# 11 Congenital Pouch Colon

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Contents



## 11.1 Introduction and Definition

Congenital pouch colon (CPC) is a common condition associated with anorectal agenesis, and is seen particularly in Asia. The condition is defined as an anomaly in which all or part of the colon is replaced by a pouch-like dilatation, which communicates distally with the urogenital tract via a large fistula. In this condition, a supralevator anorectal malformation (ARM) is associated with a colonic pouch of variable size (5–15 cm in diameter). The mesentery of this pouch is short and poorly developed, the wall is very thick, the taenia coli are absent or ill defined, and haustration and the appendices epiploicae are absent. The main pouch is supplied by the branches arising from the superior mesenteric artery, which form a leash of vessels around it.

This condition is more common in the northern Indian population and neighboring nations like Pakistan and Nepal, although sporadic reports have also come from other parts of the world. With the growing awareness, this condition is being reported with increasing frequency in children born with ARM. Its management involves a diversion colostomy at birth with or without the excision of the pouch followed by a pull-through.

## 11.2 Historical Background

The first description of this anomaly was given in 1912 by Spriggs in a London Hospital Museum specimen that exhibited absence of the left half of the colon and rectum [1]. In 1959, Trusler et al. described a pouch-like dilatation of shortened colon associated with high ARM [2]. Until then no name was given to this entity. In 1953, Spencer [3] reported 53 cases: 43 cases with exstrophy of bladder and intestines were labeled typical exstrophia splanchnica, and the remaining were labeled atypical exstrophia splanchnica. In 1967, Blunt and Rich described this condition as an absence of colon and rectum [4], and in 1971 Shafie described it as cystic dilatation of the colon [5].

The first report from India came in 1972 by Singh and Pathak who, in a series of six cases, named this condition as "short colon," and attempted to discuss its embryogenesis [6]. In 1978, Gopal called it colonic reservoir in a case with rectovaginal fistula [7], and in 1981, Li [8] from China named it congenital atresia of the anus with short colon malformation. Narsimha Rao et al. [9] in 1984 suggested the name "CPC syndrome". In 1990, Wu Yuejie [10] called this condition as association of imperforate anus with short colon and suggested that cases with exstrophy of the bladder and/or intestine can be called association of imperforate anus with exstrophia splanchnica. Recent

large series have used two terms, congenital short colon or CPC to describe this condition [11,12].

The anatomy of this malformation was first described in 1977 by Singh et al. [13] and subsequently in detail by Wakhlu et al. [14] and Chadha et al. [11]. It was in 1984 that Narsimha Rao et al. proposed an anatomical classification of this condition that has been widely accepted [9]. An important advancement in the management of this condition was the technique of coloplasty, which was developed in 1976 by Chiba et al. [15]. Subsequently this has been used with good results [11,16,17].

## 11.3 Geographic Distribution

This condition is seen much more frequently in the northern, north western, and central parts of India. Most of the patients come from the states of Punjab, Uttar Pradesh, and Delhi. A few reports have originated from China, Japan, and other parts of the world. The cause of this unique geographical distribution has not yet been ascertained.

## 11.4 Incidence

The incidence of CPC varies in different parts of the world. Except from the northern part of the Indian subcontinent, there are only sporadic case reports from other parts of the world. The incidence of CPC among all cases of ARM in the northern parts of India has been reported to be between 2.5% and 9%. Chatterjee from Kolkata reported an incidence of 2% [12]. In Bangladesh, CPC forms 1.07% of all ARM cases (personal communication, Professor Tahmina Banu, Bangladesh). Similarly, in Assam (eastern India), the reported incidence is 5.05% of all and 7.93% of high anomalies (personal communication, Dr. NC Bhattacharyya, Assam, North east India). In our series of 992 cases of all types of ARM treated during the period 1993–2005, 15.3% cases were of CPC, a much higher incidence than reported elsewhere. However, it may be possible that minor varieties of the ARM cases may not have reached us at the tertiary care hospital, hence projecting a false higher incidence of CPC.

CPC is more common in males. Interestingly, the sex ratio reported by authors outside India has been almost equal (1.27:1), while in India the reported incidence has been 3–4.3: 1 [11]. In our series (152 cases, 1993–2005), it was significantly higher in males than in females (7:1; Table 11.1).

