Translevator Anal Anomalies With Cutaneous Fistulae Passing Deep to the Scrotum

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Methods: The authors present a series of 9 patients collected from 4 centers with translevator anal anomalies, each of which has a fistula tracking forward deep to the scrotum and opening at the peno-scrotal junction. Whereas some would appear to be covered ani in type, others are deeper and would appear to fit in with an intermediate type of classification emphasizing the idea of a "spectrum" of malformation.

Results: The anatomic arrangement, associated anomalies (eg, 2 had hypospadias), and surgical management is described briefly in each case. Careful examination may be necessary to identify the fistula.

Conclusions: It is recommended that the surgery be individualized depending on the findings. On a theoretical embryologic basis there is abnormality in the formation of the outer genital folds, and there also may be abnormality in some cases of the inner genital folds.

INDEX WORDS: Anal anomaly, imperforate anus, fistula deep to scrotum, translevator.

IN THE SPECTRUM of congenital anal anomalies it would not be too surprising to find a translevator "imperforate" anus with a fistula passing deep to the scrotum and opening at the peno-scrotal junction. We present a series of 9 such patients collected from 4 centers. It is surprising that few cases have been published previously in the literature. The case of Nixon, quoted by Stephens and Durham-Smith,[1] is a patient in our series. Another similar case of the fistulous tract running deep above the base of the scrotum and opening near the penoscrotal junction has been reported.[2] In general, the nomenclature of the *Wingspread Classification* has been used.[3]

MATERIALS AND METHODS

Information was collected from the 4 centers cited. In this, a largely retrospective review, the amount of information available was variable.

Details including external findings on clinical examination, site of fistula opening (Fig 1-4), associated anomalies, surgery performed, long-term results, and histology of fistulous tracts were documented. The data were evaluated and presented in a tabulated form (Table 1).

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DISCUSSION

In 2 of the patients described, the fistulae were not diagnosed at birth, and it is therefore important to look extremely carefully for an opening at the penoscrotal junction in any boy with "imperforate anus." It is stressed that there may be nothing passing through the fistula. If this is found we would recommend gentle probing, and if the fistula appears to be deep to the scrotum, then a fistulogram should be carried out. If the lesion is of covered anus type (low), then a local anoplasty is indicated, with excision of the fistula either then or at a later date. The fistula can be dissected out by "bi-valving" the scrotum, that is, dividing it along the median raphe and dissecting it from the bowel to the peno-scrotal junction. Great care is needed to avoid entering the urethra where it is closely attached. Alternatively, it can be cored out from the bowel to its destination (Fig 3). This is facilitated by the elasticity of the scrotum, which allows it to be retracted sufficiently.

The fistulogram may indicate an intermediate lesion with anal agenesis. We use the term *anal agenesis* to indicate failure of formation of the anus, as per the Wingspread Classification, but with the only difference being that this anomaly has a fistula running forward deep to the scrotum. In this anomaly, we would recommend a colostomy initially. Subsequent to this, excision of the fistula and a posterior sagittal ano-rectoplasty (PSARP) can be performed.[4] Alternatively, a posterior approach similar to that of Peña but without dividing the muscle complex as recommended by Durham Smith (personal communication) would be possible. A totally perineal approach also is possible as long as great care is

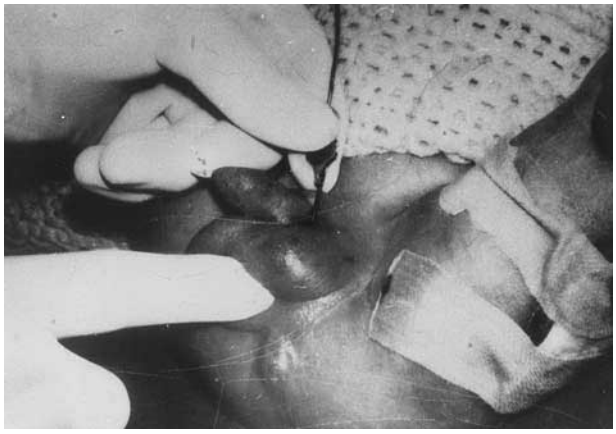

Fig 1. Probe in fistula site.

taken to ensure that the bowel is brought down in front of the puborectalis muscle. We would consider this a more difficult and dangerous approach.

