"Perineal Canal": A Rare Anorectal Malformation of Variable Complexity

Authors L. Burgos, S. Rivas, J. A. Tovar

Affiliation Department of Pediatric Surgery. Hospital Universitario "La Paz", Madrid, Spain

Key words
- perineal canal
- female
- anorectal malformation
- double vagina

Abstract

Introduction: Perineal canal (PC) is a fistulous tract that communicates the anus with the vulvar vestibule. Our aim is to show the variable spectrum of complexity of this caudal malformation.

Patients and Methods: Patient 1 had a single urogenital orifice and a normal anus. After sigmoidostomy, a uterovaginal duplication was discovered upon opening the canal that was repaired at puberty. Patient 2 had a single urogenital orifice, a huge dilatation of a single vagina and an apparently normal anus. During perineal operation without diversion, the true anus was found which opened into the PC. Patient 3 had a single urogenital orifice, a uterovaginal duplication and an apparently patent anus. After colonic diversion, the PC was taken down revealing a normal urethral opening, the absence of an anus and a high rectovaginal fistula at the intervaginal wall. A posterior sagittal approach allowed extensive mobilization of the fistula and the rectum, fashioning of a single vagina and positioning of the rectum within the striated muscle complex.

Results: The cosmetic results were satisfactory. Patients 1 and 2 have normal continence and a near-normal perineal anatomy. Patient 3 is too young to assess continence.

Conclusions: 1) Perineal canal is a malformation that may involve the genitourinary system as well as the rectum and perineum. This developmental disturbance of the caudal end is profound as judged by the vaginal duplication and the nearly cloacal pattern of some cases. 2) Repair may be as challenging as that of a regular cloaca requiring an individualized approach.

Introduction

Perineal canal (PC) is a female anorectal malformation consisting of a median perineal fistula between the anus and the vulvar vestibule [8]. Both the anus and the vulva may be normal, but in some cases they are not. This malformation has also been referred to as "double termination of the GI tract", "H-type" or "N-type" anorectal malformation and is usually classified among the rarer varieties in standard classifications [7,8,9]. The purpose of the present study is to point out that this condition is not always a simple one and that its variable degree of complexity requires an approach individually tailored to each case.

Patients and Methods

Patient 1. This patient was referred to us for constipation and abnormal genitalia at the age of 5 months. She had a single urogenital orifice and a normal anus without additional malformations. Upon endoscopy of what appeared to be a urogenital sinus, the instrument emerged at the anus through a PC. In addition, two vaginal openings were found in the depth of the PC, midway between the urethra and the anal canal. A colostomy was performed and, some weeks later, the PC was taken down by division of the superficial wall, excision of the mucosa and closure of the skin, leaving the double vaginal opening in a normal vulvar location. At puberty, both vaginas were joined by stapling and cutting of the common wall to achieve a normal vaginal width (**Fig. 1**).

Patient 2. This newborn girl was sent to us for severe constipation. In fact, she stooled with considerable difficulty through what appeared to be a narrow, well located anus. In addition, the mother had observed that some fecal material was being passed through a single urogenital orifice. There were no other associated malformations. Urogenital imaging and barium enema

received June 23, 2008
accepted after revision July 7, 2008

Bibliography
DOI 10.1055/s-2008-1038909
Published ahead of print: 14 November 2008
Eur J Pediatr Surg 2008; 18: 392 – 394 © Georg Thieme Verlag KG Stuttgart · New York · ISSN 0939-7248

Correspondence
Prof. Dr. Juan A. Tovar
Department of Pediatric Surgery
Hospital Universitario "La Paz"
Paseo de la Castellana 261
28046 Madrid
Spain
jatovar.hulp@salud.madrid.org

