Congenital Pouch Colon

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Contents

Introduction .............................................................................. 2
Historical Insights ................................................................. 2
Incidence ................................................................................ 3

Etiology ................................................................................ 3

Pathology ................................................................................. 4
Gross Pathology ....................................................................... 4
Histopathological Findings ....................................................... 5
Classification .......................................................................... 6

History and Physical Examination ........................................... 6

Presentation .............................................................................. 6

Diagnosis ................................................................................. 7

Management ........................................................................... 7
Preoperative Management .......................................................... 7
Operative Management ................................................................ 8
Postoperative ........................................................................... 10

Complications ......................................................................... 10

Outcomes ............................................................................... 10

Conclusion and Future Directions ............................................... 10

Cross-References .................................................................... 10

References .............................................................................. 11

Abstract

Congenital pouch colon (CPC) is a rare form of anorectal malformation, in which the entire colon is replaced by an enormously dilated pouch that communicates with a fistula to the genitourinary tract. CPC is a condition which comprises of a high form of anorectal malformation which is associated with large
variations in the size of the dilatations of the affected colonic segment. Surgical management depends on the type of CPC, and outcomes are variable depending on the length of colon affected and the anorectal muscle complex.

Keywords
Congenital pouch colon · Congenital malformation · Pull-through procedure · Protective colostomy · Coloplasty

Introduction

Congenital pouch colon (CPC) is a congenital malformation of the large bowel in which the entire large bowel or segments of varying lengths of the large bowel exhibit enormous dilatations in the form of a pouch and communicate distally through a fistula with the urogenital system. CPC is a condition which comprises of a high form of anorectal malformation which is associated with large variations in the size of the dilatations of the affected colonic segment (Fig. 1). The scarce reporting of this condition in the 1980s missed its inclusion in the Wingspread classification of anorectal malformations; however, with the increasing number of reports and detailed investigation of the condition, CPC has been recognized as a rare form of anorectal malformation and has been included in the Krickenbeck classification (Holschneider et al. 2005).

Historical Insights

Though CPC is a rare congenital malformation which is almost nonexistent outside the Indian subcontinent, this anomaly was initially recognized in England during the beginning of the twentieth century. The first description of this anomaly was documented by Spriggs from a specimen carefully observed at the London Hospital Museum in 1912 (Spriggs 1912). The specimen was described by him to have an absence of half of the large bowel and rectum, where the dilatation was presumed to be the result of a congenital occlusion of the gastrointestinal tract. The following reporting of this congenital malformation was published almost half a century later in an article from Canada in 1959. In this publication, a more accurate account was provided by Trusler et al. who described more typical characteristics of this malformation such as the pouch-like dilatation of the shortened large bowel and its association with a high form of anorectal malformation (Trusler et al. 1959). Later on in 1967, El-Shafie described this malformation in detail as a congenital shortening of the small intestines which accompanied a cystic dilatation of the colon associated with an ectopic anus (El-Shafie 1971).

![Fig. 1](image_url) Distribution of anorectal malformations in 426 patients from 1995 to 2007
CPC was first reported from India in 1972, when Singh and Pathak coined the term “short colon” after observing this condition in a series of six patients (Singh and Pathak 1972). The authors also speculated in this report on the possible embryogenesis of the malformation. Later on, in a successive report in 1977, a description of the anatomy of this malformation was published (Singh et al. 1977). An important contribution to CPC was made by Chiba et al. in 1976, who not only made the first attempts to classify this malformation but also reported on the management of this malformation with the technique of “coloplasty” (Chiba et al. 1976). The term “colonic reservoir” was coined by Gopal in 1978 in a report with this malformation with the distal end of the reservoir terminating into the female genitals through a rectovaginal fistula (Gopal 1978). Further efforts to describe this malformation as a “short colon malformation” associated with an atresia of the anus were done by Li in 1981 (Li 1981).

