Prune belly syndrome with pouch colon with scaphoid megalourethra: a newer embryological and prognostic perspective

Saurabh Garge, Monika Bawa and Katragadda Lakshmi Narasimha Rao

We here report a rare association of megalourethra with pouch colon with prune belly syndrome. We also provide a newer embryological and prognostic perspective to this association. *Ann Pediatr Surg* 11:42–45 © 2015 Annals of Pediatric Surgery.

Annals of Pediatric Surgery 2015, 11:42–45

Keywords: megalourethra, prune belly syndrome pouch colon, scaphoid

Department of Pediatric Surgery, Advanced Pediatric Center, PGIMER, Chandigarh, India

Correspondence to Saurabh Garge, MCh, Department of Pediatric Surgery, Advanced Pediatric Center, PGIMER, Chandigarh 160012, India
Tel: +91 172 274 7585 x5320; fax: +91 172 274 4401/274 5078;
e-mail: saurabhgarge8@gmail.com

Received 15 November 2012 **accepted** 26 May 2014

Introduction

Prune belly syndrome (PBS), also known as Eagle–Barrett syndrome, comprises a triad of anomalies that include abdominal wall flaccidity, urologic abnormalities, and bilateral cryptorchidism. The incidence of PBS is between 1 in 29 000 and 1 in 40 000 live male births, with incidence four times higher in twins [1–6]. Male individuals are affected 20 times more often than female individuals. The underlying etiology of the urinary tract obstruction, severity of associated abnormalities, and the degree of pulmonary hypoplasia influence the survival and outcome of infants with PBS. Congenital pouch colon is characterized by replacement of varying length of colon by a dilated pouch that almost invariably has a wide high fistulous communication with the genitourinary tract. There have been four instances where pouch colon has been found associated with prune belly. We here describe the fifth case of such occurrence and also review the previous cases (Table 1) [1–3,7]. Our case also had scaphoid megalourethra with urethral stenosis, which is the first instance of this rare occurrence. The previous report of megalourethra was truly speaking an anterior urethral diverticulum, without any proximal or distal obstruction [1].

Case report

A term male neonate, born through normal vaginal delivery of an uneventful supervised pregnancy, was brought to our hospital at 28 h of life because of an imperforate anus and progressively increasing abdominal distension. The baby was dehydrated and hypothermic, with a birth weight of 2.05 kg. His abdomen was flabby, with visible loops of intestine. Two lumps were seen, one over the right hypochondrium and another over the hypogastrium. The lump over the right hypochondrium was 7 cm × 5 cm in its maximum horizontal and vertical dimensions, respectively. It was cystic in consistency, transilluminant, and ballotable. The lump over the hypogastrium was because of a distended bladder palpable 6 cm from the pubic symphysis. The bladder was expressible, with urine getting collected in the dilated urethra. A scaphoid megalourethra with pinpoint meatus with absent corporal tissue was found. On pressing the urethra, patient voided turbid purulent urine. The scrotum lacked rugosities and bilateral testis were undescended. They were not palpable even in the inguinal region (Fig. 1). The anal opening was absent, with poorly developed buttocks and median raphe. A cross-table prone lateral radiograph and erect anteroposterior abdominal radiograph were suggestive of a high rectal anomaly with a pouch colon (Fig. 2). Ultrasonography of the abdomen showed bilateral hydronephrosis with right-sided anteroposterior diameter of 5.5 cm. The ureters were not visualized due to excessive bowel gas. Echocardiography revealed presence of coarctation of aorta with patent ductus arteriosus and atrial septal defect of 5 mm.

Exploratory laparotomy revealed deficient muscular tissue in the abdominal wall. Intraoperatively, type IV pouch colon with a wide colovesical fistula was present. It lacked haustrations, appendices epiploicae, and taenia coli. There was an abrupt transition from the normal caliber proximal bowel to the distal dilated pouch. The bladder was grossly distended and contained purulent urine (Fig. 3). The colovesical fistula was ligated, the pouch colon was excised, and an end descending colostomy was fashioned. The bladder opening was closed and a suprapubic cystostomy was placed as the urethra was not catheterizable. Bilateral intra-abdominal testis was also found.

A dye study at the bed side showed scaphoid megalourethra with large bladder (Fig. 4). The intraoperative urine culture grew *Escherichia coli*. Blood cultures grew *Klebsiella pneumonia*. The patient developed sepsis and died on the fifth postoperative day, despite all possible measures.