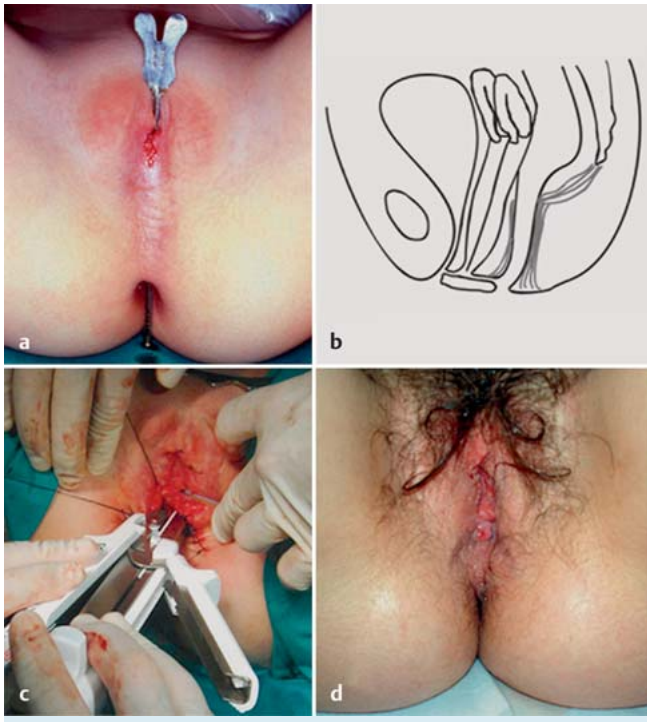

Fig. 1 a to **d** Patient 1 had a single urogenital orifice and a normal anus (**a**). A perineal canal communicated with both orifices. A double vagina drained into the canal (**b**), which was longitudinally opened. Both vaginas were joined by stapling and section at puberty (**c**), resulting in a normally appearing vulvo-perineal area (**d**) with a normal vaginal caliber.

showed a narrow hindgut exit, a normal bladder and a single, largely dilated vagina. Endoscopy confirmed that the malformation fitted within the PC pattern and it was decided that simple opening of the fistula would suffice. Therefore, the canal was taken down with diathermia over a probe, whereupon it was found that, in fact, the true anus and the vaginal orifice were hidden by the thick cutaneous-subcutaneous labio-perineal fusion that formed the superficial wall of the PC. The edges of the open canal were individually sutured, producing a near-normal vulvo-perineal anatomy (**Fig. 2**).

Patient 3. This newborn girl had abnormal genitalia and a narrow, normally located orifice that appeared to correspond to the anus. The urethral orifice was patent, but the vaginal opening was partially covered by a labial fusion with only part of the hymen visible immediately behind the urethra. The "anal" orifice was narrow and no crypts were clearly visible. She also had a left crossed renal ectopy and scoliosis with an L2 hemivertebra and posterior anomalies of L5 and S1. Upon endoscopy, the PC could be diagnosed and, due to the abnormal aspect of the anus, a sigmoidostomy was performed. A few months later, in the prone position, the superficial wall of the canal was longitudinally opened, revealing the absence of an anus underneath it and the presence of a clear anal dimple in a normal anatomical position. The posterior opening of the canal was, in fact, the result of the anterior labio-perineal fusion. There was a wide vaginal opening, corresponding to an incomplete vaginal duplication and the urethra was normal. PSARP procedure uncovered a high rectovaginal fistula opening at the midline on the common mucosal wall of an incomplete vaginal duplication. The fistula was divided, the rectum widely mobilized, both vaginas were joined, the terminal colon was placed within the striated muscle complex after dissecting 2 centimeters of thin fistula and the perineum was refashioned as in a regular cloaca (**Fig. 3**). The postoperative course was uneventful, and the colostomy was closed 3 months later after anal calibration.