It is tempting to engage in some theoretical embryology relating to these anomalies. It would seem that they fit into the spectrum of anorectal malformations between rectobulbar urethral fistula and anocutaneous fistula in the Wingspread classification.[3]

From the descriptions of the 9 cases given, there appears to be a spectrum between an intermediate type with anal agenesis to a type with a truly covered anus. According to Stephens and Smith,[5] after the partition of

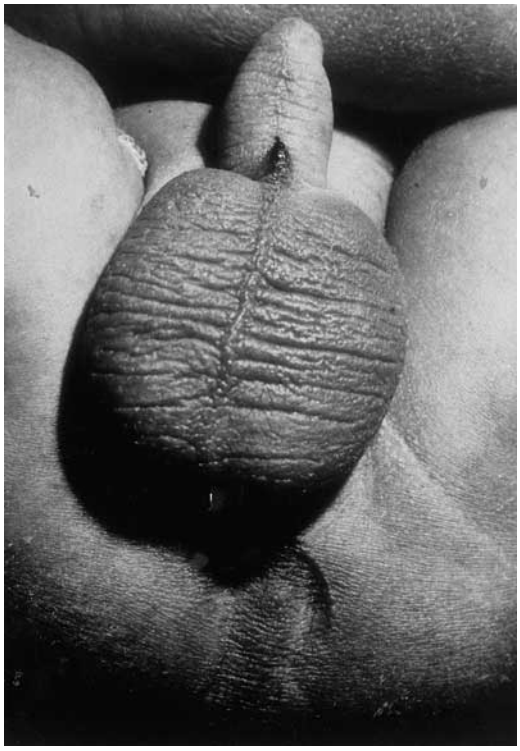

Fig 2. Meconium visible in bulla at fistula opening.

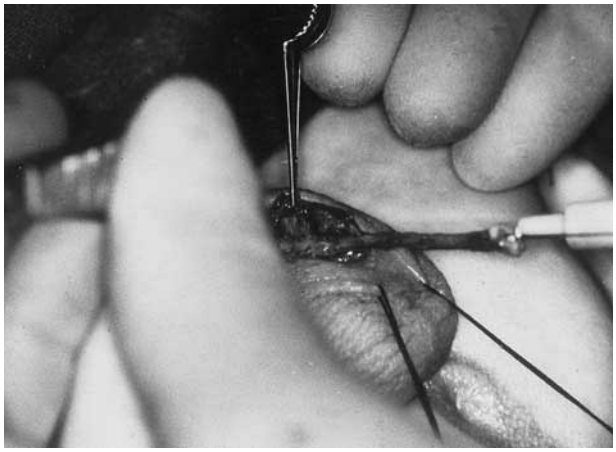

Fig 3. Fistulous tract being brought out at peno-scrotal junction having been dissected through an incision in the scrotum.

the cloaca by the urogenital septum in boys, the lower part of which represents the perineal mound, the inner genital folds close in the anterior urethra by a process of midline fusion. This begins posteriorly at the anus and extends forward to the penile meatus. The outer lateral folds also migrate medially blending into the inner folds and meeting in the midline to form the raised up perineal raphe characteristic of the male and enlarge locally to form the scrotum. If the perineum has fully developed with migration of the anus almost to the normal site, excessive posterior fusion of the folds may cover the anus and project a fistulous tract a short distance anteriorly in the perineum, the anal cutaneous fistula. With only partial development of the perineum, the covering process may roll the fistula more anteriorly in the raphe to the scrotum. With poor development of the perineal mound, the rectum remains close to the urethra, and the fistula then communicates with the bulb of the urethra, the rectobulbar fistula. In this condition, the urethra and rectum initially have separate openings, which subsequently become enclosed by the genital folds to issue to

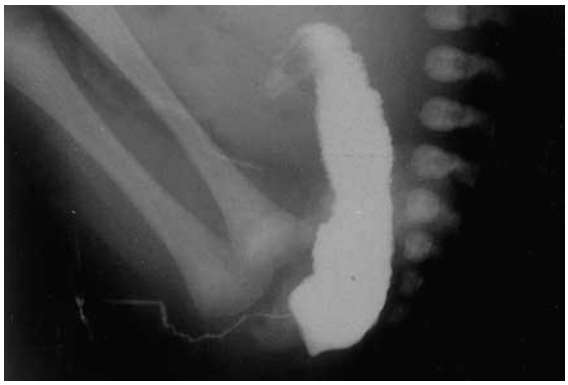

Fig 4. Fistulogram shows narrow fistula opening into large bowel.

