SMGroup

Congenital Pouch Colon

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Published Date: February 15, 2015

ABSTRACT

Congenital pouch colon (CPC) is a congenital anomaly associated with anorectal malformations (ARM) in which whole or part of the colon is replaced by a pouch like dilatation which communicates distally to the urogenital tract with a fistula. This anomaly is also referred as *congenital short colon* and *pouch colon syndrome*. Narsimha Rao, et al in 1984 classified the CPC into four types, and Wakhlu, et al in 1996 simplified the classification of CPC as partial short colon, and complete short colon. CPC, which was not mentioned in the Wingspread classification of ARM in 1984, has now been accepted and included in the new International classification (Krickenbeck) of ARM in 2005, as a rare anomalies, (being only regional). CPC is seen much more frequently in Northern, North Western, and Central part of India, though there have been case reports from other part of the India and rest of the World. The incidence of CPC among all the cases of ARM has been reported to occur in 2-18% and among the high ARM is 10-26%. CPC is more common in males, with sex ratio of 3 to 4.3: 1. Male newborns present as an absent anal opening with distension of the abdomen, and 60-75% of them also present with history of meconium in urine. In females, they present as an absent anal opening, with history of passage of meconium / stool through an abnormal opening. Female child may not always present during neonatal period due to the deflation of the bowel / pouch colon through fistulae. A large loop of bowel with single air fluid level occupying more than half of the total width of the abdomen on the plain abdominal x-ray either erects or inverted, is almost diagnostic of CPC, and it is possible to diagnose CPC pre-operatively in more

than 3/4th of the cases. Prior to definitive procedure, USG of abdomen and distal cologram is a must and other investigations like, MCU, IVP, CT scan of pelvic, also needed depending upon the need. The standard procedure for the management of the CPC cases is three-staged procedures, although it is also possible to manage by single-stage or two-staged procedures. The cases of incomplete / partial pouch colon (type III and type IV) are best managed by the excision of the pouch and pull-through of the proximal normal colon. The cases of complete pouch colon (type I and type II) are best managed by tabularizing / coloplasty of the remaining colon, as in these cases an adequate length of colon is absent. Mortality following initial surgery for fecal diversion is quite high due to various reasons.

Keywords: Anorectal malformations; Congenital malformations; Congenital pouch colon; Pouch colon syndrome; Congenital short colon; Staged-procedures; India

DEFINITION

Congenital pouch colon (CPC) is a congenital anomaly in which whole or part of the colon is replaced by a pouch like dilatation that communicates distally with the urogenital tract by means of fistula and is uniformly associated with anorectal malformations (ARM). This anomaly is also described as "congenital short colon" and "pouch colon syndrome". Most commonly used terminology for CPC are, congenital pouch colon, and congenital short colon [1-6].

HISTORICAL ASPECT

CPC-like condition was first described in 1912 by Spriggs in a London hospital museum specimen with absence of left half of the colon and rectum, but it was not named at that time [7]. Trusler, et al in 1959 from Canada described a pouch like dilation of shortened colon associated with high ARM, but that time also no name was given [8]. Singh and Pathak from India in 1972 first reported six cases associated with imperforate anus and named this anomaly as 'Short Colon' and attempted to discuss its embryogenesis [9]. Chiba, et al in 1976 proposed coloplasty for CPC [10]. Wu, et al from Henan, China in 1990 reported eight cases of imperforate anus with short colon and suggested the name as "association of imperforate anus with short colon [11]. Narsimha Rao, et al in 1984 suggested the name 'Pouch Colon Syndrome' and based on the length of the normal colon present; they also proposed a widely accepted anatomical classification of this condition [2]. Wakhlu, et al in 1996 simplified the classification of CPC as "partial short colon" and "complete short colon" and also advocated that a staged management (three - staged) for complete CPC provides considerable better result [4]. Gangopadhyay, et al in 2005 suggested single-stage management for all types of pouch colon in newborns[12]. Gupta, et al in 2005 classified the CPC into two as complete CPC and incomplete CPC [13]. Saxena, et al in 2008 proposed a classification of CPC based on anatomic morphology [14]. Wakhlu, et al in 2009 published their experience with long term results of coloplasty for CPC [15]. Ghritlaharey, et al in 2013 published their experience with two-staged management for all types of CPC [16].

GEOGRAPHICAL DISTRIBUTION AND ETIOLOGY

CPC is seen much more frequently in Northern, North Western, and Central part of India. Almost all the series on CPC are reported from India; Chandigarh, Delhi, Lucknow, Varanasi, Rohtak, Udaipur, Jaipur, Bhopal, and Srinagar [1,3-6,12-19]. A series of 17 cases of CPC has been reported in 2007 from Aurangabad, Maharashtra, India [20]. A series of 7 cases of CPC has been reported in 2009 from Kingdom of Saudi Arabia [21].Sporadic case reports of CPC have also been reported from other part of India and the rest of the World including; Hungary, USA, UK, Sweden, Turkey, Kingdom of Saudi Arabia, Nepal, France, Pakistan [22-34].

