomalies such as renal malformations, urethral hypoplasia or atresia, and tracheoesophageal fistula with or without atresia. In our review, we did not find any male patient with a congenital anoperineal fistula, as in patient 3—an anomaly more similar to that described in females. Tagart[15] reported an 11-month-old boy with a "paraanal sinus" having some features in common with our patient, but the sinus did not communicate with the anal canal.

Bryndorf and Madsen[16] were the first to describe a female patient with a rectovaginal fistula and a normal anus. Many reports have since appeared, the largest series being from Asia—Chatterjee from India,[7] Tsuchida[8] and others[5,6] from Japan, and Ninh and Bohn[17] from Vietnam. These patients usually presented with passage of stool or flatus per vaginam in the first year of life. The fistula has occasionally been described as rectovaginal, but in most cases appears to be anovestibular; Stephens and Smith[11] call this malformation a perineal canal. Over 50 cases have been reported, none with major malformations. The only anomalies were a stenotic or anteriorly placed anus, and the presence of two or more openings in some cases. Many patients presented an abscess at some point in their course. This is thought to be secondary to infection in a congenital blind-ending sinus with secondary fistulization, or infection in an existing fistula leading to additional openings.[11]

In boys with urethral fistulae, the congenital nature of the lesion is undisputed. In patient 3 and in girls, some could argue that these are acquired perineal abscesses that fistulize and become epithelialized with time. However, the early onset of symptoms and the absence of inflammation in many cases are more indicative of a congenital origin, either sinus or fistula. It is interesting to note that >90% of infants with

fistula-in-ano are males,[18,19] and a hormonal imbalance acting on the anal glands has been proposed as the etiology.[18] Others have proposed that abnormally deep crypts of Morgagni were at the origin of the fistula-in-ano,[19] and some even challenge the classic teachings about the anal glands.[20] The fact remains that infants with fistula-in-ano usually have inflamed fibrous tracts adherent to surrounding tissues, showing granulation tissue and no epithelial lining upon microscopic examination if they are excised. Patients with congenital anovestibular or anoperineal fistula with a normal anus are all girls, with the exception of patient 3, and there is a strong racial predisposition. Our Korean boy may belong to this Asian group predisposed to low types of fistulae without severe malformations.

The embryologic basis for congenital fistulae with a normally placed anus[12] remains speculative and may not be the same in males and females. In some cases, the fistula in the male appears more like a urethral duplication emptying in the anal canal.[1] More recently, van der Putte[21] published a new theory on the development of the anorectum, which satisfactorily explains most cases of anorectal malformations, including fistula with normal anus. He suggests that the urorectal septum never joins the cloacal membrane to divide it into anal and urogenital membranes as is currently thought; anomalies of various parts of the cloacal membrane would lead to localized failure of its regression and explain most malformations of the area.

The ideal surgical technique to excise anovestibular fistulae appears to be the one described by Tsuchida et al.[8] Recurrences are rare; functional and cosmetic results are excellent. Simple excision of the tract without pull-through of the anterior wall of the rectum leads to a higher risk of recurrence.[3,7,8] In boys with urethral fistulae, the treatment is more difficult and must be preceded by thorough investigations. Some have used the urethroanal fistula to bypass a hypoplastic perineal urethra.[4] When the urethra is of adequate caliber, the fistula can be excised through an anterior perineal or a posterior sagittal approach. A preliminary colostomy is indicated, at least when the fistula opens in the rectum. When it opens in the anal canal, a pull-through of the anterior wall (as performed by Tsuchida et al[8] for girls) might yield equally good results. Another way to deal with the fistula, also useful to avoid recurrence, is to perform a Soave-type endorectal pull-through by simply ligating the fistula at the level of the muscular cuff.[12] Recurrences are frequent with any technique, and were observed even with the Soave pull-through.[4] Postoperative anal dilatations are indicated only if concomitant anal stenosis exists, and were not used in our cases.

The anomaly of a congenital anal fistula without imperforate anus is rare, representing $<1\%$ of anorectal malformations. It should be classified under the category of rare anorectal malformations in the recent "Wingspread" classification.[22] The anomaly is less severe in girls, most of whom are Asian.

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