Narasimha et al. in 1984 proposed the term “pouch colon syndrome” for this malformation and after observing variations in the presentation of this malformation presented a classification based on the length of normal colon preceding the pouch dilatation (Narasimha Rao et al. 1984). Cloutier et al. in 1987 described this malformation as a “rectal ectasia” in a newborn with a low anorectal malformation and reported an incidence of 5% of terminal bowel ectasia in their patients with low anorectal deformities (Cloutier et al. 1987). Elaborate terms to describe this malformation by Wu et al. in 1991 were suggested with acronyms such as “association of imperforate anus with short colon” (AIASC) or “association of imperforate anus with exstrophy splanchnica” (AIAES) (Wu et al. 1990). Also in 1990, Wardhan described this entity as imperforate anus with congenital short colon (Wardhan 1990). However, Chadha et al. in 1994 coined the term “congenital pouch colon” which is the nomenclature to aptly describe this anomaly (Chadha et al. 1994).

Incidence

The largest patient series in CPC are being reported exclusively from India. Smaller series of patients are being reported from the neighboring countries in the Indian subcontinent such as Pakistan and Nepal, however, with a low incidence reported from Bangladesh (Gupta and Sharma 2006, 2007). Sporadic cases of CPC are being reported from the Middle East, Far East, Europe, and North America (Al-Salem 2008; Wester et al. 2006; Herman et al. 2000; Arestis et al. 2005). The incidence of CPC is the highest in the North-West regions of India and is estimated to be 5–18% of the total number of neonates managed for anorectal malformations. Among the Indian Tertiary Care Centers reporting on large series of CPC, Udaipur in the Western State of Rajasthan has reported the highest incidence of CPC in India accounting for 37% of the high forms of anorectal malformations (which is more than double reported in Delhi, 15.2%) (Mathur et al. 2002). CPC more frequently affects the male population with a male-female distribution of 4:1.

Etiology

The etiopathogenesis and embryology of CPC are poorly understood. The widespread use and direct contact with pesticides in agriculture-based communities have been regarded as the possible factor in the triggering of events that lead to CPC. It is also important that the effect of these factors influence the fetus after conception at a time when the hindgut is differentiating into the urinary and colonic tracts.

Various theories have been hypothesized to explain the formation of the pouch. The chronic obstruction theory proposed that the expansion of the large bowel was a result of chronic obstruction of the distal colon (Trusler et al. 1959). However, this theory has not been accepted since the dilated pouch does not return to assume normal proportions even after a colostomy placement to relieve the obstruction. Another hypothesis is the interference of hindgut growth and migration theory.
proposed by Dickinson (1967). In this hypothesis, it was proposed that the interference in the longitudinal growth of the hindgut (distal to the allantois) and failure of its migration into the pelvis following the obliteration of the inferior mesenteric artery in the early embryonic life were responsible for the formation of the short colon. The *altered hindgut stimulation theory* was hypothesized by Chatterjee (1991). This theory proposed that the cecum and the right colon normal development was stimulated by the normal developing hindgut and development alterations in the hindgut resulting from a primary disorder were responsible for the altered development of the cecum or the right colon. The *faulty rotation and fixation theory* proposed by Wu et al. hypothesized that the faulty rotation and fixation of the large bowel were responsible for the disturbances in longitudinal growth (Wu et al. 1990). The *vascular insult theory* was proposed by Chadha et al. in which degrees of vascular insult at the time of the partitioning of the cloaca by the urogenital septum were deemed responsible for the malformation and could explain its variations (Chadha et al. 1994). The *vascular insult theory* was also supported by Mathur et al. in explanation of double pouch formation in CPC (Mathur et al. 2002). At present, vascular insults best explain the formation of CPC, which is evident by the abnormal vascular supply to the pouch. Also, the overwhelming vascular support provided by the superior mesenteric artery to the entire distal bowel supports this view, since the inferior mesenteric artery has been identified only in few patients with CPC during surgery. Recently, Mathur et al. (2018) performed a whole exome sequencing of CPC samples and reported significant associations in patients with rare mutations and variants.

**Pathology**

CPC is recognized by certain pathological characteristics that are solely found to be associated with this congenital malformation (Fig. 2).