The causes and this unique geographical distribution of the CPC cases are not known. Some of the authors suspected environmental factors with deficiency of iodine and vitamin B complex as one of the causative factors for CPC. Use of pesticides by farmers is also suspected as one of the factors for this unique geographical distribution. As most of the cases of ARM are occurring in low socio-economic families and in rural area and this also support that above factors as causative for the occurrence of CPC cases in this geographical area. Although; Trusler, Wu YJ, Chatterjee, Gangopadhyay, Chadha, and Wakhluproposed different theories for the embryogenesis of CPC, but none is fit to explain all the details of CPC [3,5,8,11,13,14,19,35-37].

CLASSIFICATION OF CPC

Chiba, et al in 1976 first classified the short colon into five types (Table I) [35]. Narsimha Rao, et al in 1984 classified CPC based on the length of the presence of normal colon proximal to the dilated pouch, and they classified it into four types. This is a most widely accepted classification for CPC (Table 2) [2,35,36]. Wakhlu, et al in 1996 simplified the classification of CPC based on the length of normal colon in relation to the need for coloplasty, and they classified CPC into two types (Table 3) [4,35,36]. Gupta, et al in 2005 classified CPC based on the absence or presence of adequate length of normal colon for pull-through procedures and they classified CPC into two types as complete congenital pouch colon and incomplete congenital pouch colon (Table 4) [13,37]. Saxena - Mathur in 2008 classified CPC based on anatomic morphology into five types (Table 5) [14,38]. CPC was not mentioned in the Wingspread (USA) classification of ARM in 1984, has now been included in the new International classification of ARM 2005 (Krickenbeck-Germany classification), as rare anomalies, (being only regional) [39,40].

Table 1: Classification of short colon[10].

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 extrophy of bowel and bladder (small and narrow colon).
5.	Short colon due to abnormal vessels and the like.

Table 2: Narsimha Rao, et al (1984) Classification of Congenital Pouch Colon:(Based on the length of colon present proximal to pouch).

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.

Table 3: Classification of Congenital Pouch Colon:(Based on presence or absence of colon for coloplasty)[4].

Type A: Partial short colon: Length of normal colon proximal to pouch > 8 cm. **Type B: Complete short colon**: Either absent normal colon or length of colon proximal to pouch < 8 cm.

Table 4: Classification of Congenital Pouch Colon:

(Based on presence or absence of adequate length of colon for pull-through)[39].

1.	**Complete congenital pouch colon**: Either no or little normal colon left and is not enough for performing the pull-through, and needed coloplasty for preserving the colon.
2.	**Incomplete congenital pouch colon**: The length of normal colon is adequate enough for performing the pull-through, without the need for doing coloplasty.

Table 5: Classification of Congenital Pouch Colon:

(Based on anatomic morphology) [14].

Type 1: Normal colon is absent, and ileum opens into pouch colon. Type 2: Ileum opens into a normal cecum that opens into pouch colon. Type 3: Normal ascending colon and transverse colon opens into pouch colon. Type 4: Normal colon with rectosigmoid pouch. Type 5: Double pouch colon with short normal interposition colon segment.

ANATOMY OF CPC

Gupta, et al suggested that CPC should have the following anatomical criteria: anorectal agenesis with shortened colon, formation of varying length of pouch (saccular or diverticular), presence of abnormal blood supply to the pouch, presence of long and large genitourinary fistula, absence of transitional zone between pouch colon and normal bowel, with thickened wall of pouch colon and presence of mucosal hypertrophy [37].

INCIDENCE OF CPC

Incidence of CPC amongst reported series were 2 to 18% among all the cases of ARM, and 10 to 28% among the high ARM, and almost all the series reported male preponderance as 2.1 to 11.7:1 [3,12,13,16,18,21,36,37]. A high incidence (60.2%) of CPC with high ARM has been reported from Udaipur Rajasthan, India [14,38].

ANOMALIES ASSOCIATED WITH CPC

Associated anomalies with CPC are reported to occur in up to 40% of cases. Genitourinary system is most commonly involved followed by gastrointestinal and others systems [12,13,15,18-20,35,37,38,41-46].

Genitourinary System

The most commonly associated anomalies are; hydronephrosis, hydroureter,renal aplasia/ dysplasia, renal ectopia, vesicoureteral reflux. Almost all the male child with CPC have colovesical fistula, very rarely may have colo-ureteral fistula.Male child may also have hypospadias, undescended testis, bifid scrotum, bifid penis, bladder extrophy. Many of the cases of CPC in girl child have fistulae and are;colo-cloacal fistula, colo-vestibular fistula, colo-vesical fistula, colo-vaginal fistula and colo-uterine fistula. Other than above girl child may also have associated double vagina /uterus, vaginal septum,bicorniate uterus, etc[3,13,15,18-20,26,28,34,37,38,41-44].

