Review Article

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Congenital pouch colon - Then and now

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ABSTRACT

Congenital pouch colon (CPC), a condition associated with anorectal agenesis has been reported mainly from the Indian subcontinent though there have been few case reports from other areas. The pouch differs from a normal colon structurally, histologically and functionally. The management involves a diversion colostomy at birth with or without the excision of pouch followed by pull through. This review article attempts to compile all the available literature on pouch colon along with the authors' experience and discuss the relevant issues for proper diagnosis and management. A changing trend has been seen in the most common type of CPC seen over the years from complete congenital pouch colon that accounted for more than 70% of cases earlier to incomplete pouch colon that is more commonly seen now.

KEY WORDS: Anorectal malformation, coloplasty, congenital pouch colon, Krickenbeck

INTRODUCTION

The condition is defined as an anomaly in which all or part of the colon is replaced by a pouch-like dilatation that communicates distally with the urogenital tract by a large fistula.[1] A supralevator anorectal anomaly is associated with a colonic pouch of variable size (5-15 cm in diameter). This condition has now been included under rare anomalies and given due recognition in the Krickenbeck classification.[2,3] Though the condition is more common in northern Indian population and neighboring countries like Pakistan and Nepal with about 90% of the cases being reported from India, with the growing awareness, there are few reports pouring in from around the globe[4] The mortality has decreased from previously reported 40% to 15% if identified and managed properly.[5]

HISTORICAL BACKGROUND

This anomaly was first described way back in 1912 by Spriggs in a London Hospital Museum specimen with absence of the left half of colon and rectum.[6] Trusler in 1959 described a pouch like dilatation of shortened colon associated with high ARM.[7] Till then no name was given to this entity. Spencer in 1965[8] reported 53 cases, 43 with exstrophy of bladder and intestines were called typical exstrophia splanchnica and the remaining were called atypical exstrophia splanchnica. Blunt in 1967 discussed this condition as an absence of colon and rectum.[9] Shafie in 1971 described it as cystic dilatation of colon.[10]

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.[11] Gopal in 1978 called it colonic reservoir in a case with Rectovaginal fistula.[12] In 1981, Li[13] from China named it congenital atresia of anus with short colon malformation. Narsimha Rao *et al*[14] in 1984 suggested the name 'Pouch Colon syndrome' and proposed an anatomical classification of this condition which has been widely accepted. In 1990, Wu[15] called this condition as Association of Imperforate anus with short colon (AIASC) and suggested that cases with exstrophy of bladder and/or intestine can be called association of imperforate anus with exstrophia splanchnica (AIAES). The two terms most commonly used to describe this anomaly are congenital short colon and congenital pouch colon.[16-24] The anatomy of this malformation was first described in 1977 by Singh *et al* and subsequently in detail by Wakhlu *et al* and Chadha *et al*.[25-27] An important advancement in the management of this condition was the technique of coloplasty in 1976 by Chiba *et al*.[28] Subsequently this has been used with good results.[27,29,30]

INCIDENCE

The incidence of pouch colon varies in different parts of the world. Apart from the northern part of the Indian

subcontinent, there are only sporadic case reports from other parts of the world including China, Japan, Sweden, UK and USA.[1,4,31] The incidence of pouch colon among all cases of anorectal malformation, in the northern India has been reported to be between 5% to 10%. The incidence in tertiary centers is between 10-15% of all cases of anorectal malformation and even upto 20% of all cases of High anomalies.

In the recently conducted survey from various pediatric surgical centers in India, Pakistan, Bangladesh, Nepal, Srilanka, Italy, Sweden and Japan on this anomaly, the incidence was reported to be the highest in North India (Kashmir, Chandigarh, Delhi, Lucknow, Varanasi) but decreased as we proceeded towards east. It was uncommon in Bangladesh (1.07%) [Figure 1] However, in Pakistan, the incidence was as high as 8-10% of all anorectal malformations. From Sweden, Japan, Italy only sporadic cases were seen and reported as curiosities. (Personal communications).

Pouch colon 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 the authors series (152 cases, 1992-2005), it was significantly higher in males than in females (7:1)

AETIOLOGY AND EMBRYOGENESIS

The exact embryogenesis is not known. Trusler in 1959 proposed that the dilatation was because of chronic obstruction but this theory was discarded as the pouch fails to decrease in size even after colostomy.[7] Another theory proposed was aborted hindgut development following obliteration of the inferior mesenteric artery early in fetal life.[32] Chatterjee proposed that the caecum and right colon develop normally from the post-axial midgut when this portion of the midgut is stimulated by normally developing hindgut.[17] Thus improper development of the postaxial midgut or presplenic gut is due to a primary disorder of the proximal end of the hindgut or post-splenic gut.