Table 1. Clinical Presentation and Management of Nine Patients With Translevator Anal Anomalies

Patient No./ Surgeon	Year of Birth	External Findings	Opening	Surgical Procedure		Associated Anomalies	Outcome	Histology
				Early	Late			
1/Nixon	1951	Covered anus	Fistula opening at penoscrotal junction, meconium present	Fistula probed, cutback		Sagittal craniosynostosis, left corneal opacity, micrognathia, kyphoscoliosis, rib anomalies, tetralogy of Fallot	Normal stools, Died at 6 months of pneumonia	None
2/Bjordal	1975	Covered anus	2 bullae at penoscrotal junction, filled with meconium	Bullae penetrated, probed (Fig 1), cutdown onto probe at anal site	At 8 months, scrotum laid open and fistula excised 2 cm from anal verge	None	At 10 years, fully continent, occasional constipation	None
3/Bisset	1976	Covered anus	Fistula opening at penoscrotal junction, small hillock of skin at anal site, small cleft to right of anal hillock, meconium present (Fig 2)	Incision over cleft to right of anal hillock, perineal anoplasty, fistula probed, proximal and distal ends laid open		Absent left kidney, malrotated right kidney with single ureter opening into left side of bladder, ureterovesical obstruction	Fully continent, constipation	None
4/Bjordal	1976	Anal agenesis, intermediate type, 4 cm lipoma left perineum	No visible opening	Colostomy	At 7 months, found to have intermediate anomaly with narrow fistula opening near urethral meatus, incision at anal site, no pouch visible, thus laparotomy, pouch 2 cm from skin, rectal pouch sutured to skin, lipoma excised	Perineal hypospadias, bifid scrotum	Fully continent, constipation	None
5/Guiney	1976	Covered anus	Bulla at penoscrotal junction with meconium	Bulla burst, fistula probed, cutdown anoplasty	At 1 month, anal stenosis, further anoplasty. At 4 years, fistula laid open and excised	Hypospadias, duplex collecting system left side	Fully continent	Fistulous tract, squamous epithelium
6/Nixon	1977	Covered anus	Opening at ventral surface of penis, discharging meconium	Fistula probed, cutdown onto probe, perineal anoplasty	Faecaluria, fistula tract excised	Diverticular Müllerian remnant in posterior urethra	Fully continent	
7/O'Donnell	1984	Covered anus	Fistula opening at penoscrotal junction, meconium present	Anterior based flap turned into Anoplasty	At 6 weeks, anal stenosis, further anoplasty	Spina bifida occulta	Incontinent	
8/Fitzgerald	1986	Anal agenesis, intermediate type	No visible opening	Sigmoid colostomy	Day 6, bead of meconium at penoscrotal jct, tiny fistula opening closely adherent to urethra, at 2 weeks fistulous tract laid open (Fig 3), brought to neo-anal site and excised at bowel level, pouch 1.5 cm from skin, mobilized and sutured to skin, fistulogram illustrates narrow fistula (Fig 4), colostomy closed at 4 months	None	Fully continent, constipation	Fistulous tract, squamous epithelium
9/Monclair	1993	Covered anus	Bulla at penoscrotal junction	Bulla opened, tract probed, fistula laid open, proximal end brought down to anoplasty site		None	Fully continent	None

NOTE. Translevator anal anomalies with cutaneous fistulae passing deep to the scrotum.

the exterior through and along a bulbo-urethral conduit. This is associated with anal agenesis.

It is possible, in the cases in our series that appeared to be covered ani, the lateral genital folds had formed excessively, but the scrotum was normally formed pushing the opening distal to the scrotum. In the deeper varieties, it would seem likely that both the internal and external genital folds form excessively with some deficiency of the perineal mound.

It is interesting that 2 of the anal agenesis patients had hypospadias with the urethral opening at the site of the fistula with rather intimate walls. This would perhaps suggest an excessive production of the inner genital fold proximally and a deficiency anteriorly leading to the hypospadias.

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