Gastrointestinal System

The commonly associated anomalies with CPC are; double appendix, absent appendix, Ladd's band, hind gut duplication, double caecum, Meckel's diverticulum, esophageal atresia, rectal atresia, double Meckel'sdiverticulae, exomphalos, etc[13,15,18,21,24,37,45].

Other System

Ventricular and atrial septal defects, absent radius, hemi-vertebrae, cleft palate, sacral agenesis, myelomenigocele, Prune belly syndrome, etc[13,15,19,21,37,46].

CLINICAL PRESENTATION

All the males present with absent of anal opening with gross distension of the abdomen. As there is colovesical fistula, and therefore meconium may be there in urine. Fifty to 75% of thenewborns male with CPC also have history of passage of meconium in urine. Females present with absent anal opening and passage of meconium / stoolthrough abnormal opening. All the female cases of CPC may not present at neonatal period. Depending upon the diameter of the fistulae, the bowel / pouch deflation may occur and may present late in childhood. Female child with CPC may have a fistula as either of one; colo-cloacal, colo-vesical, colo-vaginal, colo-vestibular, colo-uterine, and the diagnosis of CPC is not possible on clinical examination only. Clinically it is difficult to suspect the case of ARM as a CPC, and always need radiological investigations for documentation and diagnosis. Rarely pouch colon perforation is there and newborn babies may present as ARM with perforation peritonitis with features of septicemiain poor general condition, and the diagnosis of pouch colon is being made during surgery. In some of the cases of CPC, stoma is created by general surgeon / surgeons not aware of CPC, and referred to higher centers for the management of complications of stoma or for definitive procedures for ARM. In these cases the diagnosed of CPC is made during distal cologram or during abdominal exploration for definitive procedures for ARM. Some of the cases of CPC clinically may also present with symptoms related to the associated anomalies along with symptom related to ARM[4-6,12,13,15,18-20,35-38,41].

INVESTIGATION AND DIAGNOSIS OF CPC

A large loop of bowel with single air fluid level occupying more than half of the total width of

the abdomen on the plain abdominal x-ray, either inverted or erect is almost diagnostic of CPC (Figure 1a and Figure 1b).The pouch is proximal to the pubococygeal line in the invertogram. Pre-operatively, it is possible to diagnose pouch colon with the help of radiological investigations in about 75 to 85% of the cases. As colovesical fistula (CVF) is always there in male child with CPC, and in theses newborns, air in the urinary bladder may be detected in invertogram or plain x-ray of abdomen, along with a large air fluid level, suggestive of CPC with colovesical fistula (Figure 1c and Figure 1d). In cases of pouch colon perforation it is not possible to suspect CPC, either clinically or on radiological investigations, and the diagnosis of CPC is confirmed during surgery. Rarely, a case of ARM with history of meconium in urine investigated (by refereeing physician) by plain x-ray abdominal and micturitingcystourethrogram (MCU) and referred to higher centres for the management of ARM (Figure 1e). Female child needs varieties of investigations to for the confirmation of the CPC and fistulae [2-6,12-14,18-20,35-38,47].

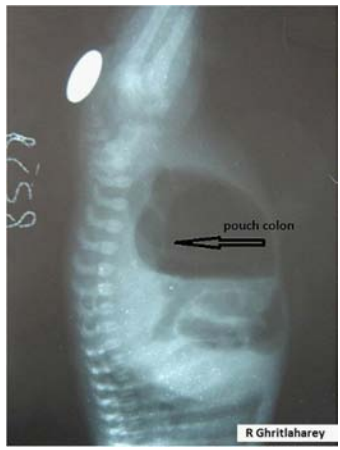

Figure 1a: Invertogram showing a large loop of bowel, suggestive of CPC.

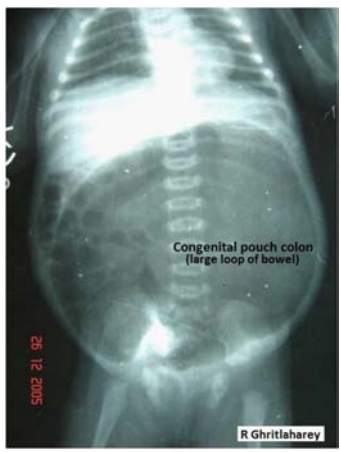

Figure 1b: Plain x-ray of abdomen showing a large loop of bowel, suggestive of CPC.

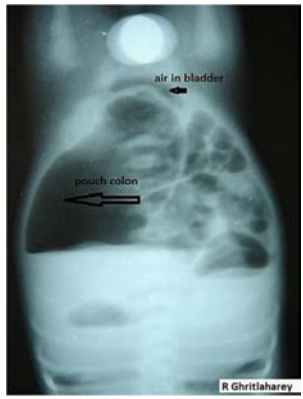

Figure 1c: Invertogram showing a large loop of bowel & air in urinary bladder, suggestive of CPC with CVF.