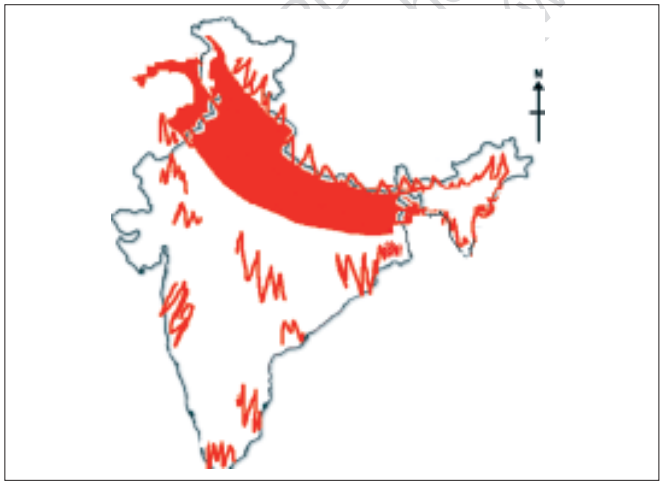

Figure 1: Geographical distribution of congenital pouch colon. It comprises of 5-15% of all cases of anorectal malformation in Northern India, 8-10% in Pakistan and 1.7% in Bangladesh. More than 500 cases in 10 years are seen in 4 major cities (Chandigarh, Delhi, Lucknow and Varanasi).

Wu Yuejie suggested that faulty rotation and fixation of the colon leads consequently to a disturbed longitudinal growth.[15] Chadha 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.[30] Wakhlu *et al* have postulated that congenital pouch colon (CPC) represents a stage in the development of cloacal exstrophy and is the combined effect of defective development of the splanchic layer of the caudal fold and failure of rotation of the gut causing defective longitudinal growth of the colon.[33]

As 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 only in 50% cases of distal pouch colon. Also a genetic predisposition needs to be ruled out.

In the authors view, the high density of cases in the northern belt of Indian subcontinent points towards environmental factors with deficiency of iodine or vitamin B as some of the possible factors contributing to this anomaly.

In north Indian belt, the land is very fertile and farming is the main occupation. Pesticides are used liberally and the population is mainly vegetarian. Also, most cases of ano-rectal anomalies belong to the low socio-economic status. All these factors suggest more of the environmental factors affecting or precipitating the anomaly at a window time after the conception when the hindgut is developing and differentiating in to urinary and intestinal tracts.

CLASSIFICATION

The term short colon has been used in the past to misleadingly describe pouch colon. However, now the term "short colon" should be used exclusively to describe a shortened length of left colon without anorectal anomaly that is also narrow in caliber and the babies are usually born to the diabetic mothers. The condition "Short Colon" was first classified by Chiba *et al*. in Table 1.

Type 3 of this classification can be described as CPC.

Table 1: 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 bowel and bladder (small and narrow colon).
5. Short colon due to abnormal vessels and the like.

Type 5 including the abnormal vessels forms the part of the pouch colon. Pouch colon also needs to be differentiated from congenital segmental dilatation of the colon, without any anorectal anomaly.[34]

A widely accepted classification is based on the length of normal colon present proximal to the dilated pouch as given by Narsimha Rao *et al*[14] [Table 2].

Wakhlu *et al*[33] simplified the classification, depending on the length of normal colon and the management planning in relation to need for coloplasty [Table 3].

In the authors view, the terms "Incomplete" and "Complete" pouch colon may be more appropriate. The term short colon should preferably be avoided for this condition to avoid confusion in terminology. Also it is the feasibility to use the remaining colon for definitive pullthrough that would be more important rather than the 8 cm length of colon. Thus a modified version of Wakhlu's classification has been proposed [Table 4, Figure 2].[1]

Initially, in India, complete congenital pouch colon was more commonly seen and accounted for more than 70% of cases till 1985. Interestingly, during the past two decades, it is the incomplete pouch colon that has become more common.