**Table 11.1** Cases of congenital pouch colon (CPC) treated between 1993 and 2005 at the All India Institute of Medical Sciences, New Delhi

| Gender | Type | Number of cases |
|---|---|---|
| Male | Colovesical | 133 |
| Female | Colocloacal | 13 |
| | Colovaginal | 6 |
| Total | | 152 |

## 11.4 Etiology and Embryogenesis

The exact embryogenesis of CPC is not known. In 1959, Trusler proposed that the dilatation was the result of chronic obstruction, but this theory was discarded as the pouch fails to decrease in size even after colostomy [2]. Another theory proposed was aborted hindgut development following obliteration of the inferior mesenteric artery early in fetal life [18]. Chatterjee proposed that the cecum and right colon develop normally from the postaxial midgut when this portion of the midgut is stimulated by normally developing hindgut [12]. Thus improper development of the postaxial midgut or presplenic gut is due to a primary disorder of the proximal end of the hindgut or postsplenic gut.

Wu Yuejie suggested that faulty rotation and fixation of the colon leads consequently to a disturbed longitudinal growth [10]. Chadha et al. proposed that varying extents of vascular insult at the time of the partitioning of the cloaca by the urorectal septum could explain the different types of the malformation [17]. Wakhlu et al. have postulated that CPC represents a stage in the development of cloacal exstrophy and is the combined effect of defective development of the splanchnic layer of the caudal fold and failure of rotation of the gut, causing defective longitudinal growth of the colon [19].

In the authors' view, the high density of cases in the northern belt of the Indian subcontinent points toward environmental factors, with deficiency of iodine or vitamin B as some of the possible factors contributing to this anomaly. In the recently conducted survey on this anomaly from various pediatric surgical centers in India, Pakistan, Bangladesh, Nepal, Sri Lanka, Italy, Sweden, and Japan, the incidence was reported to be the highest in north India (Kashmir, Chandigarh, Delhi, Lucknow, Varanasi), but decreased as we proceeded toward the east. It was uncommon in Bangladesh (1.07%); however, in Pakistan, the incidence was as high as 8% of all ARM. Only sporadic cases

have been seen from Sweden, Japan, and Italy, and were reported merely as curiosities (personal communications).

As the blood supply is always abnormal to the pouch in these patients, an early vascular insult cannot be ruled out. It is only the superior mesenteric artery that is prominent and supplies the whole distal bowel. The inferior mesenteric artery is present in only 50% of cases of distal CPC, and it is also quite insignificant. A genetic predisposition also needs to be ruled out.

The north Indian belt is also known as the stone belt (due to the deficient nutritional factors in the diet) and also for the iodine deficiency in the water there. The land is very fertile and the pesticides are used liberally in the fields. The population is mainly vegetarian and consume lot of fresh vegetables in the diet. In addition, most of the population with ARM in this region have a low socioeconomic status. All of these factors suggest environmental factors affecting or precipitating the anomaly at a window time after conception when the hindgut is developing and differentiating into urinary and intestinal tracts.

## 11.5 Classification

The term short colon should not be used to describe this ARM. The short colon is a term that should be used exclusively for the condition seen in babies born with a shortened length of left colon, which is also narrow in caliber; these babies are usually born to diabetic mothers – there is no ARM. The condition of short colon was first classified by Chiba et al. (Table 11.2) [15].

According to the current definition of CPC, type 3 of this classification can be called congenital CPC. Type 5 including the abnormal vessels forms part of the CPC. CPC also needs to be differentiated from those cases with the congenital segmental dilatation of the colon, without any ARM [20].

**Table 11.2** Types of short colon

| | |
|---|---|
| 1. | Agenesis of colon |
| 2. | Short colon without imperforate anus |
| 3. | Short colon with imperforate anus (dilated colon) |
| 4. | Short colon as a part of exstrophy of the bowel and bladder (small and narrow colon) |
| 5. | Short colon due to abnormal vessels and the like |

**Table 11.3** Types of CPC. Initially, in India, cases of types I and II were more commonly seen and accounted for more than 70% of cases until 1985. Interestingly, during the past two decades, it is types III and IV CPC that have become more common

| | |
|---|---|
| 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 cecum, 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. |