Discussion

PBS, a rare congenital anomaly, exists almost exclusively in male individuals and consists of genital and urinary abnormalities with partial or complete absence of abdominal wall musculature [1,2].

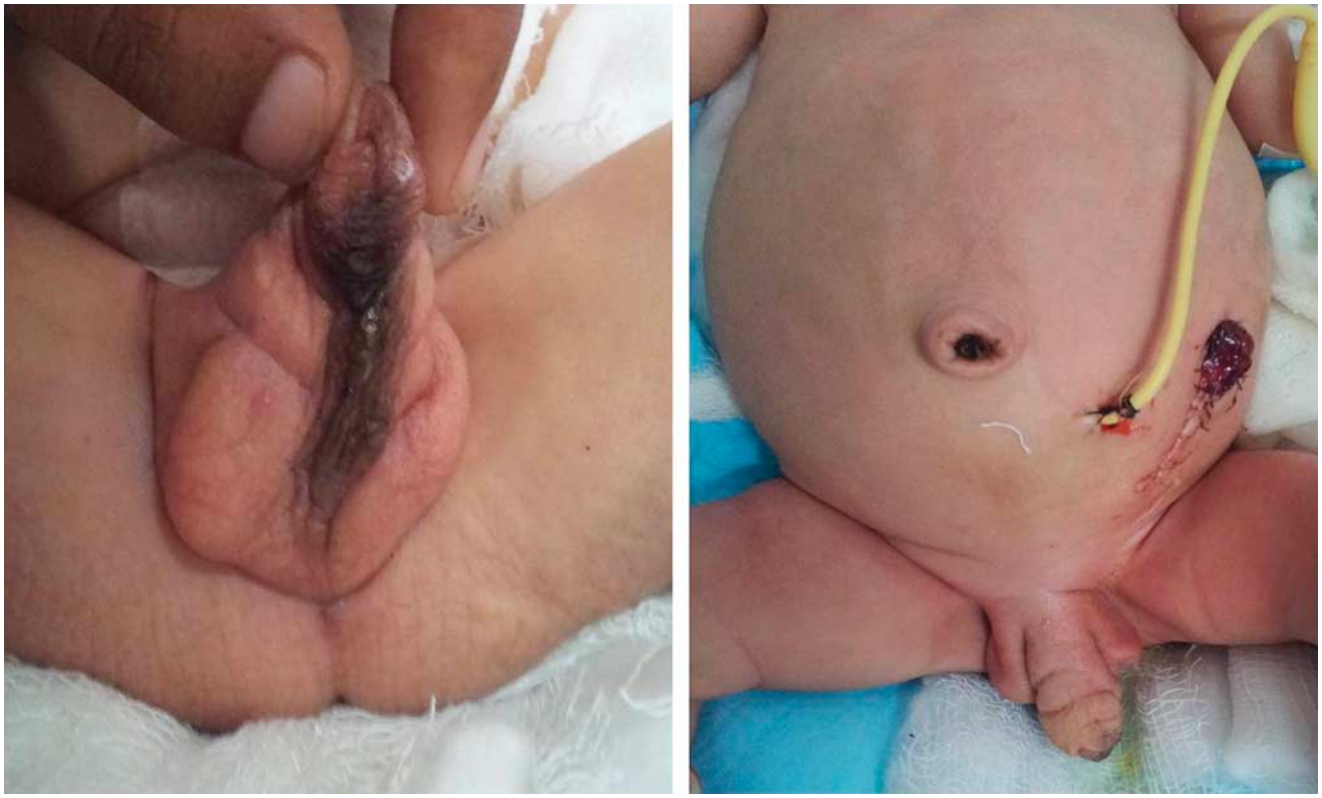
The syndrome has a broad spectrum of affected anatomy with different levels of severity. Additional associated anomalies exist involving the respiratory tract, gastrointestinal tract, cardiac system, and musculoskeletal

1687-4137 © 2015 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000452064.76439.47

Table 1 Prune belly syndrome associated anomalies

Serial numbers	References	Type of pouch colon	Urethral anatomy	Associated abnormalities
1	Bangroo *et al.* [1]	Type I	Anterior urethral diverticulum	None
2	Baba *et al.* [7]	Type IV	Microurethra	Absent dermatome
3	Raghavan *et al.* [3]	Type I	Penoscrotal hypospadias	None
4	This study	Type IV	Scaphoid megalourethra	Coarctation of aorta, patent ductus arteriosus, atrial septal defect