CPC should have the following anatomical criteria:

1. There is anorectal agenesis.
2. Total length of the colon is short
3. Colon has a pouch formation for a varying length - saccular or diverticular with the collection of meconium or fecal matter
4. The blood supply to the pouch is abnormal
5. The colon wall is thick and muscular with hypertrophied mucosa
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 colon and the normal bowel. The pattern changes suddenly and sharply.

Table 2: Types of pouch colon based on length (1984)

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: Types of pouch colon based on need for coloplasty (1996)

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

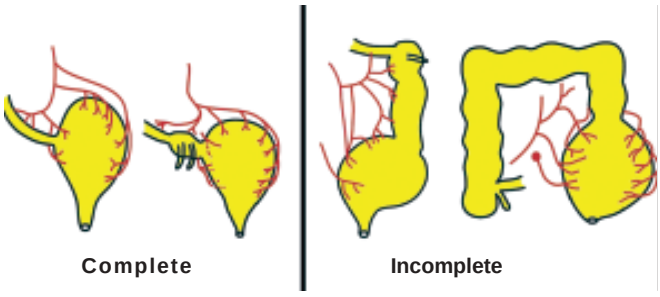

Figure 2: Types of congenital pouch colon

Table 4: Classification of congenital pouch colon (2005)

1. **Complete congenital pouch colon** - If there is either no or little normal colon left that is not enough for performing the pull through. In this situation, a coloplasty procedure would be required to retain only 15 cm length of pouch colon 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.
2. **Incomplete congenital pouch colon -** where the length of the normal colon is adequate enough for performing the pull through, without the need for doing a coloplasty. The procedure would involve excision of 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.

Cited from Gupta DK, Sharma S. Congenital pouch colon. In: Anorectal, Malformations, 1st ed. Editors. Hutson J, Holschneider A, Springer Heidelberg: 2006. Chapter 11: p. 211-22 with kind permission of Springer Science and Business Media

Associated genitourinary malformations (Cloacal anomalies, double vagina, exstrophy) are common in girls.

In complete CPC there is a large dilated thick walled pouch occupying most of the left side of abdomen. Caecum, if present almost always opens into the sac from right side. It may be associated with an absent, rudimentary or double appendix. Ileum opens into caecum or the pouch from right side and there is associated malrotation. The pouch has poorly developed mesentery and is supplied by the superior mesenteric artery on the superior and right side and an arcuate extension of superior mesenteric artery on the left side.[35] Inferior mesenteric artery is present only in incomplete

pouch colon 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 genitourinary fistula of the pouch opens into the posterior wall of the bladder in males. Fistula is usually quite broad and thick walled. In females, colocloacal fistula is the most common followed by colovaginal and colovestibular fistulas.

Pelvic musculature is variable in cases of pouch colon.

HISTOPATHOLOGY

Grossly, the pouch has a short and poorly developed mesentery. The wall of the pouch is thick, the *Taenia coli* are absent or ill defined, haustration and the appendices epiploicae are absent. The main pouch is supplied by the branches arising from the superior mesenteric artery that form a leash of vessels around it.

The pouch wall consists of normal number of ganglion cells though few have found reduced and very small ganglion cells.[12,14,25,36,37] Nerve bundle hypertrophy has also been reported but is not the regular feature.[35] Congestion of the mucosa and focal hemorrhages are seen commonly.[36,37] Ectopic heteroplastic tissue has also been reported.[38] In a detailed review of these cases, the authors found the following histological features in patients with pouch colon;

1. The muscle coat did not have the differentiation of the inner circular and the outer longitudinal muscles in most cases of pouch colon. The muscles were also arranged in a decussating pattern. The circular muscle was incomplete in 50% cases. The wall of the blood vessels was normal [Figure 3A].
2. The ganglion cells were mature and present in all cases with the presence of normal or occasionally hypertrophic nerve bundles. However, there were giant ganglia seen in 10% cases [Figure 3B].

The most salient feature is disorganization of the muscle coat in an arborhizing manner.[1,36,38] This is possibly responsible for the absence of normal peristaltic activity in these cases, requiring the removal of dilated pouch and retaining only the normal bowel.

CLINICAL PRESENTATION

Most cases present in the early neonatal period with an absent anal opening and distension of the abdomen with or without meconurea. The association of bilious vomiting with early gross distension of abdomen in a case of anorectal malformation is strongly suggestive.

In females, pouch colon is often associated with a cloacal anomaly.[39] The female baby passes meconium from abnormal opening. There may be double or septate vagina and the fistulous communication may open in one of the hemivaginae or between the two into the cloaca. In cases of colonic perforation that occurs early in cases of grossly distended pouch colon, the baby may present with septicaemia, gross abdominal distension with prominent veins, fluid and electrolyte imbalance and features of peritonitis.