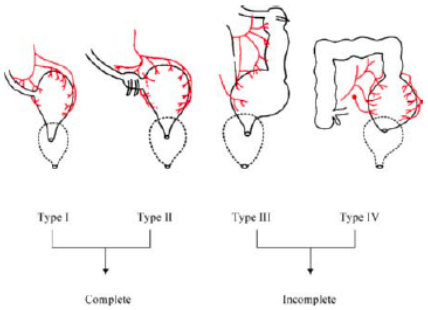


**Fig. 11.1** Modified classification of congenital pouch colon (CPC)

For all descriptive purposes, the widely accepted classification is based on the length of normal colon present proximal to the dilated pouch, as given by Narsimha Rao et al. (Table 11.3; Fig. 11.1) [15].

Wakhlu et al. [19], with their large clinical experience in this field, have simplified the classification, basing it upon the length of normal colon and the management planning in relation to need for coloplasty. They describe type A as a partial short colon, with at least 8 cm of colon proximal to the pouch. Their type B (or complete short colon) is where there is no normal colon, or a colon of less than 8 cm in length.

In the authors' view, the terms "incomplete" and "complete" CPC may be more appropriate. It is preferable that the term short colon should be avoided to avoid confusion in terminology. It is also feasible to use the remaining colon for definitive pull-through, which would be more important, rather than the 8 cm length of colon. Based on these, the authors propose a modified version of Wakhlu's classification.

1. Incomplete CPC: where the length of the normal colon is adequate for performing the pull-through, without the need for doing a coloplasty. The procedure would involve excision of the pouch with an end colostomy at birth and a definitive pull-through later. A single-stage pull-through in the newborn stage can also be undertaken if the condition of the baby permits.
2. Complete CPC: where there is either absent or insufficient normal colon left to permit a pull-through procedure. In this situation, a coloplasty procedure would be required to retain only a 15-cm length of CPC in the form of a tube, to be brought out as an end colostomy. A pull-through procedure at the time of performing coloplasty should not be preferred in the newborn stage as it is associated with high morbidity and mortality.

According to the authors, the CPC should have the following anatomical criteria:

1. There is anorectal agenesis.
2. The total length of the colon is short (Fig. 11.2).
3. The colon has a pouch with varying length; saccular or diverticular with a collection of meconium or fecal matter (Fig. 11.3).
4. The blood supply to the pouch is abnormal (Fig. 11.4).
5. The colon wall is thick and muscular with hypertrophied mucosa (Fig. 11.5).
6. The fistula with the genitourinary tract is large, muscular, and long. It is closely adherent with the bladder wall.
7. There is no transitional zone between the pouch and the normal bowel. The pattern changes suddenly and sharply.

Associated genitourinary malformations (cloacal anomalies, double vagina, exstrophy) are common in girls; however, other (nongenitourinary) associated congenital anomalies are less common.

The anatomical features vary according to the length of the colon that exhibits pouching. In complete pouching of the colon, there is a large, dilated, thick-walled pouch occupying most of the left side of abdomen. The cecum, if present almost always opens into the sac from the right side. It may be associated with an absent, rudimentary, or double appendix. The ileum opens into the cecum or the pouch from right side and there is associated malrotation. The pouch has a poorly developed mesentery and is supplied by the superior mesenteric artery on the superior and right side and an arcuate extension of the superior mesenteric artery on the left side [21]. The inferior mesenteric artery is present only in incomplete CPC and supplies the lower half of left lateral side of pouch. The pouch lacks haustrations, taeniae, and appendices epiploicae. At times the inferior mesenteric artery may be completely absent.

The distal communication of the pouch is in most instances with genitourinary system. In males, the communication is most commonly present with the bladder and the fistula opens on the posterior wall of bladder near the base. Occasionally the fistula may open higher or lower down (Fig. 11.6). The fistula is usually quite broad and thick-walled, and measures up to 1 cm in external diameter. In females, colocloacal fistula is the most commonly occurring fistula, followed by colovaginal and colovestibular fistulae.

In cases of incomplete CPC, the cecum is situated in the epigastrium or the left hypochondrium and a variable length of normal colon is found, which ends in a large sac that communicates with the bladder in males and the vagina or vestibule in females. The cecum, appendix, and the normal part of the colon are vascularized by the superior mesenteric artery. The inferior mesenteric artery, if present, supplies part of the colonic pouch.