Fig. 1



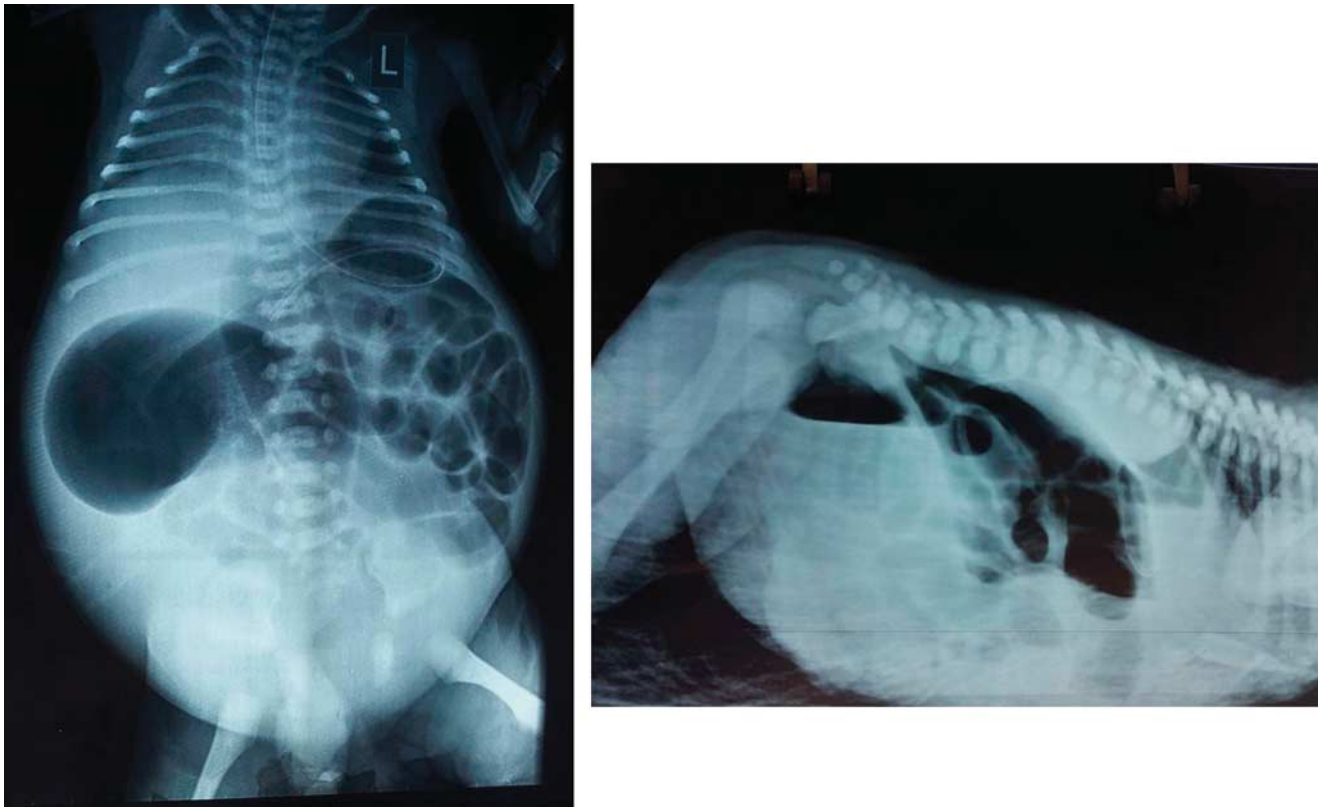
Megalourethra, empty scrotum, flaccid abdomen.

system. Gastrointestinal anomalies are seen in over 30% of these patients [3]. Malrotation, intestinal atresia, volvulus, embryonic mesentery, and wandering spleen are common gastrointestinal associations [3–6]. Exomphalos, gastroschisis, tracheoesophageal fistula, and anorectal malformations are rare gastrointestinal associations [4–6]. Pouch colon, as a rare gastrointestinal association, has been reported in only four instances before [1–3,7]. Congenital pouch colon, also known as congenital short colon, is a high anorectal malformation in which a varying length of colon is replaced by a dilated pouch that almost invariably has a wide high fistulous communication with the genitourinary tract [8]. Pouch colon has been classified into four types depending on the segment of normal colon present [8].

The anterior urethra in cases of PBS is usually normal; however, many urethral anomalies such as atresia, stenosis, and megalourethra have been reported [1,6]. Congenital megalourethra is characterized by severe dilatation of the anterior urethra and lack of penile erectile tissue development [1,6]. Absence of proximal and distal obstruction, absence of erectile tissue, and dilation of entire anterior urethra differentiate it from urethral diverticulum [1,6]. Our patient had a scaphoid megalourethra with a urethral stenosis that could not be catheterized. This is probably the first report of an association of PBS, pouch colon, and scaphoid megalourethra. Our patient lacked discernible erectile tissue; however, the previous report of anterior urethral diverticulum does not mention anything about the condition of erectile tissue [1].

