Anorectal-Vestibular Fistula without an Imperforate Anus in Female Infants: Report of Three Cases

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An anorectal-vestibular fistula without an imperforate anus in female infants is a rare clinical condition. This report is based on a series of 3 female infants who had fistulous communication between the bowel and the vestibule coexisting with a normal anus. Redness of the external genitalia over the labia major was the initial symptom in all 3 cases, and a suppurative discharge of pus mixed with stools was also found at the orifice of the fistula. Based on our experience, initial surgery with a double-barrel colostomy followed by a sequential fistulectomy might be safe and curative. We also review the related literature for the pathogenesis. *(Chang Gung Med J 2005;28:421-4)*

Key words: anorectal-vestibular fistula, imperforate anus.

"Double termination of the alimentary tract in females" was first introduced by Bryndorf and Madsen of Copenhagen in 1960.(1) An anorectal-vestibular fistula without an imperforate anus in female infants is still a rare condition, but it is encountered relatively more frequently in Asian countries than in Western countries.(2) The pathogenesis of this condition might be the presence of the congenital tract which is predisposed to secondary inflammation.(3) This report is based on a series of 3 female infants who had fistulous communication between the bowel and the vestibule coexisting with a normal anus. The clinical presentation and management of this condition are discussed, and the pertinent literature is also reviewed.

CASE REPORT

Case 1

A female infant was born to a xx-year-old, gravida 2, para 2 mother through a normal vaginal delivery at 39 weeks of gestation. At the age of 1 month, redness and swelling of the left-sided labia major was initially noted, and stool passing through the vestibular area near the vagina followed. An abnormal vestibular opening with a normal anus was suspected. A double-barrel sigmoid colostomy was first done to control the inflammation. The fistula was totally excised through a perineal incision 1 month later. The diameter of the abnormal opening at the vestibule was 0.2 cm. The external opening was about 1 cm from the anal verge. Histological examination revealed squamous epithelium in the wall of the fistula (Fig. 1). Serial follow-up revealed normal anal function and no recurrent fistula.

Case 2

A female infant was noted to have erythematous swelling over the left side of the labia major at 4 months of age. Some fecal material occasionally dripped out of the vestibular region. A tiny fistula was found in the 5 o'clock direction during the sub-

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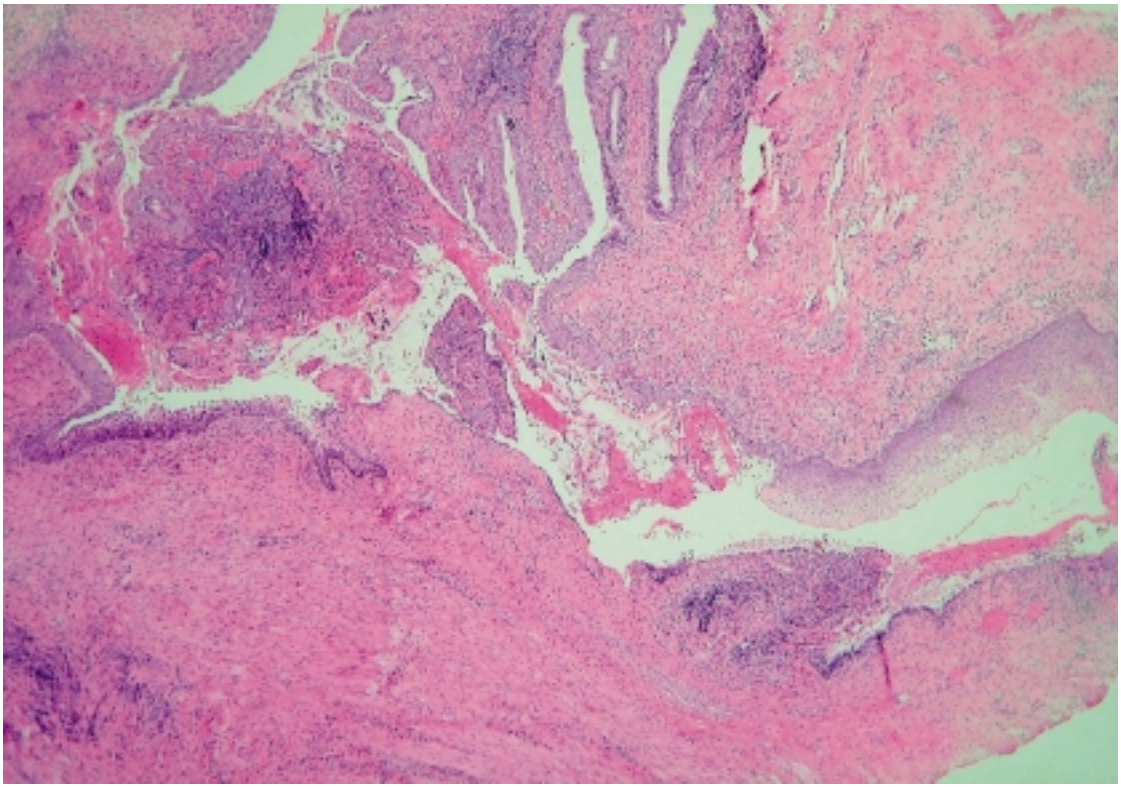

Fig. 1 Fistula tract partially lined by stratified squamous epithelium and partially by stratified columnar epithelium (H&E, ×40).

sequent colostomy procedure. The fistula was also totally excised through a perineal incision 2 months later. The external orifice of the fistula was 1 cm from the anal verge, and the squamous epithelium was also confirmed from a fistula tract specimen. There was no recurrent fistula during a 1-year follow-up.