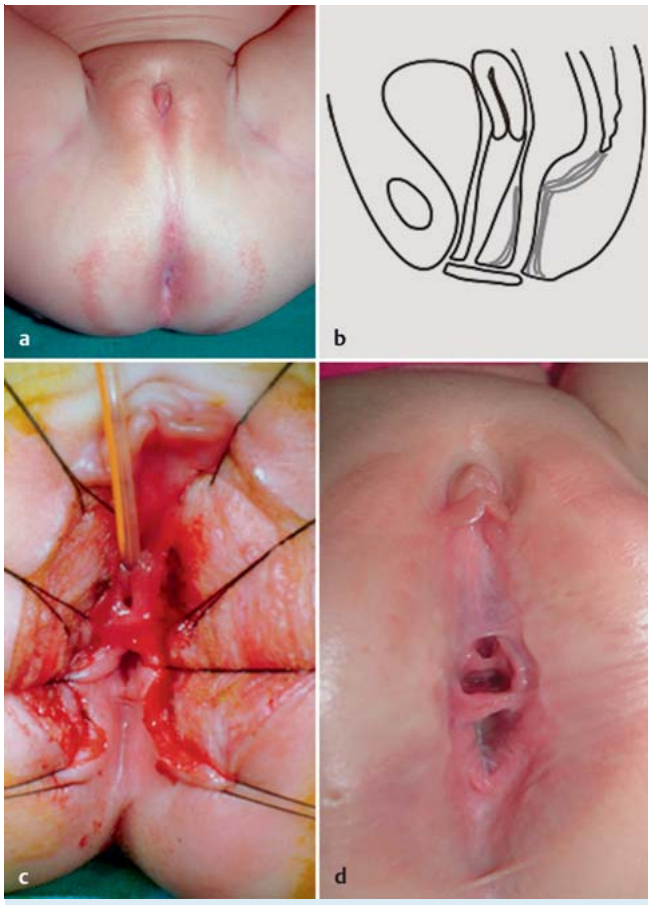

Fig. 2 a to **d** In patient 2 there was a single urogenital orifice and an apparently normal anus (**a**). Fecal material was passed through both orifices with extreme difficulty. A perineal canal was found to communicate with both orifices. In fact, the canal was an extended fusion of labio-perineal tissues which covered a rather normal anus, a single vagina and a near-normal urethra (**b**). The bridge was cut, exposing the three orifices (**c**) and, after suturing the edges, a near-normal perineal anatomy was achieved (**d**).

Results

The cosmetic results were largely satisfactory in all cases. The vaginal width returned to normal. There were no urinary infections, continence for urine and feces is normal in the first two patients and seems to be good in the third. Menstruation is normal in patient 1, the only one who has reached pubertal age.

Discussion

Perineal canal is usually classified among the "rare variants" of the anorectal malformations [7,8,9]. It has also been referred to as double termination of the alimentary tract [1,10], rectovestibular fistula [11] or H-type [3,7] or N-type anorectal malformation [13], to include in this concept some male cases with rectourethral fistulas and a normal patent anus. Probably, the term

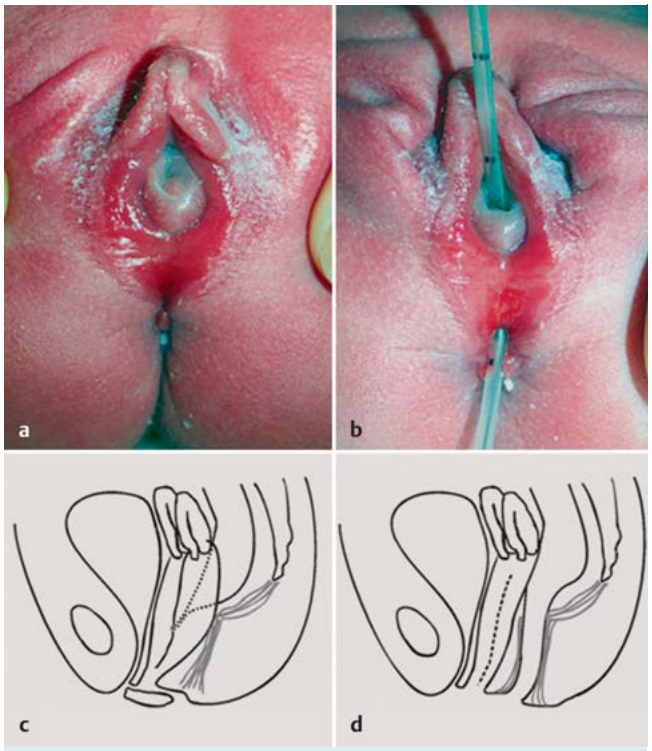

Fig. 3 a to **d** Patient 3 had a narrow "anus" and a labial fusion that partly covered the vaginal orifice (**a**). A probe was passed through the perineal canal (**b**). After sigmoid diversion, the canal was taken down, revealing the absence of an anus underneath the fusion of labio-perineal tissues, a double vagina and a high rectovaginal fistula (**c**). Repair (**d**) was performed by posterior sagittal anorectoplasty (**d**).

"perineal canal" is only appropriate when describing superficial fistulas connecting the anal canal with the vulvar vestibule as defined by Stephens and Smith [8], and all the remaining variant, including our patients 2 and 3, would be better referred to using more descriptive terms such rectovestibular or rectovaginal H-type fistula [3].

