Case Report

Congenital pouch colon in a girl associated with bilateral atresia of cervix uteri and uterus didelphys

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ABSTRACT

This report describes a girl with congenital pouch colon (CPC), uterus didelphys with septate vagina, and a cloacal anomaly. The girl underwent cloacal reconstruction at the age of 15 months. Subsequently, at puberty, the child had primary amenorrhea with severe cyclic abdominal pain due to endometriosis of both the uteruses and adnexal cysts with hematometra and hematosalpinx. Laparotomy with removal of both uteri and the left fallopian tube was performed. Both uteri had atresia of the cervix uteri. This report emphasizes the need for comprehensive evaluation and a long-term management strategy for associated gynecologic anomalies in girls with CPC, especially with regard to patency of the outflow tract.

KEY WORDS: Anorectal malformation, cervical atresia, cloaca, congenital pouch colon, septate vagina, uterus didelphys

INTRODUCTION

Congenital pouch colon (CPC) is an unusual abnormality in which a pouch-like dilatation of a shortened colon is associated with an anorectal malformation (ARM).¹,² This report describes a girl with type-III CPC and uterus didelphys with a septate vagina. At puberty, the child had primary amenorrhea that was found to be due to bilateral atresia of the cervix uteri. Similar findings in a girl with CPC have not been reported earlier in the literature.

CASE REPORT

A 3-month-old girl was brought with the absence of the anal opening and passage of urine and stools from a single perineal opening. On examination, there was a single, fairly wide, perineal opening, and a clinical diagnosis of a persistent cloaca was made. Laparotomy revealed a type-III CPC malformation with 8 cm of normal colon proximal to the distended colonic pouch that appeared to end in a colocoloacal fistula. The child had uterus didelphys with one fallopian tube and uterus flanking the distal portion of the colonic pouch and the colocoloacal fistula on each side. The fistula was ligated, subtotal excision of the colonic pouch with tubularization of its outer portion performed (tubular colorraphy) and an end-colostomy fashioned.

Postoperative abdominal ultrasonogram (US) and intravenous urogram (IVU) were normal. Radiographs showed a normal sacrum. At the age of 15 months, one-stage repair of the cloacal malformation was performed. Initial dissection by the posterior sagittal approach revealed a short but wide common channel (around 2 cm long) with a double vagina. The
anterolateral walls of the common channel were tubularized to reconstruct the urethra over an 8 Fr. catheter. With the child in the supine-lithotomy position, a short segment of ileum was isolated, its upper end sutured to the margins of the double vagina and its lower end brought to the perineum. The colostomy was mobilized and brought down to the site of the neoanus. A protective loop-ileostomy was constructed and closed 3 months later.

The girl was lost to follow-up for 5 years and brought again with complaints of urinary and fecal incontinence. The child had partial urinary incontinence (UI) with passage of urine in a stream a few times daily along with continuous dribbling of urine. On examination, the urethral and vaginal openings were separate and the vaginal opening was adequate in caliber. The child was advised daily administration of saline enema at home. A micturating cystourethrogram (MCU) showed a bladder of adequate capacity with a patulous bladder neck. The parents were not willing for clean intermittent catheterization or any further surgical management for UI.

At 11 years of age, the girl presented with primary amenorrhea and severe monthly cyclical pain in both flanks and the pelvis. Investigations were advised but the child was lost to follow-up for 3 years and brought again only when the symptoms had worsened considerably. A US of the abdomen and pelvis showed well-defined lobulated masses with cystic lesions in the pelvis and both adnexal regions. The ovaries and uteri could not be seen separately from the pelvic masses. A diagnosis of hematometra and hematocolpos was made. Magnetic resonance imaging (MRI) scan of the abdomen and pelvis [Figure 1] showed 2 uteri and fallopian tubes that were distended and filled with blood and blood products. Poorly developed cervical tissue was seen and could be traced only to the right uterus. The vagina was around 4 cm long. Bilaterally, the adnexal structures appeared to be filled with blood and hemorrhagic cysts.