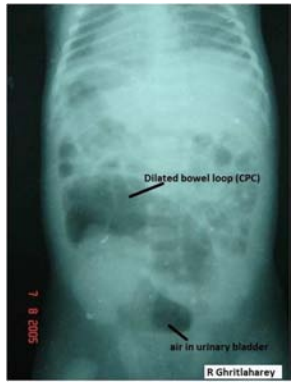

Figure 1d: Plain x-ray of abdomen showing a large loop of bowel & air in urinary bladder, suggestive of CPC with CVF.

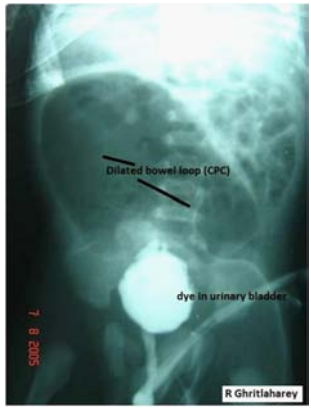

Figure 1e: MCU of a newborn showing dye in urinary bladder and a large loop of bowel, suggestive of CPC.

Prior to the definitive procedures (staged-procedures) detail investigations are required and are; USG of the abdomen, distal cologram, intravenous urogram, MCU, CT scan of pelvis, etc, and all varies from case to case and center to center. Distal colostogram is a must prior to definitive procedure, and it provides details of colon anatomy, and fistula may be detected, although it is not always possible to document the fistulous communication in all the cases (Figure 2a, Figure 2b, and Figure 2c). The pre-operative preparation prior to the definitive procedure is usually with normal saline washes twice daily for three days prior to operation and the post-operative management is as standard protocol as done for any exploratory laparotomy; ie, intra venous fluids, antibiotics, pain killer, naso-gastric aspiration, etc[4,5,13,15,16,18,35-38,48].

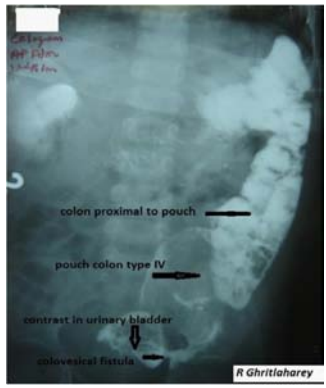

Figure 2a: Distal cologram showing CPC type IV, colon proximal to pouch, dye in urinary bladder and CVF.

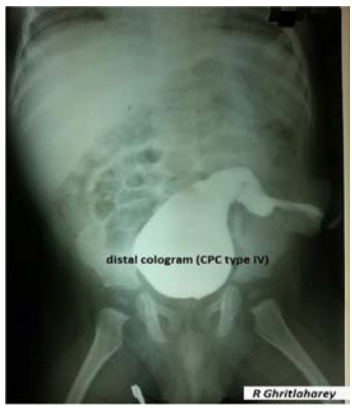

Figure 2b: Distal cologram showing CPC type IV, dye not seen in urinary bladder, and CVF not documented.

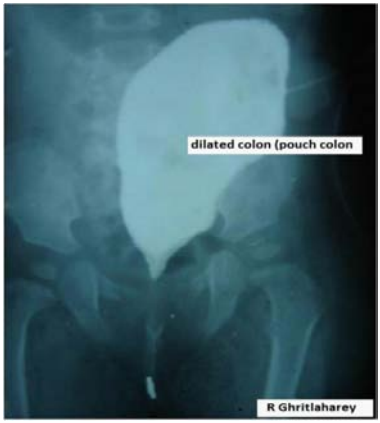

Figure 2c: Distal cologram showing dilated pouch colon, and spillage of dye in perineum in female child.

MANAGEMENT

The standard procedure for the management of CPC is a three-staged procedure, and this is the most preferred approach and also advocated by majority of the authors. The basic objectives of the surgical procedures are to utilize the available length of the colon for absorption and storage capacity, as well as for the capability to propel the faecal matter onwards with a continent anal opening. In the cases with incomplete / partial pouch colon (type III and type IV), as an adequate length of normal colon is present proximal to pouch, so that the pouch can be excised during definitive procedures while the colonic function is still preserved. In the cases with complete pouch colon (type I and II), as the adequate length of normal colon is absent, so that the objectives of preserving colon functions can be achieved only by tabularizing (coloplasty / colorrhaphy) the remaining colon / pouch [3-6,13,15,18,20,35-38,49]. CPC can also be managed by single-stage procedures or by two-staged procedures [12,16,38,49]. Laparoscopic assisted anorectal pull-through for the treatment of incomplete CPC (type III and IV) has also been reported in literature with encouraging results [33,51].The choice of surgical procedures largely depends upon the choice of the surgeons, condition of the baby at initial presentation, technical skill and expertise of the operating surgeons and the facilities available at the particular center.

