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Congenital pouch colon associated to a cloaca malformation in a syndromic newborn: A case report

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ABSTRACT

Introduction: Congenital pouch colon (CPC) is a rare intestinal anomaly usually associated with an anorectal malformation (ARM). Because of its low incidence, there are no guidelines on the correct treatment. We report the step-by-step management and challenging surgery of a newborn with CPC and cloaca to ensure the patient's best quality of life possible.

Case presentation: We describe a case of type II CPC associated with a complex cloaca with a retrovesical didelphus uterus in a 1530g preterm syndromic female. A preoperative cystoscopy identified a urogenital sinus with an anterior bladder and a posterior vagina. The abdominal exploration done on day 1 showed a dilated CPC with the cecum ending in a large structure extending toward the bladder, and a proximal diverting ileostomy was created. Eight months later, the CPC was mobilized, and the common wall of the bladder and the pouch colon was excised. The 8-cm colonic pouch was tabularized by a triangle resection resulting in a colonic tube used to create a colostomy. Because of the syndromic nature of the patient, the consensus has been not to proceed with the urogenital reconstruction and the ileo-anal pull-through.

Conclusion: CPC malformations should be suspected in the case of a single perineal orifice. There is no consensus about the best surgical approach to CPC. The operative reconstruction must be tailored to each patient in order to ensure the best possible quality of life.

1. Introduction

Congenital pouch colon (CPC) is an uncommon congenital intestinal anomaly in which the colon is partially or wholly replaced by a pouch-like dilatation distally connected to the urogenital tract through a large fistula [1]. It is associated with an anorectal malformation (ARM) [2]. In Europe this malformation is extremely rare, but it represents 4.38% up to 18.7% of all anorectal malformations in Indian newborns [3]. Its incidence is higher in boys, with a male-to-female ratio from 2.25:1 to 7:1 [4]. Narasimharao et al. classified congenital pouch colon (CPC) into four sub-types. Type I CPC involves the ileum directly opening into the colonic pouch with no normal colon, while Type II CPC has the ileum opening in the cecum and then into the colonic pouch. In Type III, a 7–8 cm normal colon is present between the ileum and the colonic pouch, and in Type IV, normal colon and a colonic pouch are formed by rectum and sigma [5]. While boys typically present with CPC that culminates in a colo-vesical fistula, with the diagnosis made at birth, female newborns may be diagnosed at birth or later, particularly if meconium evacuation occurs through the fistula [6,7]. The rarity of

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this congenital malformation precludes the existence of guidelines or standardized protocols describing the appropriate management of this patient group. This report details a case of a neonate presenting with CPC and associated cloaca.

This manuscript was prepared following the CARE guidelines.

2. Case description

A 1.530 g girl was born at 31 weeks of gestation by transvaginal delivery to a 33-year-old gravida. An ultrasound exam (US) conducted at the 24th week of gestation indicated a suspected diagnosis of ano-rectal malformation. At birth, she was immediately intubated and transferred to the Neonatal Intensive Care Unit. Physical examination revealed dimorphism with bulging eyes, poorly hemmed ears, a cloverleaf appearance, and a single perineal opening (Fig. 1). Meconium drained spontaneously through the single orifice within the first 24 hours of life.

An abdominal X-ray performed after birth did not show gas in the pelvis area. A spinal X-ray revealed a widened symphysis pubis, partial sacral agenesis and fusion of the anomalous lower lumbar vertebrae. A few hours later abdominal and pelvic MRI confirmed the presence of a congenital pouch colon associated with a complex cloaca, and a right ureteral-pelvic dilatation. A cystoscopy identified a urogenital sinus with a large bladder and a cervix filled with urine and stool. A genitogram done at the same time revealed a connection between the bladder and the large colonic sac at the upper left part of the bladder. Additionally, the contrast filled the digestive tract, which appeared normal. The exact course of the ureters into the cloaca malformation could not be clearly identified (Fig. 2). An abdominal exploration done 18 hours after birth, confirming the presence of a dilated CPC without a normal colon, with the cecum ending in a large structure extending toward the bladder. The pouch colon was opened and the intra-operative contrast study showed the small intestine on the right side of the abdominal cavity and the distal part of the pouch plugged directly at the cloacal orifice (Fig. 3). The surgical wound on the pouch colon was closed, and a Pezzer tube was left inside. The postoperative course was uneventful. Ultrasound (US) evaluation performed on postoperative day 4 showed a left lateral retro-vesical didelphus uterus, bilateral grade II/III uretero-vesical reflux, nephrocalcinosis and a left superior caliceal ectasia without pyelo-ureteral dilation. A cerebral-medullary MRI highlighted a left cerebral hemispheric microcephaly and partial agenesis of the corpus callosum without medullar anomaly.