Case 3

A preterm female infant, with a gestational age of 35 weeks, was noted to have fever with swelling and redness over the right side of the labia major at 1 month of age. She received empirical antibiotics of oxacillin and gentamicin, and obvious stool passage through the vestibular region was noted 1 week later. A double-barrel colostomy was initially performed and a fistulectomy through a perineal incision was done 1 month later. An anovestibular fistula (Fig. 2) was clearly noted at the 7 o'clock direction with an internal orifice about 0.4 cm from the vaginal opening. The external orifice was 0.5 cm from the anal verge. Squamous epithelium was found in the wall of the fistula. No fistula recurrence was noted during 6

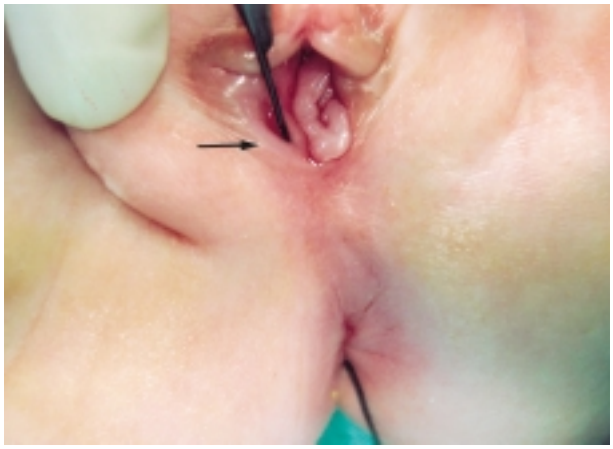

Fig. 2 An internal orifice (arrow) of the anorectal-vestibular fistula clearly noted about 0.4 cm from the vaginal opening.

months follow-up.

DISCUSSION

Since the first report by Bryndorf and Madsen of Copenhagen about the double termination of the alimentary tract in females in 1960,[1] much literature has focused on this topic. In 1969, Chatterjee and Talukder reported on 7 female infants with double termination from Calcutta, India.[3] Stephen and Smith collected similar conditions and coined the term "perineal canal" in 1971.[4] The pathogenesis of this condition might be the presence of the congenital tract which is predisposed to secondary inflammation.[3]

Anatomical levels of fistulous tracts in females can differ according to the authors. However, in reviewing a large series, Chatterjee divided these cases into 2 groups.[5] In 1 group, the tract originates in the rectum above the levator ani and resembles the usual type of rectovestibular fistula. In the other group, the tract is entirely located below the levator ani and the term "perineal canal" coined by Stephen and Smith can be properly applied.[4] White et al.[6] named it an anorectal-vestibular fistula. In our 3 cases, the fistulous tract opened uniformly below the level of the levator ani, and upon inspection, the fistula was found to open just 2 cm above the anal verge. Considering the anatomical levels of the fistulas observed in our probe identification during the operation, we believe that the term "anorectal-vestibular fistula without an imperforate anus" is accurate.

Redness with suppuration and discharge of pus over the medial aspect of the labia major was initially noted. In the meanwhile, passage of feces through the vestibule was also noted in our 3 patients. Thus presence of an anorectal-vestibular fistula was obvious. Broad-spectrum antibiotics should initially control local fistula infection. The curative surgical management is to excise the fistula tract from the surrounding tissue through a perineal approach. Adequate local incision and drainage concurrently with broad-spectrum antibiotics followed by a sequential fistulectomy might be an optional regimen. Although the role of an initial double-barrel colostomy is controversial, in our experience, we think that the first step of a double-barrel colostomy followed by a sequential fistulectomy might be a good option in the treatment of such cases not only to control local infection of the external genitalia, but also to prevent wound breakdown and recurrence after the second-stage fistulectomy.

Some controversies remain regarding the pathogenesis of this particular condition. As to the pathogenesis of the anorectal-vestibular fistula with an imperforate anus, we believe that it is an instance of mal-development in the process of the urorectal septum dividing the cloaca into a ventral urogenital portion and a dorsal rectal portion down to the pubococcygeal line in the embryo. A histological study[5] revealed squamous epithelium in the wall of the fistula, and the age of onset can confirm the "essentially congenital nature of the tract, evidence of inflammation being insignificant".

In conclusion, if a female baby is born with inflammation of the external genitalia and a normal anus, an abnormal fistula tract might be taken into consideration. Based on our experience, surgery with double-barrel colostomy followed by a sequential fistulectomy might be safe and curative.

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肛門直腸與外陰前庭間廔管發生於未合併無肛症之女嬰：三病例報告

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發生於未合併無肛症之女嬰的肛門直腸與外陰前庭間廔管在臨床上是很罕見。在此我們提出三個病例爲三位女嬰皆同時擁有一個正常的肛門以及一個肛門直腸與外陰前庭間交通的廔管。三個病例皆是以外陰部大陰唇紅腫合併前庭肛門廔管出現含有糞便之化膿分泌物。根據我們的經驗，雙管腸造口手術接續使用廔管切除術是安全且可根治的治療方式。在此我們也同時回顧醫學文獻討論可能的病理機轉及治療方針。(長庚醫誌 2005;28:421-4)

關鍵字：肛門直腸與外陰前庭間廔管，無肛症。

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