Case Report

Functional and Manometric Outcomes after a Congenital Pouch Colon Reconstruction: Report of a Case

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Congenital pouch colon is a form of anorectal malformation, rarely reported outside north India. Hallmarks of this malformation are a short colon containing a large distal pouch with a fistula connecting to the urinary system. Herein, the authors report the case of a Thai male neonate with a congenital pouch colon type II who was initially misdiagnosed as a common imperforate anus. As a result, urinary tract infection and metabolic acidosis developed after a colostomy. A definitive surgery consisting of a tabularized coloplasty and an abdominoperineal pull-through was performed at one month of age. After closure of the colostomy, the child experienced transient loose stool with perineal excoriation for about three months and then gradually improved. At three years of age, the patient had normal bowel movements and adequate sensation, and a contrast enema showed a normal sized neorectum. An anal endosonogram revealed good localization of the rectum. A rectal manometry showed spontaneous rectal contraction and a complete rectoanal inhibitory reflex. The present case provides evidence suggesting that preservation of the native pouch colon is not contraindicated in this type of congenital pouch colon syndrome.

Keywords: *Congenital pouch colon, Pouch colon syndrome, Rectal manometry*

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Congenital pouch colon (CPC) is a rare anorectal malformation variant, in which a distal portion of the colon is substituted by a pouch-like colonic dilatation that terminates in a fistula communicating with the genitourinary tract[1-3]. The condition varies in its severity from affecting only the rectosigmoid colon to an entire colonic involvement[4]. The etiology of CPC is not clear, however, there is some evidence suggesting that the anomaly shares common embryopathogenesis with cloacal exstrophy[1]. Most of the cases have been reported from south Asian countries that are predominately populated by Indo-Aryan ethnic groups such as India, Pakistan and Bangladesh[5].

In the present report, the authors described a case of congenital pouch colon in a Thai infant treated with a straight tapering coloplasty. In addition to satisfactory clinical outcome, a post-operative endosonography and manometric study gave objective data suggesting a possibility to keep the pouch in place.

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Case Report

A full-term Thai male infant, birth weight 3,150 grams, was referred to Prince of Songkla University with problems of anorectal malformation and respiratory distress. The baby was born to a 21-year-old mother by Cesarean section. Shortly after birth, the patient developed respiratory distress that mandated an endotracheal intubation and respiratory support. On examination, the infant had a markedly distended abdomen and imperforate anus. Abdominal x-rays revealed a large air-filled segment of bowel loop, occupying the lower part of the abdomen.

On the first exploration, which was done through a small incision in the left side of the abdomen, the distal colon was found to be hugely dilated and could not be fully brought out. The cecum was found to be bifid with two vermiform appendices. Incidental appendectomies were performed. There was only a 7-centimeter length of colon between the cecum and the dilated portion of the distal colon. A

Nixon's type loop colostomy was created just proximal to the pouch colon. After the operation, although the colostomy functioned well, the patient developed hyperchloremic metabolic acidosis, which prompted us to schedule him for an early definitive operation at the age of three weeks. A loopography done before the operation showed a fistula between the dilated distal colon and the bladder neck. Bilateral hydroureters and hydronephrosis were also found by a cystography.

An exploratory laparotomy was then performed, during which an examination of the distal colon revealed a congenital pouch colon type II, according to the Saxena-Mathur's classification[6], with a 0.8 centimeter wide colovesical fistula (Fig. 1). A large amount of urine was found retained in the colonic pouch. A Meckel's diverticulum was also incidentally found. After a division of the fistula, a tubularized coloplasty was performed on the pouch colon to prepare it for an abdominoperineal pull-through. On coloplasty, the out-pouching portion was removed and the remaining bowel walls were sewn together in a tapering fashion (Fig. 1). The patient had an uneventful post-operative course.

After closure of colostomy performed six months later, the patient transiently passed loose stool and had perianal skin excoriation, but this condition gradually improved over the following three months. His body weight and height increased from lower than the third percentile at the age of 6 months to the 60 percentile at the age of 12 months and sustained around 60 to 70 percentile thereafter. Oral diet was maintained with soy-bean formula and baby food. On the initial period, the patient passed more than 10 bowel movements each day. On the final follow-up before the present report at an age of three years, the child was doing well in terms of emptying function and continence, with three to five bowel movements in a day. Most of the time, the patient could tell a caretaker when he needed to go to the toilet. According to Kelly's clinical score[7], the quality of continence in this child could be rated as good (5 points). A barium enema showed a normal sized neorectum (Fig. 2A). An anal endosonography demonstrated the proper position of the pull-through rectum within the external sphincter complex (Fig. 2B). An anorectal manometric study used the same technique as presented in the authors' previous publication[8] and followed the recommendation of DiLorenzo C et al[9]. The present study showed spontaneous contraction of the rectal pouch at a frequency of 0.7 waves per second and the mean amplitude of 2.3 mmHg. The mean resting rectoanal pressure gradient was -6 mmHg. A rectal stimulation showed complete relaxation of the high-pressure zone when the rectal balloon was blown up to 20 milliliters. The relaxation resulted in a positive rectoanal pressure gradient of 2 mmHg (Fig. 3). A cystography also showed spontaneous resolution of vesicoureteric refluxes.