THREE-STAGED PROCEDURES FOR CPC

Three-staged procedure for CPC is a standard procedure [3,4,13,15,18,20,35-38,49]. It consists of the following stages;

A. First stage: This is an initial operative procedure and includes either of one (a) proximal transverse colostomy (Figure 3a), colostomy few cm proximal to pouch (Figure 3b and Figure 3c),window colostomyor ileostomy (Figure 3d) with or without fistula division, (b) division of fistula and end colostomy, excision of pouch and end colostomy, and (c) division of fistula, coloplasty and end colostomy. Rarely during colostomy, it is not possible to deliver out the colon

and may need to deflate the pouch, and this puncture site may be used for creation of a window colostomy (Figure 3e). Most commonly colostomies are being preferred for incomplete pouch colon (type III and type IV CPC) (Figure 4a and Figure 4b) and an ileostomy is usually being done in the cases of complete pouch colon (type I and type II CPC)(Figure 4c).

B. Second stage: This stage consist of definitive operative procedure, and it includes either of one (a) pouch excision and abdomino-perineal pull-through (APPT) of colon(b) pouch excision and abdomino-posterio sagittal anorectoplasty (PSARP), (c) division of fistula, coloplasty and APPT of coloplasty colon with proximal ileostomy, (d) division of fistula, coloplasty and sacro-abdomino-perineal pull-through, (e) division of fistula, coloplasty and PSARP, and (f) pouch excision and APPT of ileum

C. Third stage: This stage includes closure of colostomy / ileostomy.

Figure 3a: Transverse colostomy for incomplete pouch colon.

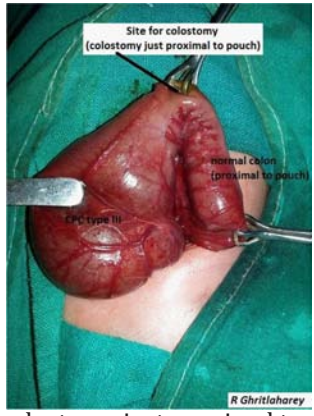

Figure 3b: Colostomy site for colostomy just proximal to pouch for incomplete pouch colon.

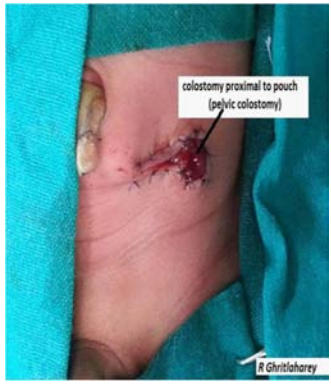

Figure 3c: Colostomy just proximal to pouch for incomplete pouch colon.

Figure 3d: Ileostomy for complete pouch colon.

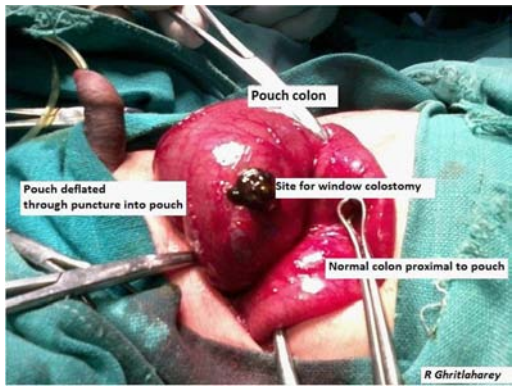

Figure 3e: Site for window colostomy for pouch colon.

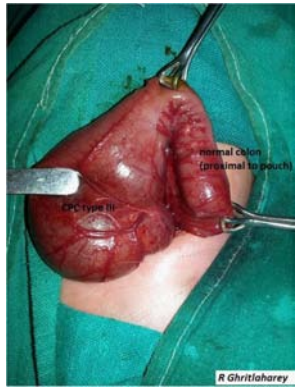

Figure 4a: Operative photograph, showing CPC type III (incomplete pouch colon).

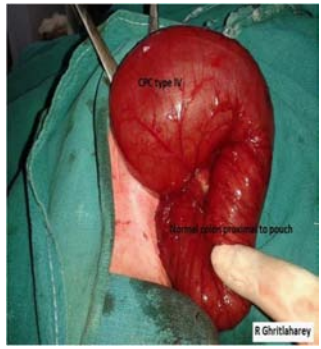

Figure 4b: Operative photograph, showing CPC type IV (incomplete pouch colon).

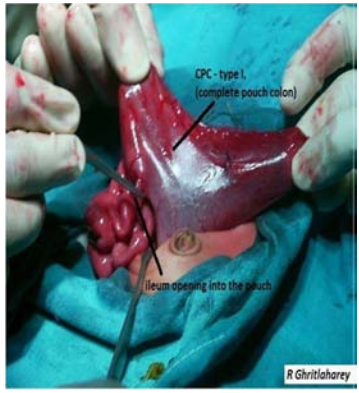

Figure 4c: Operative photograph, showing CPC type I (complete pouch colon).

TWO-STAGED PROCEDURES FOR CPC

First stage of the two-staged procedure is fecal diversion, and followed by definitive procedure without protective stoma [16]. The two - staged procedure for CPC consists of the following stages:

(A) First stage: This is an initial operative procedure and it is more or less similar to the first

stage of three-stage procedure, and includes either of one (a) colostomy just proximal to pouch, window colostomy or ileostomy with or without fistula division, (b) division of fistula and end colostomy, excision of pouch and end colostomy, and (c) division of fistula, coloplasty and end colostomy,

(B) Second stage: This stage includes the definitive procedure for CPC without protective stoma.