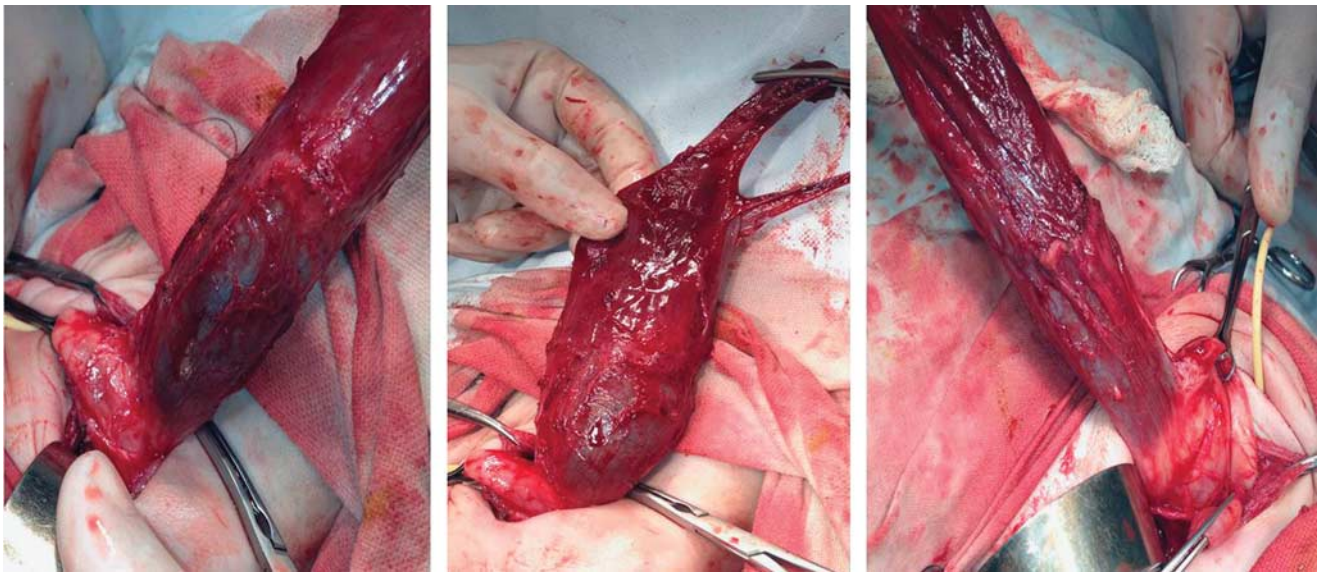
The exact etiology of PBS is unknown, although several embryologic theories attempt to explain the anomaly [1–3]. Overdistension of the abdomen due to bladder distension, mesodermal arrest, and defects in yolk sac are among the most common etiologies [1–3,7]. The association of examphalos, gastroschisis, absent dermatome, and now pouch colon with PBS lends credence to the mesodermal arrest theory, early during embryogenesis [4–7]. A vascular compromise in the region of the

Fig. 2



Erect abdominal radiograph and CTPL showing pouch colon and high ARM. ARM, anorectal malformation; CTPL, cross table pelvic lateral (view).