The incidence is apparently variable, since some relatively large series have been reported from Asia [13] with very few cases from other countries. Several authors have suggested that PC might be acquired after perineal infection [6,10]. However, there is no doubt that the histology and the anatomical patterns of most documented cases and of our own are consistent with a malformative mechanism in which excessive fusion of the ectoderm and mesenchym of the genital folds and perineum and sometimes abnormal Müllerian confluence at the cloacal region are involved [1,10]. This canal usually opens posteriorly at the anus, below or above the anal crypts and anteriorly at the vestibular area of the vulva. However, as in our patient 3 and others [3, 10], it can involve an absence of anus and rectum and a high rectovaginal fistula. In this eventuality, the pattern is cloacal-like, although the urethra opens separately and, at least in our case and in that of Tsuchida et al. [10], there was Müllerian duplication. In our case there were also vertebral and renal anomalies.

The malformation is usually diagnosed after investigation of abnormal anal and/or urogenital orifices and occasional passage of feces through the latter, but sometimes the leading symptom is constipation. Associated malformations of other organs are infrequent in females, but frequent in male patients in whom the fistula is often above the levator muscle [13]. The fact that our only case with supralevator rectal agenesis (patient 3) had vertebral and renal malformations further reinforces this concept.

Immediate repair by simply opening the PC with or without excision of the remaining mucosa [2,4,6] or perineal fistulectomy [4,10–12] are probably the best approaches in most of the simpler cases, but whenever in doubt about the anatomy, a sigmoidostomy should be performed in order to allow for a more accurate diagnosis and planning of a more appropriate surgical approach [3,4,7,11–13]. The operation itself cannot be considered a simple one [2], at least in some cases. It may involve a challenging repair similar to that of a long-channel cloaca and requires adequate positioning, muscle stimulation, careful repair of the vaginas and repositioning of the colonic fistula at the anal dimple.

This short series illustrates the need to employ all resources for the repair of severe anorectal malformations in order to solve the variable degree of complexity of this malformation. Our results were good in terms of anatomy and continence, although the final assessment of our more complex case will not be possible until an older age.

Conflict of Interest: None

References

1 *Chatterjee SK.* Double termination of the alimentary tract – a second look. J Pediatr Surg 1980; 15: 623–627
2 *Ismail A.* Perineal canal: a simple method of repair. Pediatr Surg Int 1994; 9: 603–604
3 *Kulshrestha S, Kulshrestha M, Prakash G, Gangopadhyay AN, Sarkar B.* Management of congenital and acquired H-type anorectal fistulae in girls by anterior sagittal anorectovaginoplasty. J Pediatr Surg 1998; 33: 1224–1228
4 *Mirza I, Zia-ul-Miraj M.* Management of perineal canal anomaly. Pediatr Surg Int 1997; 12: 611–612
5 *Murphy F, Puri P, Hutson JM, Holschneider AM.* Incidence and frequency of different types and classification of anorectal malformations. In: Holschneider AM, Hutson JM, eds. Anorectal malformations in children: embryology, diagnosis, surgical treatment, follow-up. Berlin, Heidelberg: Springer Verlag; 2006: 163–184
6 *Rao KLN, Choudhury SR, Samujh R, Narasimhan KL.* Perineal canal: repair by a new surgical technique. Pediatr Surg Int 1993; 8: 449–450
7 *Rintala RJ, Mildh L, Lindahl H.* H-type anorectal malformations: incidence and clinical characteristics. J Pediatr Surg 1996; 31: 559–562
8 *Stephens FD, Smith ED.* Ano-rectal malformations in children. Chicago: Year Book Medical Publishers; 1971
9 *Stephens FD, Smith ED, Hutson JM.* Congenital anomalies of the urinary and genital tracts. Oxford: Isis Medical Media; 1996
10 *Tsuchida Y, Saito S, Honna T, Makino S, Kaneko M, Hazama H.* Double termination of the alimentary tract in females: a report of 12 cases and a literature review. J Pediatr Surg 1984; 19: 292–296
11 *Tsugawa C, Nishijima E, Muraji T, Satoh S, Kimura K.* Surgical repair of rectovestibular fistula with normal anus. J Pediatr Surg 1999; 34: 1703–1705
12 *Wakhlu A, Pandey A, Prasad A, Kureel SN, Tandon RK, Wakhlu AK.* Anterior sagittal anorectoplasty for anorectal malformations and perineal trauma in the female child. J Pediatr Surg 1996; 31: 1236–1240
13 *White JJ, Haller Jr JA, Scott JR, Dorst JP, Kramer SS.* N-type anorectal malformations. J Pediatr Surg 1978; 13: 631–637