It includeseither of one (a) pouch excision and APPT of colon (b) pouch excision with abdomino-posterio sagittal anorectoplasty (PSARP), (c) division of fistula, coloplasty and APPT of coloplasty colon (d) division of fistula, coloplasty and sacro-abdomino-perineal pull-through, (e) division of fistula, coloplasty and PSARP, and (f) pouch excision and APPT of ileum. The main steps of definitive procedure (pouch excision and APPT of colon), executed by author (author of this chapter) during second stage of two-staged procedure for CPC are provided as figure 5a to figure 5f).

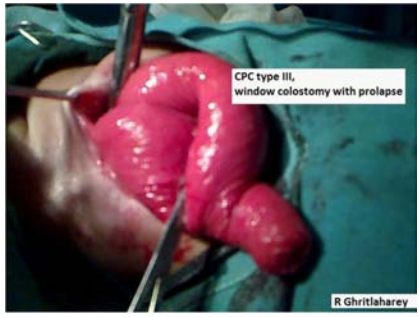

Figure 5a: Operative photograph, window colostomy (CPC type III) with stoma prolapsed.

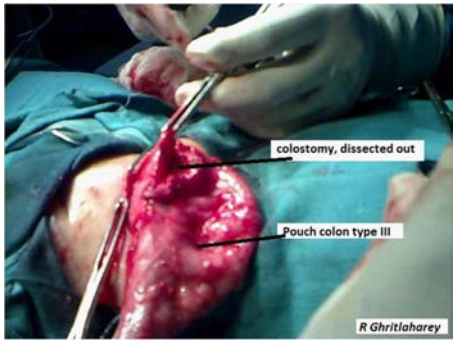

Figure 5b: Operative photograph, colostomy dissected out from abdominal wall.

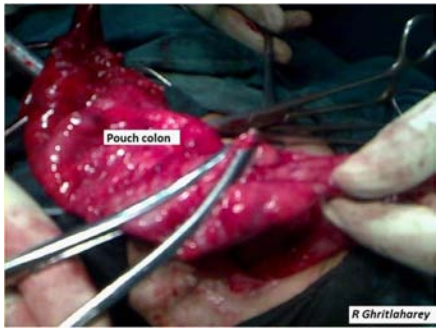

Figure 5c: Operative photograph, pouch colon disconnected from bladder after ligation of CVF.

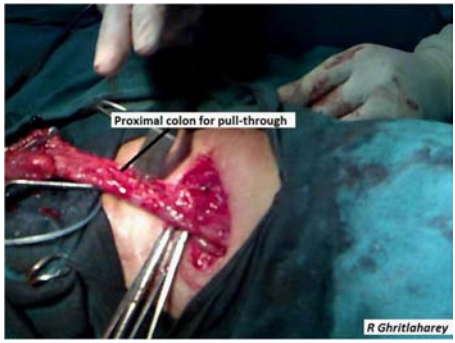

Figure 5d: Operative photograph, normal colon proximal to pouch for pull - through.

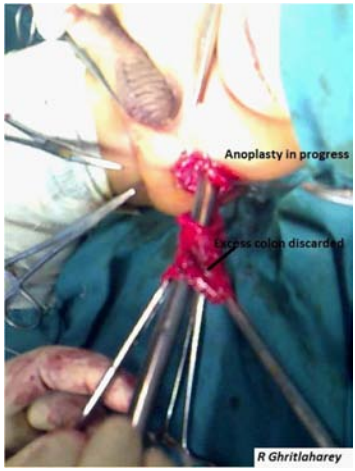

Figure 5e: Operative photograph, anoplasty in progress, diseased terminal colon discarded.

Figure 5f: Operative photograph, anoplasty completed and anal dilator to confirm patency, etc.

Advantage of Two-Staged Procedures for CPC

Two-staged procedures for CPC have certain advantage and are: (a) it provides sufficient time for proper investigations and selection of the cases for two-staged and performing the definitive procedures without protective stoma, (b) as the stoma is excised during definitive procedure in two-staged therefore; stoma closure, its cost, hospital stay and stoma related complications are avoided, (c) two-staged procedures can be performed for all types of CPC, (d) definitive procedures are well tolerated at few months of age, (e) complications following definitive procedures of two-staged are manageable with minor surgical procedures, (f) the only pre-requisite is location of stoma in left lower quadrant of the abdomen, as the left pelvic incision is preferred approach for definitive procedures for CPC and (g) best way for planning the CPC cases for future two-staged procedure is to create a stoma just proximal to pouch (2-3 cm proximal to pouch) and locate it at left lower quadrant of the abdomen [16].