The pelvic musculature is variable in cases of CPC, and when associated with a complete pouch or low vertebral anomalies, the pelvic and perineal muscles are poorly developed.

## 11.6 Histopathological Examination

The pouch wall consists of a normal number of ganglion cells, although a few authors have found reduced and very small ganglion cells [7,9,13,22,23]. Nerve bundle hypertrophy has also been reported, but is not the regular feature [22]. Congestion of the mucosa and focal hemorrhages are seen commonly [22,23]. In a detailed review of these cases, the authors found the following histological features in patients with CPC:

1. In most cases, the muscle coat did not have the normal differentiation of the inner circular and the outer longitudinal muscles. The muscles were also arranged in a decussating pattern. The circular muscle was incomplete in 50% of cases. The wall of the blood vessels was normal (Fig. 11.7 A).
2. The ganglion cells were mature and present in all cases, with the presence of normal or occasionally hypertrophic nerve bundles. However, giant ganglia were seen in 10% of cases (Fig. 11.7 B).

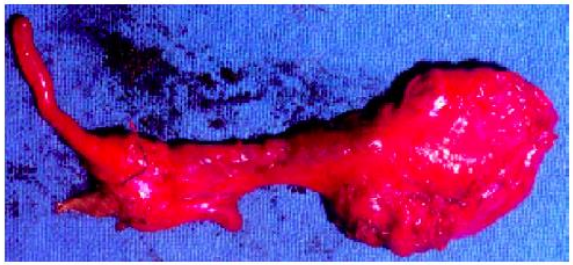

**Fig. 11.2** Complete CPC with short length of normal bowel, cecum, and appendix. The remaining normal colon is not sufficient for pull-through procedure

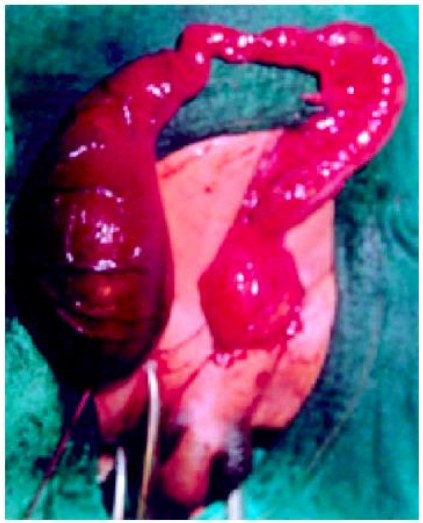

**Fig. 11.3** Incomplete CPC with dilated lower end of colon and anorectal malformation. The descending colon is normal and can easily be used for performing a pull-through procedure