When the fistulous connection with urogenital tract is large, specially in a female child with colocloacal fistula the child may present as late as few months after birth, as the child remains decompressed.. Usually at that time these children are constipated and passing faeces from an abnormal opening.

Sometimes a child may present with a colostomy done by a person not aware of this condition. Usually in these cases the presentation is with complications of colostomy

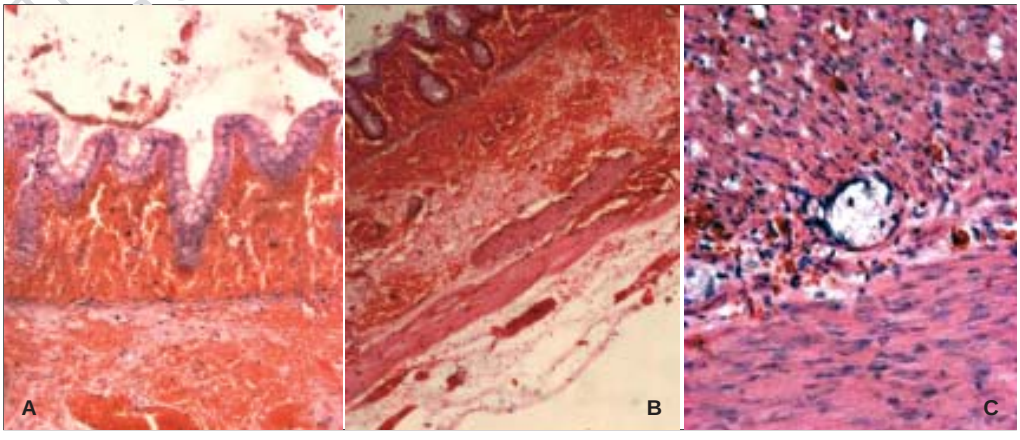

Figure 3: A) Photomicrograph showing flattened mucosa, marked congestion in the submucosa and dilated vessels (10x), B) Photomicrograph showing flattened mucosa, widened submucosa and discontinuation of circular muscle coat (10x), C) Photomicrograph showing giant ganglion cell between longitudinal and circular muscle coat (40x)

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 doing a contrast radiography and occasionally only while doing the definitive operation.

INVESTIGATIONS

Majority of the patients can be diagnosed by an erect skiagram in addition to the conventional invertogram done for anorectal malformations. 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. The pouch is proximal to the pubococcygeal line in the invertogram.

A false diagnosis can be made when there is significant dilatation of sigmoid colon or localized pneumoperitoneum following perforation in patients with anorectal malformation presenting late or in female babies with rectouterine fistula where the massive dilatation of uterus with meconium and gas may mimic pouch colon.[26]

An early perforation in cases of high ARM is suggestive of pouch colon especially if the baby comes from an area where pouch colon is commonly seen.

A detailed work up of the baby at the time of definitive surgery includes ultrasound of abdomen, Intravenous urogram and voiding cystourethrography and echocardiography to evaluate for associated anomalies. Spiral CT with 3-D reconstruction of pelvic musculature or MRI of pelvis are optional to study pelvic musculature.
Fallacies in diagnosis

The following causes may lead to fallacies in diagnosis:
- Massive dilatation of sigmoid in anorectal malformation.
- Hydrometrocolpos (with fistula).
- Pneumoperitoneum.
- If colon is fully decompressed by a large fistula, with the genito-urinary tract.

ASSOCIATED ANOMALIES

Genitourinary system is most commonly involved with associated anomalies followed by gastrointestinal and others. Table 5 summarizes associated anomalies found with congenital pouch colon.[1,35-45]

MANAGEMENT

Preoperative resuscitation includes 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 catheterization of the bladder.