EUA with cystourethroscopy showed a separate urethral orifice leading to a short urethra with a widely open bladder neck. Per-vaginal examination showed tense, hard masses on both sides in the pelvis. No cervix could be felt. Laparotomy revealed dense pelvic adhesions, uterus didelphys with both uteruses distended with blood and blood products, bilateral endometrial cysts, and a large left hematosalpinx [Figure 2]. The endometrial cysts were aspirated, the walls of the cysts were enucleated, and bilateral hysterectomy with left salpingectomy performed. There was very poorly developed cervical tissue without a lumen on the right side and complete cervical atresia on the left side. The postoperative period was uneventful. The histopathological examination of the endometrial cysts showed evidence of endometriosis. Sections from one of the uterine horns showed endometriosis while those from the other horn were unremarkable.

Presently, at the age of 16 years, the girl is relieved from her complaints of cyclical abdominal and pelvic pain. She is able to hold urine during the day for periods ranging from ½ to 2 h but has severe nocturnal UI. The child is dry after an enema for 4-5 h. The child is fairly well-adjusted psychologically. The parents are not willing for any procedure for treatment of UI.

**DISCUSSION**

Although our study[3] suggested that the anomalous clinical anatomy of CPC in girls as described by us was almost invariable, apart from a few exceptions,[4-6] other reports, including earlier ones from our center,[1,7] have not consistently described the abnormal findings of the
Mullerian structures. A likely reason is that in newborns and small infants, the anatomy of the external genitalia and perineum is not clear on clinical examination and endoscopic examination of the genitourinary tract is usually not feasible so that, as initially in our patient, the usual diagnosis would be that of a persistent cloaca. The vast majority of reports, including large series,\textsuperscript{[1,2,7,8]} have recorded only the findings at the time of primary surgery, usually in the newborn, and not the results of any detailed examination, endoscopy, and/or investigations performed later. The details of reconstruction of the lower genitourinary tract have also usually not been described.

There are very few reports of the obstetric implications of a double uterus and vagina (uterus didelphys) a finding that appears to be invariable in girls with types I-III CPC.\textsuperscript{[9]} However, a report suggested that 18% of patients with uterus didelphys had an obstructed hemivagina with hematocolpos.\textsuperscript{[9]} Between 21\\%\textsuperscript{[9]} and 32.9\\%\textsuperscript{[10]} had a miscarriage. In a recent review on the subject of gynecologic concerns in girls with ARM, Breech\textsuperscript{[11]} emphasized the role of vaginoscopy before puberty, preferably at the time of definitive repair of the ARM. Vaginoscopy allows evaluation of the vaginal anatomy and can also document the appearance, development, and position of the cervices in the vaginas and the presence or absence of mucus at the ectocervix (to infer patency).\textsuperscript{[11]} Antegrade/retrograde perturbation of the Mullerian structures during any open or laparoscopic procedure for the ARM can confirm patency of the outflow tract.\textsuperscript{[11]} Any underdevelopment of the Mullerian structures can be detected by serial US starting soon after the onset of breast development.\textsuperscript{[11]} If neglected, as in our case, onset of menarche in the presence of obstruction to the outflow tract may result in endometriosis, hematometra and/or hematocolpos, hematosalpinx, adnexal cysts, and chronic abdominal pain.\textsuperscript{[11]} Although in the past, hysterectomy was the treatment of choice for cervical atresia, current recommendations are suppression of menses with preservation of the uterus for pregnancy with reproductive assistance, or uterovaginal anastomosis.\textsuperscript{[12]} A report of 18 cases of uterovaginal anastomosis in patients with cervical atresia reported six spontaneous pregnancies in four patients, and only one required cervical cerclage.\textsuperscript{[12]} Levitt, \textit{et al.}\textsuperscript{[13]} managed a patient of vestibular fistula with the absence of vagina and cervical atresia by anastomosing a bowel graft directly to the uterus.

In conclusion, our report emphasizes the need for a comprehensive evaluation and long-term management strategy for associated gynecologic anomalies in girls with CPC, especially with regard to the patency of the outflow tract. Uterus didelphys with a septate vagina appears to be invariable in girls with types I-III CPC\textsuperscript{[9]} and needs to be assessed and managed appropriately with awareness of the possibility of obstetric complications in later life.\textsuperscript{[11,14]} Patency of the outflow tracts should be assessed at an early stage and serial US starting soon after breast development can assess the growth of both uteruses and any evidence of obstruction to the outflow tract so that early corrective treatment can be instituted.

REFERENCES