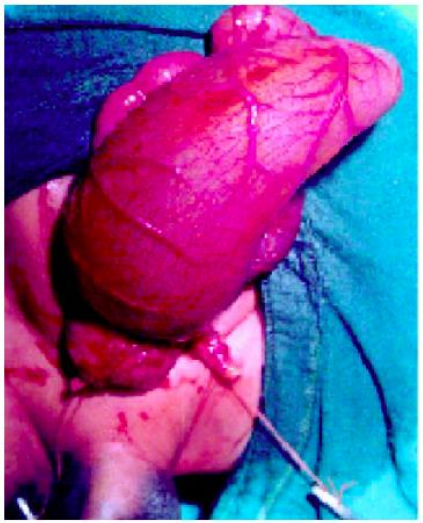

**Fig. 11.4** CPC showing abnormal blood vessels

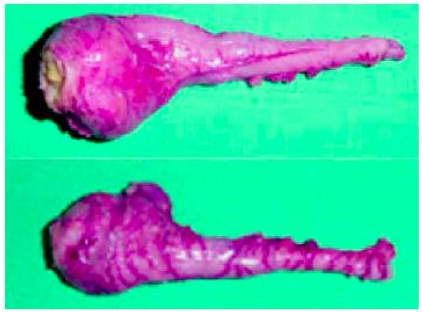

**Fig. 11.5** A, B Excised thick-walled CPC with window colostomy

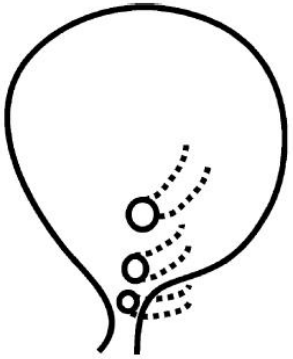

**Fig. 11.6** Diagrammatic representation of colovesical fistula sites in the bladder in congenital CPC

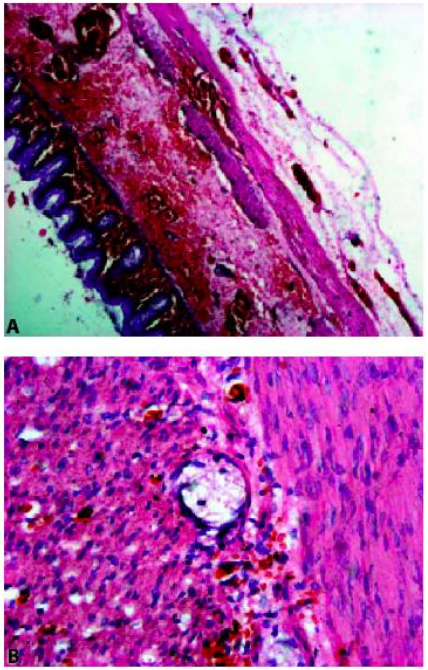


**Fig. 11.7** A Photomicrograph showing flattened mucosa, widened submucosa, and discontinuation of the circular muscle coat (magnification ×10). B Photomicrograph showing a giant ganglion cell between the longitudinal and circular muscle coats (magnification ×40)

The most characteristic finding was disorganization of the muscle coat in an arborizing manner [22]. This is possibly responsible for the absence of normal peristaltic activity in these cases, requiring the removal of the dilated pouch and retaining only the normal bowel.

## 11.7 Clinical Presentation

The majority of the patients present in the early neonatal period within the first 7 days of life. Occasionally, if the fistula is large, especially in a female child with colocloacal fistula, the presentation may be late, as the child remains decompressed. Classically, all babies present with an absent anal opening. The male baby presents early with anorectal agenesis and gross abdominal distension with or without passage of gas or meconium per urethra. The association of bilious vomiting with early gross distension of the abdomen in a case of ARM is strongly suggestive of congenital CPC.

In females, the colon is often associated with a cloacal anomaly. The female baby presents with passage of meconium from an abnormal opening, absent anus, and abdominal distension, and on examination a cloaca is usually found. Although reported by others, in this author's series, there was no case of colouterine or colovestibular fistula. There may be a double or septate vagina, and the fistulous communication may open in one of the hemivaginae or between the two into the cloaca (Table. 11.4).

**Table 11.4** Associated anomalies reported in literature ($N = 470$) [21–27]

| | Anomalies | Number of cases |
|---|---|---|
| I. | **Genitourinary System** | |
| | Posturethral diverticulum | 2 |
| | Hydronephrosis | 40 |
| | Hydroureteronephrosis | 16 |
| | Vesicoureteral reflux | 32 |
| | Renal aplasia and dysplasia | 15 |
| | Renal ectopia | 1 |
| | Pseudo exstrophy bladder | 2 |
| | Bicornuate uterus | 29 |
| | Hypospadias | 15 |
| | Cryptorchidism | 18 |
| | Duplication of the male urethra | 1 |
| | Stricture urethra | 1 |
| | Bifid penis | 1 |
| | Double uterus/vagina | 12 |
| | Septate vagina | 6 |
| II: | **Gastrointestinal System** | |
| | Double appendix | 34 |
| | Absent appendix | 25 |
| | Malrotation | 13 |
| | Duplication of the gut | 5 |
| | Duplication of the colon | 2 |
| | Double CPC | 1 |
| | Meckel's diverticulum | 11 |
| | Esophageal atresia | 7 |
| III: | **Other Anomalies** | |
| | Sacral agenesis, other vertebral anomalies | 16 |
| | Meningomyelocele | 1 |
| | Congenital heart disease | 19 |
| | Prune belly syndrome | 5 |
| | Congenital talipes equinovarus | 5 |
| | Hemivertebrae | 4 |

In cases of colonic perforation occurring early in patients with CPC, the baby may present with septicemia, gross abdominal distension with prominent veins, fluid and electrolyte imbalance, and features of peritonitis.