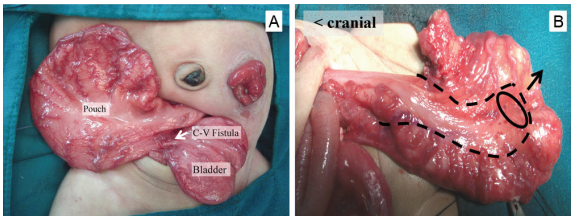

Fig. 1 A) Operative finding of a colonic pouch with a 0.8 cm wide colovesical fistula (CV fistula). B) Outline of the tabularized coloplasty performed in this patient

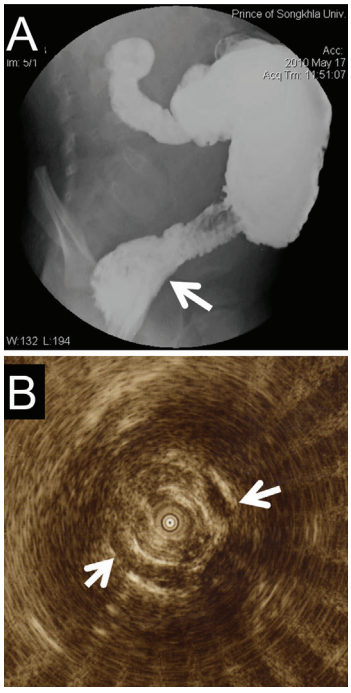

Fig. 2 A) Barium enema study performed at 20th post-operative month showed normal size neorectum (arrow) with irregular mucosal lining indicating chronic inflammation. B) Endosonography performed at 6th post-operative month showed the neoanus lying within the sphincter complex (arrows)

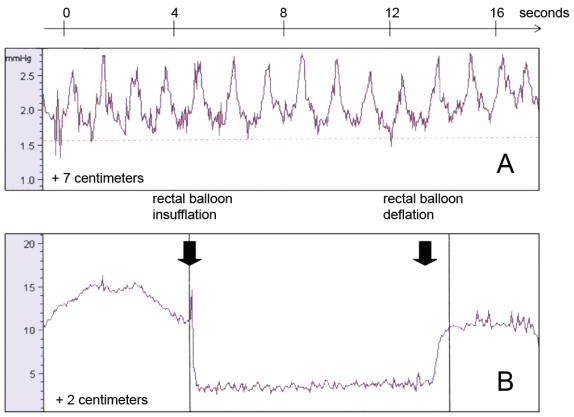

Fig. 3 Anorectal manometry with rectal pressure probe at +7 centimeters and anal probe at +2 centimeters above the anal verge; showing A) a spontaneous rectal contraction which produced mean pressure amplitude of 2.3 mmHg and frequency of 40 waves per minute B) A complete relaxation of anal pressure, responsible for rectal balloon inflation at the volume of 15 milliliters

Discussion

Although a CPC is currently recognized as a form of anorectal malformation as the nomenclature appears in the standard classification[10], this kind of anomaly remains rare outside north India. An initial film that shows a large globular structure containing both air and fluid should be a clue to the possible presence of a CPC[3]. Failure to diagnose this condition early potentially results in complications from a large fistula between the large colonic pouch and the urinary system. Mathur et al suggest that all CPCs should have their fistula ligated during the first operation, whether anorectal reconstruction is done immediately or not[3]. In the presented patient, the CPC had not been diagnosed correctly until his definitive operation and as a result, urinary tract infection and metabolic acidosis developed. However, like other common types of anorectal malformations, those problems, including vesicoureteric reflux, resolved after the colonic surgery[11].

