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 rectoanl 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 extrophy(5). 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(6).

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.

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 hydrourerets 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](image1.png) 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](image2.png) 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)
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.

References


ผลการศึกษาเชิงหน้าที่การทำงานและพลศาสตร์การทำงานของไสตรงหลังผ่าตัดรักษาในผู้ป่วยลำไส้เป็นกระพุ้งแต่กำเนิด: รายงานผู้ป่วย 1 ราย

สุรศักดิ์ สังขทัต ณ อยุธยา, ศักดา ภัทรภิญโญกุล, ปิยวรรณ เชียงไกร

ลำไส้เป็นกระพุ้งแต่กำเนิดเป็นรูปแบบหนึ่งของความผิดรูปแต่กำเนิดของทวารหนักและลำไส้ที่พบได้น้อยนอกประเทศอินเดียตอนเหนือ ความผิดรูปดังกล่าวประกอบด้วยลำไส้ที่สั้นกว่าปกติ มีส่วนปลายป่องเป็นกระพุ้ง และมีการเชื่อมกับทางเดินปัสสาวะ บทนิพนธ์นี้ได้รายงานผู้ป่วย 1 ราย ซึ่งมีภาวะลำไส้เป็นกระพุ้งแบบที่สอง ซึ่งในระยะแรกผู้ป่วยมีภาวะวินิจฉัยสะกิดเกี่ยวกับภาวะผิดรูปของทวารหนักแบบที่พบทั่วไป กระทั่งผู้ป่วยประสบปัญหาดังกล่าว ผ่าตัดเพื่อแลกเปลี่ยนและตัดเป็นกระพุ้ง ภาวะปัญหาดังกล่าวได้รับการแก้ไขเมื่อผู้ป่วยอายุ 1เดือน ด้วยการผ่าตัดสร้างทวารโดยห่อทวารที่เป็นกระพุ้งให้เป็นหูรูดและดึงผ่านกล้ามเนื้อหูรูดเพื่อเปิดออกเป็นทวารใหม่ หลังจากผ่าตัดมีความผิดรูปที่พบอย่างมีอาการเหลืออยู่ระยะหนึ่ง กระทั่งผู้ป่วยมีภาวะลำไส้ใกล้เคียงปกติ เมื่อตรวจประเมินทางรังสี การตรวจด้วยคลื่นเสียงความถี่สูงพบไส้ตรงวางอยู่ภายในวงกล้ามเนื้อหูรูด และมีการตอบสนองต่อการกระตุ้นดี ตามลำดับ กระเทาอายุ 3ปี ผู้ป่วยมีการขับถ่ายใกล้เคียงปกติ รับความรู้สึกบริเวณทวารหนักได้ดี และมีการตอบสนองต่อการกระตุ้นดี หลักฐานดังกล่าวชี้ให้เห็นว่าการเก็บส่วนของลำไส้เป็นกระพุ้งมีความเป็นไปได้ และอาจให้ผลการรักษาที่ดี