When the fistulous connection with the urogenital tract is large, the child may present as late as a few months after birth. Usually at that time these children are constipated and passing feces from an abnormal opening.

Sometimes a child may present with a colostomy performed by a surgeon who was unaware of this condition. Usually in these cases the presentation is with complications of colostomy, like stenosis or prolapse. The diagnosis may be apparent in a child with prolapse, but in a stenosed colostomy the diagnosis can be made only on performing a contrast radiography and occasionally only while doing the definitive operation.

## 11.8 Investigations

An plain erect x-ray of the abdomen in anteroposterior and lateral views in addition to an invertogram forms the mainstay of diagnosis. A large loop of bowel with single air fluid level occupying more than half of the total width of abdomen and displacing the small bowel to one side (usually right) is the classical picture (Fig. 11.8). The pouch is proximal to the pubococcygeal line in the invertogram. The majority of the patients can be diagnosed by an erect x-ray in addition to the conventional invertogram that is usually performed for ARM investigations.

The diagnosis may be missed when there is an incomplete pouch. A false diagnosis can be made when there is significant dilatation of sigmoid colon or localized pneumoperitoneum following perforation in patients with ARM presenting late, or in female babies with rectouterine fistula where the massive dilatation of uterus with meconium and gas may mimic CPC [14].

In cases of perforation of the pouch, the pneumoperitoneum may mask the diagnosis of CPC. An early perforation in cases of high ARM is suggestive of pouch colon, especially if the baby comes from an area where CPC is commonly seen.

A detailed work up of the baby at the time of definitive surgery should include ultrasound of the abdomen, intravenous urogram, and voiding cystourethrography and echocardiography to evaluate for associated anomalies. Spiral computed tomography with three-dimensional reconstruction of the pelvic musculature, or magnetic resonance imaging of the pelvis are optional for studying the pelvic musculature.

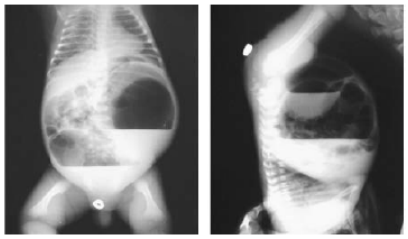

**Fig. 11.8** Invertogram: anterior–posterior (*left*) and lateral (*right*), showing large air-fluid levels, which is suggestive of CPC

## 11.9 Associated Anomalies

A large number of associated anomalies are found with CPC. The genitourinary system is most commonly involved, followed by the gastrointestinal system and others. Table 11.4 summarizes the associated anomalies found with congenital CPC [21–27].

## 11.10 Management

Preoperative resuscitation is essential with a wide bore nasogastric tube to decompress the abdominal distention, correction of dehydration and electrolyte imbalance, maintenance of body temperature, antibiotic coverage, vitamin K injection, and a urethral catheter to measure urine output as well as to decompress the bladder.

### 11.10.1 Aims of Surgical Management

The aim of surgery is to utilize the available length of colon for absorption and storage capacity as well as for propelling fecal matter onward with a continent anal opening. In incomplete CPC, an adequate length of normal colon is present so the pouch can be excised and colonic function is preserved. In complete pouching, these objectives can be achieved only by tubularizing the pouch in the form of coloplasty (Fig. 11.9). However, more complications are anticipated with the preservation and use of CPC as a tube.

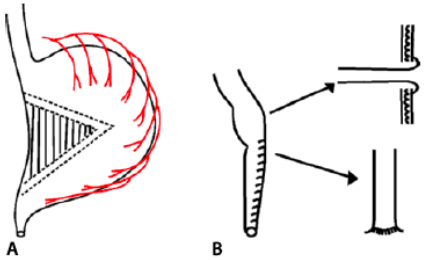


**Fig. 11.9** A Coloplasty procedure to lengthen the colon while preserving the vascular arcade. B The coloplasty may be brought out as an end colostomy on the abdominal wall for a staged procedure, or pulled through to the proposed anal site as a definitive procedure

A tube length of about 15 cm is just enough to serve the purpose of colon and to avoid the complications of a long and nonfunctional bowel.