After three weeks, a terminal ileostomy was placed in the right iliac fossa. A second cystoscopy could not clearly identify the ureteral meatus but showed two hemi-vaginas with two cervixes. During the same laparotomy, the CPC was mobilized and the common wall of the bladder and the pouch colon was identified. The upper part of the pouch colon appeared to be vascularized from the superior mesenteric artery, while the lower part in communication with the bladder seemed to be vascularized by the inferior mesenteric artery. In order to separate the colon from the bladder, the medial wall was excised and the bladder halves were sutured together to create a unique bladder cavity. The 8 cm-long colonic pouch was tubularized by resecting a triangle on the anti-mesenteric edge with 30-mm loads of an endo-GIA stapler maintaining a lumen of about 2 cm (Fig. 4). This resulted in a colonic tube measuring 9 cm in length which was used to create a colostomy at the opposite side of ileostomy trying to preserve a lowerable colonic segment. The 10 mm Blake drain was removed on post-operative day 3. She was discharged on post-operative day 10 in very good general condi-

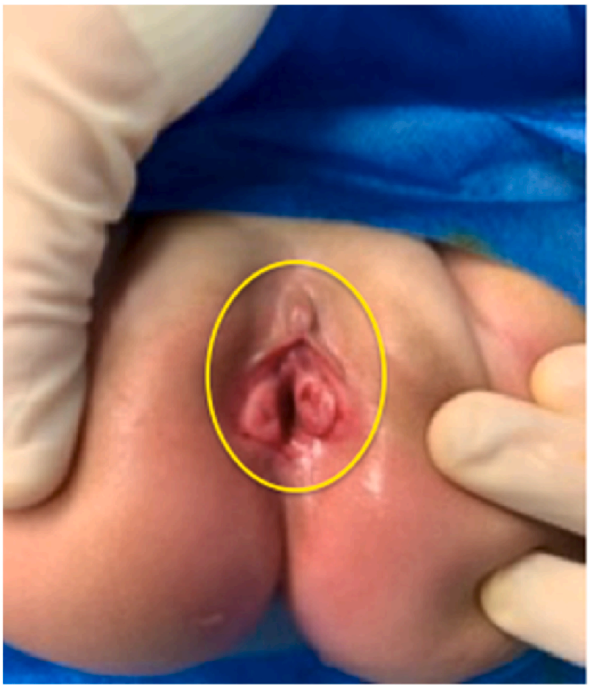

Fig. 1. Clinical examination showed a single perineal orifice with the « clover-leaf » appearance (yellow circle).

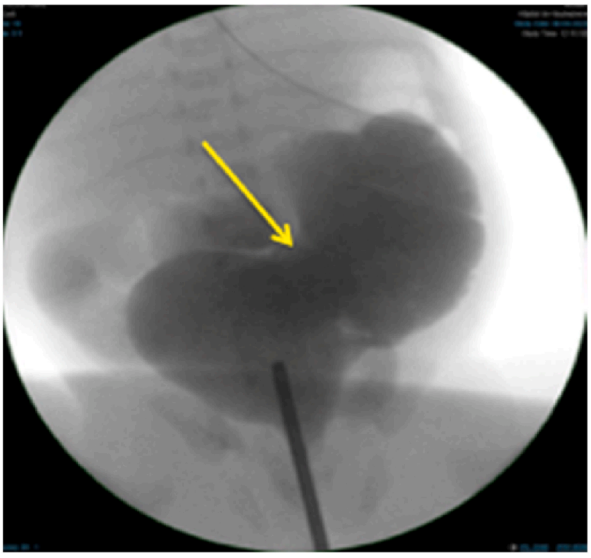

Fig. 2. Intra-operative contrast study of the lower urogenital system described the presence of a connection between the bladder and the large colonic sac at the upper left part of the bladder was identified too (yellow arrow). The contrast seemed to continue in the rest of the digestive tract. The exact course of the ureters into the cloacal malformation was not clearly identified.