Disadvantages of Two-Staged Procedures for CPC

As the definitive procedures are performed without protective stoma, so any major complications of pulled colon / ileum is difficult to manage, and definitive procedures without protective stoma may not be a good option when performing a coloplasty / colorrhaphy for preserving the colon, as there is always a risk of suture line leak [16].

SINGLE-STAGE PROCEDURES FOR CPC

Gangopadhyay, et al in 2005 advocated primary single-stage procedure for the management of all types of CPC during newborns periods with better continence and cosmesis, with low morbidity and mortality. Doing single - stage procedures at birth for CPC relieves intestinal obstruction, eliminates urinary tract contamination through fistula, and establishes anorectal continuity, with maximum potential for the development of normal defecation reflux at birth. The above all these objectives can be achieved by doing one operation rather than multiple operative procedures.

Others authors also shared their experience with single-stage procedures for CPC. A single-stage procedure includes either of one and perform without protective stoma, (a) pouch excision and APPT of colon, (b) pouch excision with abdomino-posterio sagittal anorectoplasty (PSARP), (c) division of fistula, tuboplasty / coloplasty and APPT of / tuboplasty / coloplasty colon, (d) division of fistula, coloplasty and sacro-abdomino-perineal pull-through, (e) division of fistula, coloplasty and PSARP, and (f) pouch excision and APPT of ileum [12,49,50].

COLOPLASTY

In the cases with complete pouch colon (type I and II), as the adequate length of normal colon is absent, so that tabularizing (coloplasty / colorrhaphy / tuboplasty) of the remaining colon is needed to preserve the colon functions. Surgical technique of coloplasty was introduced in 1976 by Chiba, et al for short colon and subsequently this has been used for the management of complete pouch colon with good results, and presently being used by many of the surgeons during the management of complete pouch colon. This coloplasty procedure is performed as either of one (a) during first stage of staged procedures, (b) during definitive procedure of staged procedure or, (c) during primary single – stage procedures. Variable results of coloplasty have been reported by different authors. Wakhlu, et al have the most of the experience with coloplasty and they shared their experience with coloplasty and they concluded that colonic tube must be long and the diameter of the tube small, so that it can propel stools and resist future dilatation. They also suggested that colonic tube should be made as long as possible and not more than 1.5 to 2 cm in diameter, while Gupta et al suggested that a colonic tube length of about 15 cm is enough to serve the purpose of colon. The colonic tube is made along with the mesenteric border, so that to preserve the blood supply of the tube [4,10,12,13,15,20,35,36,49].

STAGED VERSUS SINGLE - STAGE PROCEDURES OF CPC

At present moment, primary single-stage procedures for CPC is not advocated by most of the authors, due to the unacceptably high mortality associated with it, although it has certain advantages and secondly the definitive procedures for CPC at few months of age has better outcome and also well tolerated [3,4,13,15,16,18,35,38,49].

AGE FOR DEFINITIVE PROCEDURES

Doing definitive procedures for CPC at about 6 months or more is always better, as this interval also provides sufficient time for pre-operative investigations. Definitive procedure is also better tolerated at few months of age than doing it at birth. Many authors reported that the average age at definitive procedures was 15 months, ranges 3-53 months [12,15,16,35,48].

COMPLICATIONS

Complications are known to occur during the management of cases with CPC, whether managed by staged procedures or primary single-stage procedure.

Complication Occurring Following First Stage

As in the first - stage procedures for CPC, stoma is created as colostomy or an ileostomy, and the complications are related to the creation of stoma and are; stoma necrosis, wound dehiscence, peri-stomal skin excoriation, stoma stenosis (Figure 6a), stoma prolapsed (Figure 6b), stoma bleeding recurrent urinary tract infections, fecaloma formation, diarrhoea, malnutrition, etc. These complications depend upon the type of stoma, site of stoma, etc. and many of stoma related complications may require revision surgery [13,15,16,12,18,20,35,37].

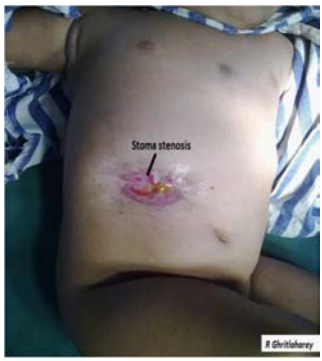

Figure 6a: Stoma stenosis following colostomy for CPC.

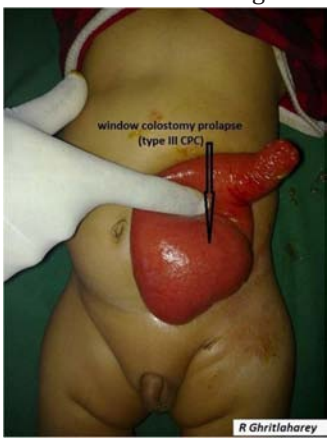

Figure 6b: Colostomy prolapsed following window colostomy for CPC.