![Operative view of the exteriorized grossly dilated congenital pouch colon segment visualized from a left lower quadrant “hockey stick” incision](image)

**Gross Pathology**

(a) The presence of anorectal malformations differentiates this entity from segmental dilatation of the colon.
(b) Irrespective of the type of CPC, there is a decrease in the length of the large bowel due to the presence of the pouch.
(c) The pouch formations may differ in length and diameter and are fecal or meconium impacted at the time of surgery.
(d) The pouch wall is thick with a stiff consistency and is abruptly connected to the normal bowel without the presence of the transition zone.
(e) There is an absence of haustrations, taenia, and appendices epiploicae in the pouch colon.
(f) An abnormal vascular supply to the pouch can always be identified during surgical exploration.
(g) A fistula can be identified to the urinary tract in male neonates (colovesical fistula) and to the genitourinary tract in females (colocloacal, colovaginal, or colovestibular fistula).
(h) Appendiceal anomalies are present and vary from complete absence to the presence of double appendices.
Histological studies of resected pouch colon demonstrate extreme variations in CPC. In most patients, acute and chronic inflammation of the mucosal and submucosal layers are present with varying degrees of hemorrhage along with the presence of disorganized muscle layers in the colon wall. Hypotrophy of the muscle layers has been observed which is more predominant in the outer muscle layer of the pouch (Agarwal et al. 2005). Extreme variations in the muscle layer (both circular and longitudinal) have also been found which range from fibrosis, atrophy, to hypertrophy along with muscle disruption (Gangopadhyay et al. 2009). Investigations have also shown variations such as the presence of normal colon wall with normal ganglion cells in some patients, to a poorly developed colon wall musculature with decreased or absent ganglion cells in others (Singh et al. 1977; Narasimha Rao et al. 1984; Chadha et al. 1994; Wakhlu et al. 1996a). Interestingly, heterotrophic tissue such as gastric mucosa, small intestinal mucosa with characteristic villi, and pancreatic tissue has also been found in pouch specimens of CPC patients (Narasimha Rao et al. 1984; Agarwal et al. 2005). Recently, Udawat et al. (2017) reported histopathological and immunohistochemical findings in 49 cases of CPC which showed distinct defects in the neuromusculature of CPC. The histopathological findings included disrupted muscle fibers, an additional muscle coat, deranged SMA, myosin and desmin expression, and a reduced number of ganglion cells. Studies have also shown that pouch colon which normally fails to show spontaneous contraction under in-vitro conditions has shown to respond to acetylcholine and histamine (Tyagi 2009).
Classification

Until recently, the most widely used classification of CPC was that described by Narasimha Rao et al. (1984) which classified CPC into four types based on the length of the normal colon proximal to the colonic pouch. Saxena and Mathur (2008) classified CPC into five types based on anatomic morphology (Fig. 3).

History and Physical Examination

The presence of an anorectal malformation with gross distention of the abdomen is the hallmark of the physical examination. In males, the pouch usually terminates in a colovesical fistula just proximal to the bladder neck. In male neonates, therefore, discharge of meconium (meconurea) or stool through the urethra via the colovesical fistula is evident, and these neonates are generally referred for treatment in the immediate neonatal period. However, in female patients, meconium and fecal discharge through a cloacal, uterine, or vaginal fistula may delay referral in a stable neonate. In the majority of female cases, the colonic pouch is reported to open into either vagina or in a persistent cloaca (Chadha and Khan 2017). Girls usually have a double vagina with a wide intervaginal bridge.

Presentation

The majority of patients with CPC present in the immediate neonatal period due to the anorectal malformation. The absence of the anal canal and excessive distention of the abdominal cavity are the two characteristic signs that raise suspicion of CPC. Although in CPC the pouch ends through a fistula in the genitourinary tract, meconurea may be present or absent. Another common symptom in these neonates is serial episodes of bilious vomiting which is a major symptom that leads to the referrals. Delayed referrals or grossly distended pouch colon are associated with colonic perforations, which present a major challenge in the management of the newborn with septicemia and peritonitis which further deteriorates the respiratory distress present due to the massive abdominal distension. Delay in diagnosis and late referrals even in the neonatal period have been largely responsible for the high mortality in CPC. Awareness of the condition and development of proper management strategies especially through improvement in neonatal intensive care at the tertiary centers in India have drastically reduced the mortality from 40% to the present rate of 15% (Sharma et al. 2005).