Fig. 3

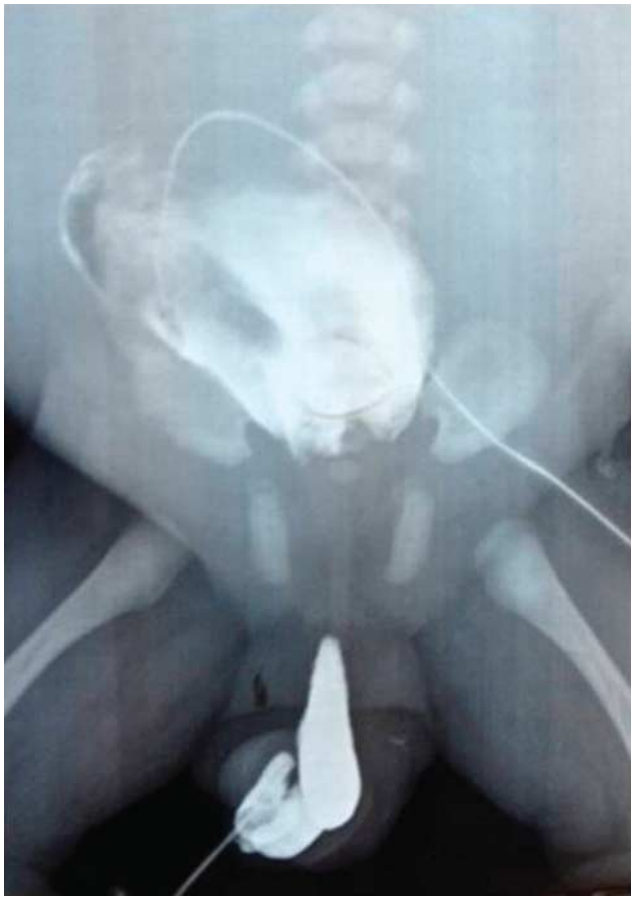


Intraoperative images.

hindgut during early intrauterine development could account for pouch colon malformation [1]. We feel that the presence of abnormally long mesentery and nonfixation of colon to posterior abdominal wall may lead to antenatal vascular events that may have led to pouch colon formation. However, it seems more plausible that both mesodermal arrest and vascular events early in embryogenesis may be responsible.

Pouch colon association with PBS can be associated with poorer prognosis than an isolated PBS. This association can have both antenatal and postnatal

Fig. 4



Dye study showing large bladder and scaphoid megalourethra.

concerns. Antenatally, the pouch colon adds to added distension of abdomen, both physically and also due to its contribution to urethral obstruction. The thick meconium in the bladder coming through the colovesical fistula or the angulations of the bladder neck due to the fistula may hinder micturition. This may add to associated oligohydramnios and lead to more severe pulmonary hypoplasia. Furthermore, pouch colon has been found to be associated always with vesicoureteric reflux, which is a cardinal part of PBS too [1]. Postnatally, after excision of pouch, these patients have high chances of developing neurogenic bladder. There can be three causes for this: first, the overdistended bladder due to outlet obstruction; second, surgical dissection posterior to the bladder to reach and ligate wide colovesical fistula; and third, the suture line of the wide colovesical fistula.

The patients of PBS may need a supravesical diversion (ureterostomies), cutaneous vesicostomy, anterior urethral reconstruction, reduction cystoplasty, ureteral reconstruction, and reconstruction of anterior abdominal wall [1,2,6,7]. In patients with pouch colon and megalourethra, another issue during surgery is regarding urinary diversion [6]. Presence of a cutaneous vesicostomy and colostomy nearby may be a cause of urinary tract infections. Thus, in this case, we did a suprapubic cystostomy, which was placed away from the stoma.

Conclusion

Thus, the association of pouch colon and megalourethra with PBS has an overall poorer prognosis. The anomaly may be a result of unique combination of vascular events and mesodermal defects early in embryogenesis. Proper preoperative assessment and explanation of poor prognosis, intraoperative surgical acumen for posterior bladder dissection, placement of vesicostomies and colostomies, and postoperative expert nursing care are major prerequisites for proper management of this rare association.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

1 Bangroo AK, Tiwari S, Khetri R, Sahni M. Congenital pouch colon with prune belly syndrome and megalourethra. *Pediatr Surg Int* 2005; **21**:474–477.
2 Yadav K. Short colon associated with the prune belly syndrome. *Int Surg* 1979; **64 (No. 4)**:83–85.
3 Raghavan M, Haripriya U, Pradeep PV. Rare association of prune belly syndrome with pouch colon. *Pediatr Health Med Ther* 2011; **2011**:9–12.
4 Short KL, Groff DB, Cook L. The concomitant presence of gastroschisis and prune belly syndrome in a twin. *J Pediatr Surg* 1985; **20**:186–187.
5 Walker J, Prokurat AI, Irving IM. Prune belly syndrome associated with exomphalos and anorectal agenesis. *J Pediatr Surg* 1987; **22**:215–217.
6 Mahajan JK Ojha S, Rao KL. Prune-belly syndrome with anorectal malformation. *Eur J Pediatr Surg* 2004; **14**:351–354.
7 Baba AA, Hussain SA, Shera AH, Patnaik R. Prune belly syndrome with pouch colon and absent dermatome. *Afr J Paediatr Surg* 2010; **7**:25–27.
8 Rao KLN, Menon P. Congenital pouch colon associated with anorectal agenesis (pouch colon syndrome). *Pediatr Surg Int* 2005; **21**:125–126.