### 11.10.2 Single-Stage versus Staged Surgery

At present, single-stage surgery for CPC is not advocated as there is an unacceptably high mortality associated with it. Although there are certain advantages of single-stage surgery, they are not sufficient to warrant a major surgical undertaking in a neonate with associated anomalies and complications.

### 11.10.3 Staged Surgery

There are two or more steps in the staged procedure depending upon the choice of procedure, which in turn depends upon the condition of the baby at presentation, the technical skill of the surgeon, and the availability of facilities for major neonatal surgery and postoperative care (Fig. 11.10). Proximal diversion may take place in the form of:

1. End colostomy after division of the fistula and excision of the pouch in incomplete CPC (preferred approach).
2. End colostomy after division of the fistula and coloplasty in complete CPC.
3. Window colostomy, in which an opening is made on the anterior surface of the colonic pouch without attempting to ligate the fistulous connection.
4. Proximal ileostomy in complete CPC
5. Transverse colostomy in incomplete CPC

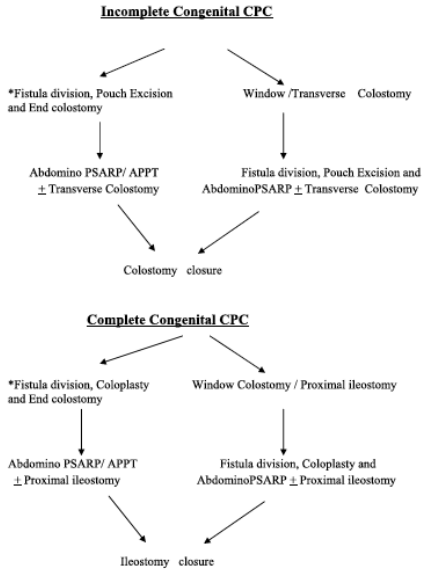


**Fig. 11.10** Algorithm for the management of congenital CPC. *APPT* Abdominoperineal pull-through, *PSARP* posterior sagittal anorectoplasty. **Preferred approach*

Excision of the pouch with an end colostomy is the procedure of choice. Window colostomy is simple surgery, can be performed with minimum anesthesia time in a sick neonate, and provides adequate decompression and a time period to allow for weight gain and fitness for the second stage. However, it has certain disadvantages, as will be described later. The mortality following window colostomy is reported to be in the range of 15–20% [21]. However, the mortality following coloplasty with end colostomy in the newborn period is higher, hence this is better done at a later date.

### 11.10.4 Definitive Procedure

In incomplete CPC with a colostomy already performed, the operative steps include dismantling the colostomy and an abdomino-posterior sagittal anorectoplasty (PSARP) for creation of a new anus. A proximal diversion with an ileostomy may be considered in selected cases to protect the neoanus.

In complete CPC without the previous coloplasty, the operation involves ligation of fistula, coloplasty, and abdomino-PSARP. A proximal ileostomy is preferred and would need to be closed in next stage.

Appendectomy should be performed at the time of pull-through to prevent misdiagnosis in the event of appendicitis occurring at a later date.

The coloplasty is performed after mobilizing the pouch completely by division of the inferior mesenteric artery (if present) and incising the pouch on the antimesenteric border, thus preserving the vascularity. The tube is fashioned over a red rubber catheter to obtain a uniform diameter (Fig. 11.9). Variable results have been reported by different authors [12,14,15,28–30]. The author's institutional experience is presented in Table 11.5.

**Table 11.5** Various types of CPC and the surgical procedures carried out at the All India Institute of Medical Sciences, New Delhi

| Type of CPC | Surgical procedure | *N* |
|---|---|---|
| **First stage** | | |
| Incomplete CPC 106 cases (69.7%) | Excision of pouch and end colostomy | 84 |
| | Pouch excision, end colostomy and abdominoperineal pull-through | 12 |
| | Referred from outside with | |
| | -window colostomy | 3 |
| | -transverse colostomy | 7 |
| Complete CPC 46 cases (30.3%) | -Excision of pouch and end colostomy/ileostomy | 4 |
| | -coloplasty and end colostomy | 36 |
| | -coloplasty, end colostomy with proximal ileostomy | 6 |
| **Second stage** | | |
| | Abdominal pull-through | 61 |
| | Abdominoperineal pull-through with proximal colostomy/ileostomy | 49 |
| **Third stage** | | |
| | Colostomy/ileostomy closure | 46 |
| **Outcome** | | |
| | Completed surgery | 107 |
| | Mortality | 11 |
| | Awaiting second or third stage | 8 |
| | Dilatation of pouch requiring pouch excision and redo pull-though | 3 |
| | Lost to follow-up | 26 |
| | Abdominoperineal pull-through | 61 |
| | Abdominoperineal pull-through with proximal colostomy/ileostomy | 49 |
| **Third stage** | | |
| | Colostomy/ileostomy closure | 46 |
| **Outcome** | | |
| | Completed surgery | 107 |
| | Mortality | 11 |
| | Awaiting second or third stage | 8 |