Table 5: Associated anomalies reported in literature (N=566)[1,35-45]

Anomalies	No. of cases
I. Genitourinary system	
Post urethral diverticulum	2
Hydronephrosis	40
Hydroureteronephrosis	16
Vesicoureteric reflux	36
Renal aplasia and dysplasia	15
Renal agenesis	8
Renal ectopia	3
Pseudo exstrophy bladder	3
Bicornuate uterus	29
Hypospadias	15
Cryptorchidism	18
Duplication of male urethra	1
Megalourethra	1
Stricture urethra	1
Bifid penis	1
Double uterus/vagina	12
Septate vagina	11
II. G.I. System	
Double appendix	34
Absent appendix	61
Malrotation	29
Duplication of gut	5
Duplication of colon	2
Double pouch colon	1
Meckel's diverticulum	11
Oesophageal atresia	7
Other anomalies	
Sacral agenesis, other vertebral anomalies	24
Meningomyelocele	2
Congenital heart disease	19
Prune belly syndrome	6
Congenital talipes equinovarus	5
Hemivertebrae	4

The aim of surgery is to utilize the available length of colon for absorption and storage capacity as well as capability for propelling faecal matter onwards with a continent anal opening.

In incomplete congenital pouch colon, an adequate length of normal colon is present so the pouch can be excised and still the colonic function is preserved. In complete congenital pouch colon these objectives can be achieved only by tubularizing the pouch in the form of Coloplasty. However, more complications are anticipated with the preservation and use of pouch colon as a tube. 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 non-functional bowel.

At present single stage surgery for congenital pouch colon is not advocated as there is unacceptably high mortality associated with it. Although there are certain advantages

of single stage surgery but they are not sufficient to warrant a major surgical undertaking in a neonate with associated anomalies and complications.

Figure 4 outlines the algorithm for the surgical management of pouch colon. The procedure adopted depends upon the choice of the surgeon, condition of baby at presentation, technical skill of the surgeon and the availability of facilities for major neonatal surgery and post-operative care.

Proximal diversion may be in the form of:

a) End colostomy after division of fistula and excision of the pouch in incomplete pouch colon (preferred approach).
b) End colostomy after division of fistula and coloplasty in complete pouch colon.
c) Window colostomy in which an opening is made on the anterior surface of the colonic pouch without attempting to ligate the fistulous connection.
d) Proximal ileostomy in complete pouch colon
e) Transverse colostomy in incomplete pouch colon.

Excision of the pouch with an end colostomy is the procedure of choice. Window colostomy is a simple surgery, can be done with minimum anaesthesia time in a sick neonate, provides adequate decompression and a time period to allow for weight gain and be fit for second stage. However, complications with window colostomy are known [Table 6]. A window colostomy should be condemned and a proximal diverting stoma or tabularization of the pouch with a terminal stoma should

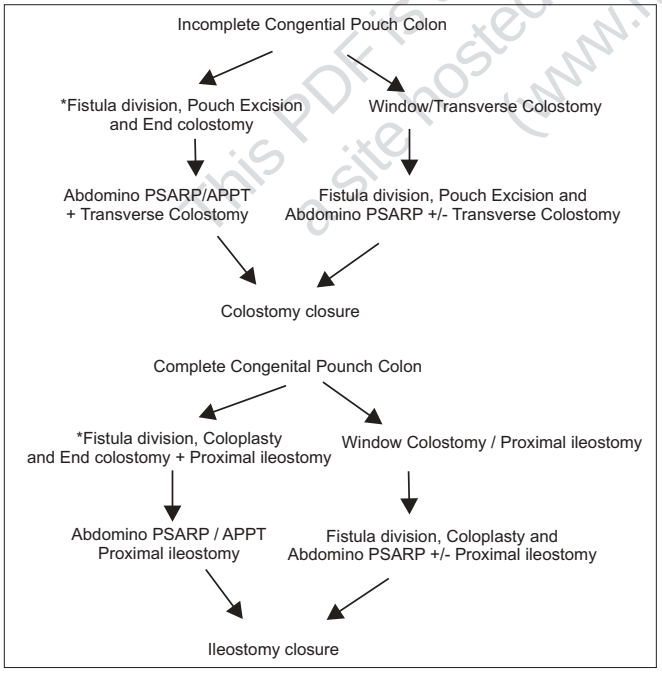

Figure 4: Algorithm for management of congenital pouch. *Preferred approach APPT - Abdominoperineal pullthrough

Table 6: Various complications associated with congenital pouch colon

1.	Related to window colostomy: Recurrent urinary tract infection. Incomplete decompression Massive prolapse Bleeding from the prolapsed bowel Recession and stenosis Pouchitis Enterocolitis Adhesive obstruction Septicaemia Failure to thrive
2.	Related to colostomy- Anemia, excoriation of skin, diarrhoea, poor weight gain, prolapse, stenosis
3.	Related to coloplasty- Suture line leak, wound dehiscence, mortality
4.	Related to pullthrough - Mucosal prolapse, anal stenosis, colonic dilatation.
5.	Related to short length of colon - Recurrent watery diarrhoea, poor weight gain

be preferred for the initial surgical management The mortality following window colostomy was reported to be in the range of 15-20%.[35] However, the mortality following coloplasty with end colostomy in the newborn period is higher, hence it may be wiser to perform it at a later date.