While earlier studies recommended colonic wall reconstruction in a curved tube fashion (so called tabularized coloplasty) in order to preserve colonic length[12,13], more recently, some authors have chosen to excise the entire pouch colon based on their belief that the part contains poorly developed colonic musculature[14-16] and such a defect carries the risk of development of rectal dilatation or megarectum change[2]. In the presented patient, the authors chose to keep a portion of the pouch colon for two main reasons: to avoid taking down the colostomy and to keep a functioning colon. The authors do not believe that weak pouch musculature alone will cause a megarectum formation, but rather, as other studies have suggested also, that features such as post-operative failure of the emptying function caused by poor toilet training, or a pathological defecation mechanism, are more likely to be responsible for dilatation of the rectal reservoir[16].

The authors' operative technique was a straight tabularized coloplasty, slightly modified from the technique proposed by Wakhlu[12]. The authors removed the thin out-pouching part and kept the thicker core of pouch colon along the main tinea coli based on the understanding that this part represents a near normal colonic structure. In the medium term follow-up period as the present paper was written, the patient showed adequate emptying function and the normal features were confirmed by radiologic study. A manometric study also confirmed that the pull-through colon had spontaneous contraction and normal distal relaxation, responsive to proximal proprioceptive stimulation. This evidence contradicts the hypothesis in previous works that suggested the redilatation of the tapering colonic pouch would be a major cause of coloplasty failure[12,13,17]. The authors postulate that early reconstruction and defecation training were important factors in the success of the authors' treatment.

In summary, a case of type II CPC in a Thai infant who was treated with a three-staged coloplasty is presented. The clinical outcome, supported by a manometric study, indicates this type of conservative surgical approach for CPC can be successful.

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Potential conflicts of interest

None.

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ผลการศึกษาเชิงหน้าที่การทำงานและพลศาสตร์การทำงานของไส้ตรงหลังผ่าตัดรักษาในผู้ป่วยลำไส้เป็นกระพุ้งแต่กำเนิด: รายงานผู้ป่วย 1 ราย

สุรศักดิ์ สังขทัต ณ อยุธยา, ศักดา ภัทรภิญโญกุล, ปิยวรรณ เชียงไกรเวช

ลำไส้เป็นกระพุ้งแต่กำเนิดเป็นรูปแบบหนึ่งของความผิดรูปแต่กำเนิดของทวารหนักและไส้ตรงที่พบได้น้อยนอกประเทศอินเดียตอนเหนือ ความผิดรูปดังกล่าวประกอบด้วยการที่ลำไส้ใหญ่สั้นกว่าปกติ มีส่วนปลายป่องเป็นกระพุ้ง และมีช่องเชื่อมกับทางเดินปัสสาวะ บทนิพนธ์นี้ได้รายงานผู้ป่วย 1 ราย ซึ่งมีภาวะลำไส้เป็นกระพุ้งแบบที่สอง ซึ่งในระยะแรกรับได้รับการวินิจฉัยสับสนกับภาวะความผิดรูปของทวารหนักแบบที่พบทั่วไป กระทั่งผู้ป่วยประสบปัญหาติดเชื้อของทางเดินปัสสาวะและเลือดเป็นกรดหลังจากผ่าตัดเปิดทวารเทียม ภาวะปัญหาดังกล่าวได้รับการแก้ไขเมื่อผู้ป่วยอายุ 1 เดือน ด้วยการผ่าตัดซ่อมสร้างทวารโดยห่อทวารที่เป็นกระพุ้งให้เป็นท่อ และดึงผ่านกล้ามเนื้อหูรูดเพื่อเปิดออกเป็นทวารใหม่ หลังจากผ่าตัดปิดทวารเทียมผู้ป่วยยังถ่ายเหลวอยู่ระยะหนึ่ง และดีขึ้นตามลำดับ กระทั่งอายุ 3 ปี ผู้ป่วยมีการขับถ่ายใกล้เคียงปกติ รับความรู้สึกบริเวณทวารหนักได้ดี และมีไส้ตรงขนาดปกติเมื่อตรวจประเมินทางรังสี การตรวจด้วยคลื่นเสียงความถี่สูงพบไส้ตรงวางอยู่ภายในวงกล้ามเนื้อหูรูด และการตรวจพลศาสตร์การทำงานของไส้ตรงและทวารพบลำไส้ส่วนที่ซ่อมสร้างมีการบีบคลายตัว และมีการตอบสนองต่อการกระตุ้นให้คลายตัวด้วยแรงดันได้ดี หลักฐานดังกล่าวชี้ให้เห็นว่าการเก็บส่วนของลำไส้ที่เป็นกระพุ้งมีความเป็นไปได้ และอาจให้ผลการรักษาที่ดี