Complications Occurring Following Colorrhaphy / Coloplasty / Tuboplasty

Coloplasty is requiring for the preservation of colon in cases of complete pouch colon. The complications related to the coloplasty are; suture line leaks, wound dehiscence, stricture of coloplasty colon, dilatation of coloplasty colon, constipation, diarrhoea, etc. During the long-term follow up, dilatation of coloplasty colon may occur, and may not function well. This dilatation is due to the fact that colonic pouch is abnormal with disorganized muscle coat with poor and abnormal peristalsis, and with tendency to dilate. Gangopadhyay, et al recommended utilization

of a shorter segment of the pouch for tuboplasty, while Wakhlu, et al recommended a long colonic tube with only 1.5 to 2 cm of diameter for coloplasty[12,13,35,52].

Complications Occurring Following Definitive Pull-Through Procedures

Complications are frequently observed following definitive procedures done for CPC and are; post-operative wound infections, wound dehiscence, intestinal obstruction, anal stenosis, mucosal protrusion, constipation, diarrhoea, peri-anal soiling, and stricture of pulled-through colon / bowel (Figure 6c). Many of the complications that occurred following definitive procedures for CPC may require additional surgical procedures for correction [12,13,15,16,18,20,35,37,49,51,53].

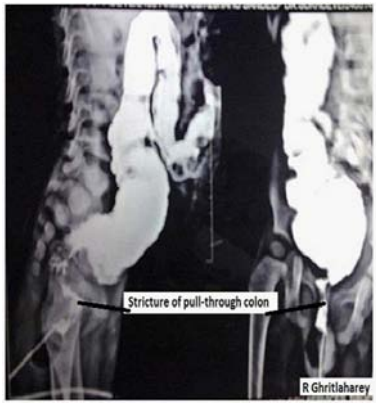

Figure 6c: Stricture of pull - through colon following single- stage procedure.

Complications Occurring Following Stoma Closure

During staged - procedures for CPC various types of stoma are being created and needed stoma closure few months after definitive procedures. This stoma closure (ileostomy / colostomy closure) may also lead to certain complications following its closure and are; wound infection, colo-cutaneous fistula, intestinal obstruction, constipation, diarrhoea, etc[12,15,18,35].

HISTOPATHOLOGY OF CPC

The gross examination of the pouch, in most of the case there is short and poorly developed mesentery and the main blood supply is from branches of superior mesenteric artery. There is absence of haustrations and teniacolli and the wall of the pouch is thicker than the normal colon. On histological examination of the excised pouch the most salient features is disorganization of the muscle layers, focal or generalized thinning of muscle layers, although ganglion cell are present but decreased number of mature ganglion cells, nerve bundle hypertrophy has also been reported, acute and chronic inflammations of the mucosa and submucosa. A study conducted for assessing the functional status of the pouch colon muscle fibres revealed that the pouch colon is either having deficient or poorly developed receptors and fails to function normally. The above neuromuscular abnormalities explain a weak or poor peristalsis as well as propensity to undergo

marked dilatation even after tubularization / coloplasty, and therefore for the same reasons the pouch should be excised for better results [13,18,22,35,51,54-57].

PROGNOSIS

The overall mortality of CPC are reported to occur in 7 to 45% of cases and this is attributed to delayed presentation, poor general condition with poor weight, presence of sepsis, types of CPC, associated lethal congenital anomalies, pouch perforation peritonitis, and complications following operative procedures, and majority of deaths occurred during initial surgical procedures [6,12,13,18,35,38]. Mortality following coloplasty has been now reduced to less than 5%, since it is being performed as a staged procedure [15]. Mortality following single-stage procedures for all types of CPC during newborn period is reported in 11%, and majority were attributed to other associated malformations [12]. There was no mortality following definitive procedures performed in 11 cases, in two-staged procedures for CPC [16]. Overall the prognosis is better for incomplete pouch colon than the complete pouch colon. The cases of complete pouch colon either managed by coloplasty or ileal pull-through, and responsible for many of the long term complications [59]. The one of the main objective during pull-through procedures is also to achieve a good continent new anus, and every effort should be made to achieve this goal, although the fecalcontinence results vary in different series [4,12,13,15,16,35,36,49,58].

CONCLUSIONS

Congenital pouch colon is a congenital anomaly associated with high anorectal malformations in which whole or part of the colon is replaced by a pouch like dilatation, and communicates distally to the urogenital tract with a fistula. This anomaly is also referred as congenital short colon and pouch colon syndrome and reported to occur more frequently in Northern, North Western, and Central part of India, with case reports from other part of the India and rest of the World. The incidence of CPC among all the cases of ARM has been reported to occur in 2-18% and among the high ARM is 10-26%, and more common in males. All males newborns presented with absent anal opening with abdominal distension and more than half of them also have meconium in urine. Female child may not always present during newborn period, due to deflation of pouch / bowel through fistula. A characteristic radiological picture on plain x-ray of abdomen is diagnostic and possible to diagnose CPC pre-operatively in more than 3/4th of the cases. The standard procedure for the management of CPC is three-staged procedures, although it can also be managed with single-stage or two-staged procedures.

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