CASE REPORT

Use of congenital pouch colon for augmenting the neurogenic bladder in a child: a 13-year follow-up

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SUMMARY

Congenital pouch colon is an anomaly always associated with anorectal malformation, where the colon is replaced by or terminates into a large aperistaltic intestinal pouch. Vertebral anomalies leading to neurogenic bladder are rare associated malformations. The pouch is aperistaltic and thus a poor rectal substitute but this very property makes it ideal for bladder augmentation. We report the first case where the pouch has been used to augment a high-pressure neurogenic bladder.

BACKGROUND

Congenital pouch colon is an uncommon malformation seen almost exclusively in the Indian subcontinent. It comprises 5–15% of all cases of anorectal malformation in Northern India, 8–10% in Pakistan and 1.7% in Bangladesh, with sporadic case reports from other parts of the world.¹ The colon is replaced by or terminates into a large aperistaltic intestinal pouch. While this is always associated with anorectal malformation anomalies of the vertebra, the genitourinary tract and the gastrointestinal tract are rare associations.^{2–5} We describe a child with type 4 congenital pouch colon (as per the classification given by Narasimharao *et al*) and partial sacral agenesis where augmentation of the high pressure poorly compliant neurogenic bladder was performed using the pouch colon, which otherwise would have been excised.

CASE PRESENTATION

A 6-year-old boy presented to our institution in the year 2000, with uncorrected anorectal malformation and urinary incontinence. He had been diverted with a transverse loop colostomy on day three of life.

INVESTIGATIONS

At the time of presentation, the patient had a creatinine level of 1 mg/dL with creatinine clearance of 59.4 mL/min.

A pressure augmented distal colonogram revealed a type 4 congenital pouch colon with a fistula connecting the pouch to the bladder neck (figure 1).

He also had a partial sacral agenesis with resultant neurogenic bladder. Ultrasound revealed bilateral hydronephrosis.

Micturating cystourethrogram (MCU) showed right major vesicoureteric reflux and renogram revealed a right contracted poorly functioning kidney.

Cystometrogram showed a high pressure poorly compliant bladder.

TREATMENT

The patient was planned for corrective surgery. A Pfannenstiel incision was made. The baggy pouch at the terminal end of the colon situated just behind the bladder was seen (figure 2A). The fistula connecting the pouch with the bladder neck was disconnected. The normal colon proximal to the pouch was divided just above the pouch and brought down as the neorectum via a channel within the levator complex, which had already been defined by a sacral approach. The anterior wall of the pouch and the posterosuperior wall of the bladder were opened longitudinally and a wide bladder to pouch anastomosis was performed, thus completing the augmentation.

The right ureter was transected at its lower third and a right-to-left transureteroureterostomy was performed. The refluxing distal stump of the right ureter was placed in a submucosal channel extravesically and its upper end brought out to the skin to form a catheterisable channel (figure 2B). The post-operative period was uneventful. The colostomy was closed at a later date.

OUTCOME AND FOLLOW-UP

The patient has been followed up for 13 years and is continent of urine and stool due to the

Figure 1 A distal colonogram showing the pouch colon with a faecaloma inside (curved white arrow on left side) and subsequent filling up of the bladder (broad white arrow on right side).

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Figure 2 (a) Schematic diagram of intraoperative findings showing a baggy pouch colon (P) at terminal end of colon situated just behind the urinary bladder (B). A fistula (F) connecting the pouch colon with bladder neck shown by arrow. (b) Schematic diagram of procedures undertaken in operation: the fistula connecting the pouch (P) with the bladder neck was disconnected. The normal colon (C) proximal to the pouch was divided just above the pouch and brought down as the neorectum. The anterior wall of the pouch and the posterosuperior wall of the bladder (B) were opened longitudinally and a wide bladder to pouch anastomosis was performed thus completing the augmentation. The right ureter was transected at its lower third and a right-to-left transureteroureterostomy was performed. The refluxing distal stump of the right ureter was placed in a submucosal channel extravasically and its upper end brought out to the skin to form a catheterisable channel (M).

intermittent catheterisation via the ureteric catheterisation port and bowel management programme. He has had no urinary tract infections in the interim period. His current serum creatinine level is 1.75 mg/dL with creatinine clearance of 63 mL/min as compared with a creatinine level of 1 mg/dL with creatinine clearance of 59.4 mL/min at the time of presentation. Ultrasound showed stable hydronephrosis on the left side and a contracted non-functioning right kidney. MCU showed a good capacity bladder with no reflux while cystometry showed a compliant bladder with a capacity of 430 mL and maximum filling pressure of 15 cm of water (figure 3).