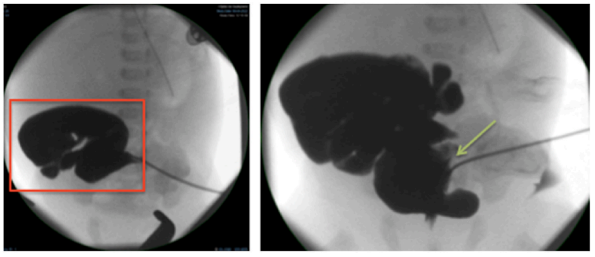

Fig. 3. A, B intra operative opacification through the pouch colon described the small intestine on the right side of the abdominal cavity (red square) and the distal part of the pouch plugged directly at the cloacal orifice (green arrow).

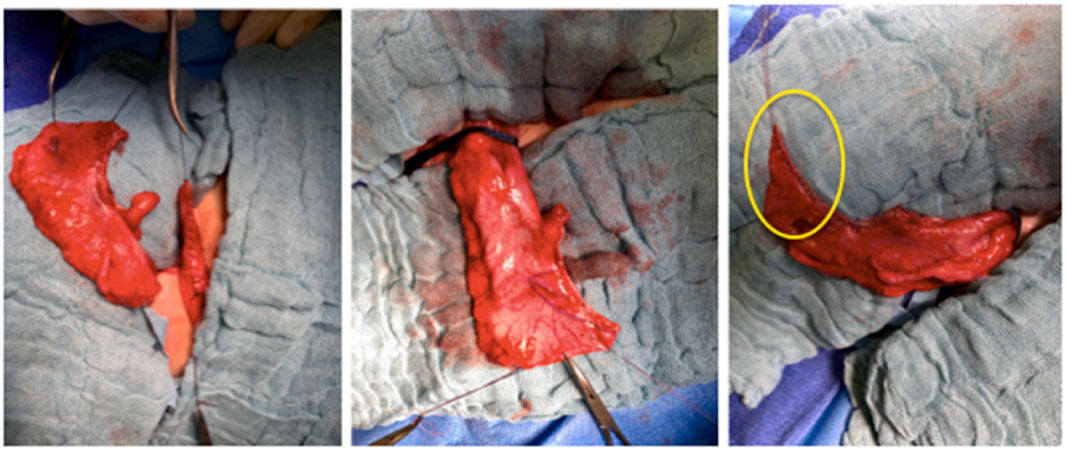

Fig. 4. A, B, C pouch colon tabularization.

tions, with a colostomy and a productive ileostomy in the right iliac fossa. Two months later, she was readmitted to the hospital for an occlusive syndrome with biliary and fecaloid vomiting, general discomfort and dehydration. Urgent US showed both dilated and empty bowel loops and abdominal X-ray showed direct and indirect signs of perforation. She underwent an urgent third explorative laparotomy showing an internal hernia at the level of the colonic segment and a bridle strangling the small bowel with a singular intestinal perforation and a consequent ischemic syndrome. The 5 cm colon causing the internal hernia and the strangulated small bowel by the bridle were resected. An end-to-end anastomosis was performed. She was finally discharged on postoperative day 17 in

good general condition. At the last post-operative clinical control, oral feeding was well tolerated, and her bowel habits were regular. A DMSA renal sonogram revealed two functional kidneys with normal relative function. At present, urogenital reconstruction and ileoanal pull-through have been discouraged due to the associated high risk of internal bladder hypertension, urinary and faecal incontinence and unsatisfying quality of life. However, physicians agreed to plan urogenital sinus' surgical treatment in the future.

3. Discussion

Congenital pouch colon (CPC) is a rare congenital anomaly in which the entire colon or part of it is replaced by a pouch-like dilatation distally connected to the urogenital tract through a fistula [1,8]. CPC was described for the first time in 1977 by Singh et al. [9,10]: in CPC the total length of the colon is shorter than normally expected (usually between 5 and 15 cm), there are no taenia coli, haustrations or epiploic appendices, and the development of the mesentery of the pouch is inadequate. Furthermore, the pouch colon is usually vascularized by the superior mesenteric artery in types I/II and by the inferior mesenteric artery in types III/IV. Anorectal malformations (ARM) can be present simultaneously [11]. The pouch-like colon ends with a colo-vesical or a colo-urethral fistula in boys, while in girls the pouch colon fistula opens into the vestibule, vagina, uterus, perineum or a persistent cloaca [2,8].