Despite CPC being a high anorectal anomaly, major associated malformations are relatively uncommon (Chadha and Khan 2017). The most common associated malformations are urological including hydronephrosis and vesicoureteral reflux.

Fig. 4 Plain abdominal erect radiograph demonstrating the classic grossly dilated pouch colon segment occupying over 75% of the left abdomen, thereby displacing the small intestines toward the lower quadrant in the right abdomen.
Diagnosis

Plain erect abdominal radiographs performed to diagnose patients with CPC demonstrate a classical solitary grossly dilated air fluid bowel loop that occupies more than 75% of the abdominal cavity with displacement of the small intestines (Fig. 4). The position of the pouch and the displacement of the intestinal loops depend on CPC type (Wakhlu et al. 1996a). Although plain abdominal radiographs can predict the CPC type, the definitive diagnosis and the CPC type can be determined only after surgical exploration (Mathur et al. 2010). False diagnosis of CPC based on plain radiographs is possible in patients with (a) significant dilatation of the sigmoid colon, (b) pneumoperitoneum after perforation in anorectal malformations due to late presentation, and (c) in females, neonates with rectouterine fistula when severe dilatation of the meconium filled uterus and gas exhibit the classical images of CPC radiographs (Wakhlu et al. 1982).

In the majority of CPC cases, the diagnosis is made on an abdominal radiograph. An enormous gas shadow or air fluid level is seen on the left side of the abdomen, occupying more than 50% of the abdominal width and the small bowel loops are displaced to the right. A prone cross table or lateral film is better for visualizing gas within the bladder. Prior to the surgical management, further investigations such as abdominal ultrasonography, intravenous pyelography, or voiding cystourethrography and echocardiography are mandatory since a wide range of genitourinary, gastrointestinal, and other forms of associated anomalies have been found in patients with CPC. (Table 1).

Management

Preoperative Management

The preoperative management is broadly dependent on the condition of the pouch (intact versus perforated). In stable neonates, preoperative management includes gastric decompression using a nasogastric tube, intravenous fluid replacement

| Genitourinary anomalies          | Gastrointestinal anomalies        | Other organ anomalies             |
|---------------------------------|-----------------------------------|----------------------------------|
| Hydronephrosis                  | Absent appendix                   | Sacral agenesis                  |
| Vesicoureteral reflux           | Double appendix                   | Congenital heart disease         |
| Bicornuate uterus               | Malrotation                       | Myelomeningocele                 |
| Cryptorchidism                  | Colon duplication                 | Prune belly syndrome             |
| Hydroureteronephrosis           | Meckel’s diverticulum             | Hemivertebrae                    |
| Hypospadias                     | Double Meckel’s diverticulum      | Congenital talipes equinovarus   |
| Renal aplasia/agenesis          | Esophageal atresia                | Perineal teratoma                |
| Renal dysplasia                 | Small intestinal duplication      | Absent ribs                      |
| Double uterus                   | Rectal atresia                    | Down’s syndrome                  |
| Double vagina                   |                                    |                                  |
| Septate vagina                  |                                    |                                  |
| Ectopic kidney                  |                                    |                                  |
| Urethral duplication (males)    |                                    |                                  |
| Urethral diverticula            |                                    |                                  |
| Bifid penis                     |                                    |                                  |
| Megalourethra                   |                                    |                                  |
| Urethral strictures             |                                    |                                  |
| Bladder extrophy                |                                    |                                  |
| Duplicate bladder extrophy      |                                    |                                  |