Figure 3 Cystourethrogram of augmented bladder.

DISCUSSION

Anorectal malformation with congenital pouch colon is an anomaly seen almost exclusively in the Indian subcontinent.^{1 6 7} In this case whole or part of the colon is replaced by a pouch-like dilatation, which in turn may communicate with the genito-urinary tract through a fistulous track.⁸

The first description of this anomaly was given by Spriggs in 1912.⁹ The term 'congenital pouch colon' was coined by Narasimharao *et al* in 1984 who also proposed the most widely accepted anatomical classification.

The classification is as follows:

Type I: Normal colon is absent and the ileum opens directly into the colonic pouch.

Type II: The ileum opens into a short segment of caecum, which then opens into the pouch.

Type III: Presence of a significant length of normal colon between the ileum and the colonic pouch.

Type IV: Presence of near normal colon with only the terminal portion of colon (sigmoid and rectum) converted into a pouch.⁸

The pouch is described to be of varying diameters ranging from 5 to 15 cm, with a short, poorly developed mesentery, thick walls, absent haustrations, absent appendices epiploicae and an absent or ill-defined taenia coli. The blood supply of the pouch is usually from the branches of the superior mesenteric artery.² The presence of the inferior mesenteric artery is variable and is seen only in type 4 congenital pouch colon as in our case.

The exact cause for congenital pouch colon is unknown. Genetic and environmental factors have been proposed as aetiological factors.^{1 10} Histological examination of the pouch has shown absence of normal differentiation of the muscle coat into the inner circular and outer longitudinal layers, with the muscles arranged in a decussating manner.¹ This has been postulated as the reason for absence of peristaltic activity in the pouch.

In most instances, the distal end of the pouch communicates with the genitourinary tract. In males, the pouch communicates more commonly with the bladder and less commonly with the urethra.² In females, a cloacal communication is most common, with vaginal and vestibular communication being less common.^{1 2}

Associated anomalies are rarely seen in the vertebra, the genitourinary tract and gastrointestinal tract.^{1 2} The incidence of vertebral anomalies as per a recent review by Gupta *et al*¹ has been put at 4.2%, that is, 24 of 566 cases.

The standard surgical management for congenital pouch colon is a staged procedure. A proximal diversion is made initially followed by ligation of the fistula and excision/tubularisation of the pouch with abdominoperineal pull-through. The undiversion is done at a later stage.^{1 11}

In this child, there was type 4 congenital pouch colon along with a partial sacral agenesis. This resulted in a high pressure poorly compliant neurogenic bladder with upper tract changes. Augmentation of the bladder was indicated to relieve the pressure. The pouch was situated posterosuperior to the bladder and, being aperistaltic, was an excellent choice for augmentation

of the bladder without mobilisation or detubularisation. Over the past 13 years, the pouch has remained a compliant augmentation providing low pressure urinary storage.

Association of pouch colon with vertebral anomalies and neurogenic bladder is rare.² However, surgeons should be aware of the potential need for augmentation in such patients and conserve the pouch in the presence of a neurogenic bladder.

Competing interests None.

Patient consent Obtained.

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Learning points

- Congenital pouch colon is an uncommon malformation.
- Vertebral anomalies leading to a neurogenic bladder are rare associated malformations.
- The pouch is aperistaltic and thus a poor rectal substitute but this very property makes it ideal for bladder augmentation.
- We report the first case where the pouch has been used to augment a high-pressure neurogenic bladder.

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