## 11.11 Complications

### 11.11.1 Window Colostomy

Complications related to window colostomy include recurrent urinary tract infections due to persistent colourinary fistula, associated vesicoureteric reflux, incomplete decompression of pouch through the window colostomy requiring regular washouts, massive prolapse requiring revision, recession and stenosis requiring dilatation, pouchitis (inflammation in the pouch), enterocolitis, adhesive obstruction and septicemia.

### 11.11.2 Colostomy

Complications related to colostomy include anemia, excoriation of skin, diarrhea, poor weight gain, prolapse, and stenosis.

### 11.11.3 Coloplasty

Complications related to coloplasty include suture line leak (has become negligible since the introduction of the proximal ileostomy) and wound dehiscence (minor wound dehiscence occurs in 4–5% of patients, but full-thickness major dehiscence is uncommon and is usually associated with leak from the coloplasty). Mortality following coloplasty has been reduced to less than 5% since being performed as a staged procedure.

### 11.11.4 Pull-Through

Complications related to pull-through are mucosal prolapse (easily managed by excision), anal stenosis due to noncompliance with dilatation, and colonic dilatation. The latter sometimes occurs following coloplasty in long-term follow-up [31]. This may be due to the fact that the colonic pouch is abnormal histologically and has a tendency to dilate. The utilization of a shorter segment of the pouch for tubularization is recommended.

### 11.11.5 Short Colon Length

Complications related to short length of colon are recurrent, watery diarrhea and poor weight gain.

## 11.12 Follow-Up

The follow-up examination is performed initially after 15 days and then after 1 month. The patient is subsequently called every 3 months for 1 year and every 6 months thereafter. Anal dilatation is started 3 weeks after surgery and continued as required. Initially the baby passes frequent loose stools, but subsequently the frequency of defecation decreases and the consistency becomes semisolid to solid. The colon on follow-up examination exhibits a normal caliber in most cases; however, dilatation of the tube coloplasty is a serious problem, though rare.

## 11.13 Prognosis

The prognosis depends upon the weight of the child, age at presentation, presence of sepsis and perforation, associated congenital anomalies, and most importantly on the length of colon that has pouching. The prognosis is better in cases of incomplete CPC as cases of complete CPC suffer from recurrent watery diarrhea due to the short length of the large bowel. Window colostomy performed in the pouch also does not allow complete evacuation of the contents and is frequently associated with massive prolapse, bleeding, and recurrent urinary tract infection.

## 11.14 Overall Results

In the authors' experience, as the anatomy and the histology of the CPC is abnormal, even the tube made from the dilated pouch does not work well; it does not contribute effectively to colonic motility. Rather, the postoperative complications like mucosal prolapse, incontinence, mucus discharge, skin excoriation, and colonic ectasia are more common than in those with ARM. Window colostomy is associated with serious complications and is thus not favored. Wherever feasible, an excision of the pouch in toto with an end colostomy (using normal colon) is the preferred procedure. An attempt should be made to excise the pouch even in cases with colonic perforation.

The overall mortality of CPC was previously as high as 30–40%, but has now come down to 10–20% as a result of the growing awareness of this condition and improvements in surgical management and neonatal care. Prognosis depends on the aforementioned factors, with the most important factor being the extent of the malformation. Excision of the pouch and

end enterostomy has been associated with maximal survival (92.3%) in good-risk patients [32]. Babies with incomplete CPC fair well with normal continence, physical, motor, and behavioral development. Cases of complete CPC suffer from increased frequency of stools for the initial 3–6 months, although the frequency decreases with the growth of the child and dietary modifications.

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