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

The coloplasty is done after mobilizing the pouch completely by division of inferior mesenteric artery (if present) and incising the pouch on the antimesenteric border, preserving the vascularity. The tube is fashioned over a red rubber catheter to obtain uniform diameter. Variable results have been reported by different authors.[29,46-48]

The pouch has been reported to be split longitudinally in analogy with Bianchi's intestinal lengthening procedure and used to create a vagina and reconstruct the anorectum with preserved blood supply.[49] Some authors have even reported satisfactory results with a patch graft of the pouch colon over the pulled-through ileum in complete pouch colon.[50]

COMPLICATIONS

Table 6 outlines the various complications associated with congenital pouch colon. Most of these are related to window colostomy. Recurrent urinary tract infection occurs due to persistent colourinary fistula and associated Vesicoureteric reflux. Incomplete decompression of pouch through window colostomy requires regular washouts. Massive prolapse requires revision, may be associated with bleeding from the prolapsed bowel and

herniation of bowel loops.[51] Suture line leak following coloplasty has become negligible ever since proximal ileostomy has been used. Minor wound dehiscence occurs in 4-5% of patients. Mortality following coloplasty has been reduced to less than 5% since it is being performed as a staged procedure. Colonic dilatation has been a problem following coloplasty in long-term follow-up.[52] This may be due to the fact that the colonic pouch is abnormal histologically and has a tendency to dilate because of disorganized muscle coat and abnormal peristalsis. The utilization of a shorter segment of the pouch for tuboplasty is thus recommended.

PROGNOSIS

The prognosis depends on the weight of the child, age at presentation, presence of sepsis and perforation, associated congenital anomalies and most importantly on the type of pouch colon**.** The prognosis is better in cases of incomplete pouch colon as cases of complete pouch colon suffer from recurrent watery diarrhoea due to short length of large bowel.

RESULTS

The postoperative course depends on the condition of the perineum, muscle complex and associated anomalies. The presence of a good perineum is essential to attain good continence results and prevent mucosal prolapse. A bad perineum with sacral agenesis in a complete congenital pouch colon may suffer from poor continence status postoperatively.

During the follow-up examination, 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 shows normal caliber in most of the cases; however, dilatation of the tube coloplasty is a serious problem, though rare.

In authors experience, as the anatomy and the histology of the pouch colon is abnormal, even the tube made from the dilated pouch does not work well. It does not contribute effectively to the colonic motility. Rather, the postoperative complications like mucosal prolapse, incontinence, mucus discharge; skin excoriation and the colonic ectasia are more common than those with other forms of anorectal malformation. Window colostomy is associated with serious complications like massive prolapse, incomplete evacuation of pouch, bleeding and excoriation and is thus not favoured. Excision of the pouch in toto with an end colostomy (using normal colon) is the preferred procedure, wherever feasible. An attempt should be made to excise the pouch even in cases with colonic perforation.

The continence results vary in different series. This is mainly because of the difference in the available infrastructure and the surgical expertise. While some report a poor results following definitive surgery, others have attained good continence. Babies with incomplete pouch colon fair well with normal continence, physical, motor and behavioral development. Cases of complete pouch suffer from increased frequency of stools for the initial 3-6 months though the frequency decreases with growth of the child and dietary modifications.

Excision of the pouch and end enterostomy has been associated with maximal survival.[53] Overall mortality of congenital pouch colon in the newborn age was previously as high as 30-40% but has now come down to 10 to 20% with growing awareness of this condition and improvement in the surgical management and neonatal care.

The crux of the treatment lies on timely diagnosis and planned management. The length of bowel used for coloplasty for cases with complete pouch colon should not be larger than 15cm in length for adequate propulsion of faecal matter as well as to utilize the colonic surface for water absorption and form formed stools. With the most common type of CPC now being incomplete congenital pouch colon than the complete congenital pouch colon that was more commonly seen earlier, better results of operation are expected as it is preferred to excise the pouch in cases of incomplete pouch colon and do away with the abnormal colon and its related complications.

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