List of associated anomalies reported in congenital pouch colon patients

Table 1: List of associated anomalies reported in congenital pouch colon patients distributed under genitourinary, gastrointestinal, and other organ manifestation categories
to correct the effects of dehydration and electrolyte imbalance, and placement of a urinary bladder catheter. Antibiotic therapy is commenced and extended depending on the state of the inflammation in the pouch colon evaluated during the surgery. In neonates presenting with pouch perforations along with signs of peritonitis or septicemia, aggressive intensive care management is necessary to stabilize the neonate for the emergency surgical procedure which is limited to evacuation of the meconium or stool from the peritoneal cavity, placement of an ileostomy or colostomy, and closure of the perforation site. The intention in surgical management in neonates with perforations is to perform the surgery with the smallest possible incision and to complete the procedure in a short time.

**Operative Management**

Management algorithm of CPC is based on the type of pouch according to the Saxena-Mathur classification (Fig. 5) (Mathur et al. 2009). An abdominal incision in the lower left quadrant in the shape of a “hockey stick” has been found to offer optimal access to inspect the malformation with the primary intention of fistula ligation irrespective of the CPC type. After the fistula has been exposed and ligated, the condition of the pouch dictates the further operative strategy. Staged procedures, employing the placement of a protective ileostomy or colostomy with ligation of the fistula in the first stage, followed by an abdominoperineal pull-through in the second stage, still offer the safest option when compared to one-stage surgery which is associated with a higher incidence of morbidity and even mortality. The intention of surgical management in CPC is to evaluate the amount of large bowel affected and to salvage its maximum length in order to restore or partially restitute the function of the large bowel such as absorption, transportation, and containment.

In type 1 CPC and type 2 CPC, a one-stage procedure (pouch excision and pull-through) or three-stage procedure (ileostomy, pouch coloplasty with pull-through, and ileostomy closure), depending on the condition of the pouch (ischemic or healthy), can be performed. In case
of severe ischemia and perforations, resection of the pouch remains the only alternative. If the pouch is resected, either a direct pull-through of the ileum or placing a protective ileostomy with delayed ileum pull-through offers the best surgical options. However, if the pouch is healthy, the surgical approach is focused on attempts to rescue and taper the pouch and to perform a pouch coloplasty through pouch tubularization. Pouch tubularization is performed after pouch mobilization and longitudinal incision on the antimesenteric side (to preserve the vascular supply) with edge reapproximation over a catheter. Although tubularized pouch coloplasty is performed in both type 1 CPC and type 2 CPC, it is not uncommonly associated with increased morbidity in terms of incontinence and complications resulting from redilatation of the tubularized pouch (Wakhlu et al. 1996b; Chadha et al. 2002).

The significance of the Saxena-Mathur classification in differentiating type 1 CPC and type 2 CPC is to recognize the presence of the normal cecum in type 2 CPC which is underestimated in the classifications on surgical approach. Various investigations and experimental studies have demonstrated the significance of the cecum and ascending colon or parts of the ascending colon in the absorption of sodium which influence the exchange of chloride and bicarbonate in these tissues (Hatch and Freel 1988; Fromm et al. 1990). However, investigations need to be performed to validate these differences in patients and compare type 1 CPC and type 2 CPC patients. Similarly, the absorption of potassium, which is significant in the descending colon in experimental studies (Yau and Makhlof 1975; Sweiry and Binder 1990), highlights the difference between the type 3 CPC and type 4 CPC since the normal functioning descending colon is only present in type 4 CPC.

The presence of considerable lengths of normal colon in the type 3 CPC and type 4 CPC enables total resection of the pouch and a staged abdominoperineal pull-through of normal colon. The approach to type 3 CPC and type 4 involves the ligation of the fistula, excision of the pouch, and placement of a stoma which is followed by a delayed abdominoperineal pull-through. Although it is debatable if a protective colostomy should be placed, or a prior high colostomy be closed during the pull-through procedure, the authors prefer the first option and place a protective colostomy during the pull-through which is returned return at a later point of time. In type 3 CPC and type 4 CPC, if the high colostomy is opted (instead of the end colostomy), appropriate placement of the colostomy is advocated since improper placement of a high colostomy may interfere with the pull-through procedure especially if the length of the colon to be pulled through is too short.