The exact incidence rate of CPC associated with ano-rectal malformations (ARM) is unknown, but it is rare worldwide [9]. Most reported cases occur in northern India, where it accounts for 7% of all ARM with a higher incidence in boys [2,11–13]. In 1984, Narsimha Rao et al. introduced the concept of CPC syndrome and proposed the first classification based on the length of the normal colon proximal to the pouch [5]. In type 1, there is no normal tubular colon and the ileum ends directly into the pouch; in type 2 cecum and a short segment of colon extend proximal to the pouch; in type 3 a normal colon extends up to the hepatic flexure; in type 4 there is a normal colon for the terminal portion, which forms the pouch [5]. Several years later, Kirhan proposed a simpler classification: type A is a partial short colon with a normal colon of varying length and type B is a complete short colon with or without a cecum [14]. Kirhan type B and Narasimaharao types 1 and 2 are the most common ones [11]. Our case reported a type 2 (according to Narasimaharao) CPC associated with a persistent cloaca. The exact embryogenesis of CPC has not been completely elucidated yet, but its etiology might be related to a decreased vascular supply to the cloaca in utero [8,14].

CPC is often associated with other congenital gastrointestinal, genitourinary, or brain anomalies [2,8]. The diagnosis of CPC can be challenging. Most of the times it is made at birth, but it can also be missed, particularly in cases of large fistulas with spontaneous evacuation of gas and meconium [2]. CPC can present with varying symptoms, from abdominal distention with or without bilious vomiting to poor general conditions with fever, bowel obstruction and shock, in case of perforation [8]. When CPC is suspected, an abdominal anteroposterior and lateral X-ray, and an invertogram are necessary to confirm the diagnosis. The radiographic hallmarks include a large loop of bowel with a single air-fluid level, in a meconium-filled pouch occupying most of the abdominal cavity [15]. If the abdominal x-ray fails to provide a conclusive diagnosis, an ultrasound and/or magnetic resonance can be of help [2].

The management of CPC is complex. Surgery is always required, and can be done in a single step or in a staged repair, depending on the clinical condition of the patient and the type of CPC. The surgical procedure aims to preserve a sufficient length of the colon, to preserve of the ileo-cecal valve, and to preserve anal continence. There are, however, no established gold standard for the repair of a CPC. A staged repair seems to be the most effective surgical option. Two procedures have been described for the correction of CPC: the first involves the resection of the pouch, followed by pulling through the ileum or the proximal colon, and the second involves the mobilization and tubularization of the pouch, followed by a corrective anorectoplasty 6–8 months later [11,14]. Both procedures are associated with potential side effects: the first option carries a high risk of incontinence, while the second one is associated with motility alterations which necessitate repetitive dilatations of the tubularized pouch [15,16]. The dilated proximal vagina, ileum, sigmoid colon and rectum can be used to reconstruct the vagina [17]. Additionally, the vaginal reconstruction can be done using the pouch colon according to Bianchi's technique of lengthening the small bowel. The pouch can be split longitudinally, with both flaps still receiving an intact blood supply. These flaps can be used to create a vagina and reconstruct the anorectum [18].

The prognosis for children suffering from CPC depends on the type of CPC, the child's general health and any associated anomalies. Mortality for CPC in the neonatal period was reported to be 8.8% by Gupta and Sharma [2]. In order to evaluate the final urinary and fecal functional outcome, long-term follow-up is mandatory.

4. Conclusion

CPC malformations should always be suspected, especially in endemic areas, in the presence of a single perineal orifice. There is no consensus on the best surgical approach, and the treatment must be tailored to each patient to ensure the best possible quality of life possible.

Statement on informed consent

Informed written consent was obtained from the parents of the child described in this study to publish the clinical data and photographs.

Informed consent

Written informed consent was obtained from the parents of the minor patient, who are the legal guardian of the baby, for the publication of any potentially identifiable images or data included in this article.

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Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

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Authors contributions

Dr. FN had primary responsibility for protocol development, patient screening, enrollment, outcome assessment, preliminary data analysis and writing the manuscript. Dr AL operated the baby. Drs FB, AL and IT participated in the development of the protocol and analytical framework for the study and contributed to the writing of the manuscript. Dr CM contributed in writing the manuscript. Dr FN did the final data analyses and contributed to the writing of the manuscript under supervision of Prof. FB, Prof IT, Dr AL, Prof FM and Prof RA.

Declaration of competing interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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List of abbreviations

CPC = Congenital Pouch Colon
ARM = ano-rectal malformations
US = ultrasound
DMSA = di-mercapto-succinil acid

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