Management of type 5 CPC requires the approach to two pouch colon segments that are separated by a segment of normal colon and is best treated by a three-stage procedure (Mathur et al. 2002). The first procedure involves the ligation of the fistula, excision of the distal pouch, tubularization of the proximal pouch, and the placement of a protective ileostomy. The second procedure involves the abdominoperineal pull-through keeping the protective ileostomy. In the third procedure, the protective ileostomy is returned. Another option type 5 CPC would be to tubularize both the proximal and distal pouch.

Surgical management of female neonates with pouch colon is complex due to the frequent association with the cloaca as well as the associated anomalies of the genital system (Chadha et al. 1999, 2015; Sarin et al. 2007). Depending on the complexity of the genital anomalies, a one-stage or three-stage approach is preferred; however, there is no consensus on the approach to date. The pouch colon has been tubularized to create a neo-vagina and reconstruct the anorectum with preserved vasculature also using the longitudinal incision technique (Wester et al. 2006). Also, pouch colon patch graft on pulled through ileum has been reported in an isolated case with good clinical outcomes (Ratan and Ratan 2004).
**Postoperative**

The postoperative management depends on the condition of the pouch. In patients with pouch perforation, intensive care support along with parenteral nutrition is necessary until complete recovery. The use of antibiotics and the duration of treatment are also dependent on the surgical findings. In patients with protective colostomy, feeds are commenced earlier than those who have undergone tubularization and pull-through procedures. Monitoring of bowel movements is necessary to achieve success in these patients (Gharpure 2007). Regular postoperative follow-up is necessary to document the bowel movements which could range from diarrhea when the pouch is resected to obstipation which could result in tubularized pouch redilatation and necessitate further surgical procedures.

**Complications**

Complications in the management of CPC can be divided into five distinct categories which are related to (a) window colostomy, (b) protective colostomy, (c) tubularized pouch colon, (d) complete pouch resection, and (e) pull-through procedure. Window colostomy leads to a wide range of complications such as incomplete pouch decompression, prolapse, stoma recession, stoma stenosis, pouchitis, enterocolitis, and failure to thrive (Singhal and Bhatnagar 2006). The complications of protective colostomy or pull-through procedures are general complications associated with these procedures and are not specific for CPC. Tubularization of the pouch colon could be associated with complications of leakage along the suture line with consequent rupture. Also, redilatation after tubularized colon has been observed after long-term follow-up in patients with salvaged pouch colon (Chadha and Khan 2017; Chadha et al. 2002; Budhiraja et al. 1997). Complete pouch resection is associated with the complications of recurrent watery diarrhea, excoriation around the anus, perineum or genitals, and poor weight gain. Severe complications in neonates with pouch perforations are not only limited to the septicemia and peritonitis but have also been found to negatively influence the respiratory status as well as the anesthesia efforts intraoperatively and postoperatively with lethal outcomes (Ghritlaharey et al. 2007).

**Outcomes**

Although primary pull-through as a definitive treatment was proposed (Yadav 1983), the treatment of this entity is more complex as the knowledge of the types of pouch colon have become more clear. CPC is a complex form of high anorectal malformation, and the muscle complex responsible for maintenance of continence directly influences the long-term outcomes in these patients. In types of CPC where sufficient normal bowel is present (type 3 CPC and type 4 CPC), better results have been achieved in patients when the entire pouch has been resected and the normal large bowel is pulled through. The colonic pouch has histopathological abnormalities of neuromusculature and therefore not suitable for pull-through. Detailed long-term follow-up studies are required to properly assess continence in the patients.

**Conclusion and Future Directions**

Surgical management depends on the type of CPC, and outcomes are variable depending on the length of the colon affected and the anorectal muscle complex (Shinde et al. 2017). Future studies will be directed to investigate the etiology of CPC and details of colon histological characteristics which were still unknown.

**Cross-References**

- Anorectal Anomalies
- Colonic and Rectal Atresias and Disorders of Anus and Rectum
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