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Original Research Article

Primary single stage repair of newborn babies with pouch colon (anorectal malformation) in a tertiary setup

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ABSTRACT

Background: The standard procedure in the management of pouch colon is the staged procedure (SP), which has well-known disadvantages. We believe that staged procedure is unnecessary and single-stage procedure (SSP) can be done in the congenital pouch colon cases.
Methods: Patients with pouch colon who underwent SSP (90 cases from 2008 to 2013) and SP (100 cases from 2003 to 2008), 65 and 55 of whom, respectively, were in regular follow-up for more than 3 years, were evaluated.
Results: In SSP, male/female ratio was 87:3; in SP-94:6. Mean age of patient in SSP was 3.7 days and in SP 15 months. The distribution of cases into types I, II, III, and IV was 7, 16, 18, and 24 in SSP, and 6, 14, 15, and 20 in SP, respectively. The ratio of PSSP/SP for partial pouch colon (I and II) was 23:20 and for complete pouch colon (III and IV) was 42:35. The ratio of SSP/SP in terms of continence, mortality, and cost was 81:60, 17:53, and 1:6, respectively.
Conclusions: Primary single-stage procedure gives better continence and cosmesis, with low morbidity and mortality at a low cost, and hence is recommended.

Keywords: Pouch colon, Single stage procedure, Staged procedures

INTRODUCTION

Pouch colon, the globular dilatation of a shortened colon associated with anorectal malformation (ARM), also known as pouch colon syndrome or congenital short colon, accounts for 2% to 3.3% of all ARMs and 26.6% of all high ARMs.[1-6] Management of pouch colon is controversial with very poor final result and outcome. Classical management options include initial management with a) diversion (window colostomy/proximal colostomy/ileostomy) with or without division of fistula, b) excision of the pouch and end colostomy after division of fistula, and c) coloplasty and end colostomy.

This is followed by a definitive procedure in the form of a) partial pouch colon: (types III and IV) pouch excision with abdomino–posterior sagittal anorectoplasty (PSARP) / abdominoperineal pull-through, b) complete pouch colon: (types I and II) division of fistula with coloplasty and abdomino-PSARP with proximal ileostomy, c) if coloplasty and end colostomy were done earlier after colostomy mobilization, abdomino-PSARP+proximal ileostomy.

In the staged procedure (SP), after a relatively small initial procedure, with time the proximal bowel and distal pouch would not get decompressed properly because of functional obstruction (stenosis/prolapse) and even continence results are very poor with high morbidity and mortality.[7] This study tries to highlight the ease and advantages of the single-stage procedure (SSP) irrespective of type of pouch colon at birth without a prior diversion procedure.

METHODS

All the cases of pouch colon diagnosed clinically supported by radiological evidence (babygram, invertogram, cross-table prone lateral view, ultrasonogram) admitted at S.G.R.D. Hospital, Department of Paediatric Surgery, Amritsar, during the period 2003 to 2013 were included in the study. During this period, 100 cases were managed before 2008 with standard SP, and 90 cases were managed with SSP after 1997. Of these, 55 cases of SP and 65 cases of SSP who were in regular follow-up of more than 3 years were studied. All the patients were evaluated for associated congenital anomalies with X-ray abdomen, ultrasonography abdomen, micturating cystourethrogram, and echocardiogram whenever possible. After exploratory laparotomy with left lumbar hockeystick incision, the type of pouch colon was assessed according to the classification of Narasimharao et al, and decision was made accordingly.[1] In the follow-up period, all the cases were evaluated for continence with Kelly's scoring, mortality, and cost of treatment.

The SSP includes laparotomy, ligation of the fistula, followed by primary abdominoperineal pull-through/ abdomino-PSARP after pouch excision (partial/total pouch colon). Abdomino PSARP includes initial posterior sagittal approach and placement of tube stent, followed by change of posture for abdominal approach with dissection and division of fistula, followed by anoplasty with the pulled-down colon. The distribution of various operative procedures performed in both groups (SP and SSP) (Table 1).

Table 1: Distribution of various operative procedures performed in both groups (SP and SSP).

	n (%)	APPT	Abdominal PSARP
Staged procedure (55) stage 1			
Proximal colostomy	22 (40 %)		
Ileostomy	18 (32.7%)		
Fistula division and end pouch colostomy	15 (27.2 %)		
Staged procedure stage 2			
Partial pouch, pouch excision		12 (21.8%)	8 (14.5 %)
Complete pouch, pouch excision and proximal ileostomy		22 (40%)	13 (23.6%)
Stage III Ileostomy closure	35 (63.6%)		
PSSP (65 cases)			
Pouch excision + Fistula ligation		45 (69.23%)	20 (30.77 %)

RESULTS

In our study, pouch colon accounted for 30.64 % of all ARM (total ARM cases during the period 2003 - 2013 was 620) and 44.1 % of all high ARMs (high ARM cases were 430). Male predominance was noted in our series (9:1).

All the patients with congenital pouch colon had a high variety of ARM and presented with absence of anal opening at birth with abdominal distension. Patients presented to us at a median age of 3.1 days. Incidence of various types of pouch colon (Table 2).

Table 2: Incidence of various types of pouch colon.

	Types	SP (%)	SSP (%)
Complete pouch colon	1	6 (10.9 %)	7 (10.7 %)
	2	14 (25.4 %)	16 (24.6 %)
Partial pouch colon	3	15 (27.2 %)	18 (27.6 %)
	4	20 (36.3 %)	24 (36.9 %)

Average birth weight was 2240 g. Associated anomalies were found in 31%, and the genitourinary and cardiac anomalies were predominant. Clinically and radiologically, pouch colon was suspected and confirmed in 75% of cases; in the rest, pouch colon was diagnosed at the time of surgery especially in some of the type 4 cases. In our study as we see majority of the cases were of type 4 pouch colon (36.6 %) . Complications encountered are listed in Table 3.

In our study in the SP group, the incidence of diarrhea was 47.2%, and constipation was seen in 16.3%, whereas in SSP group, diarrhea was seen in 15.3%, and constipation in 9.2% cases.

Rest of the complications are seen inumerated in Table 3.

Table 3: Complications of SP and SSP of pouch colon.

SP	n (%)	SSP	n (%)
Related to initial diversion		**Early**	
Prolapse	15 (27.2%)	Constipation	10 (15.3%)
Stenosis	7 (12.7%)	Diarrhoea	15 (23%)
Diarrhoea	20 (36.3%)	Septicaemia	12 (18.4%)
Malnourishment	22 (40%)	Wound breakdown	5 (7.6%)
Retraction	5 (9%)		
After definitive procedure and stoma closure		**Late**	
Constipation	9 (16.3%)	Constipation	6 (9.2%)
Diarrhoea	26 (47.2%)	Diarrhoea	10 (15.3%)
Wound infection	3 (5.4%)		
Incisional hernia	7 (12.7%)		

In our series, the median age at the time of initial diversion procedure was 3.1 days, age at the time of definitive procedure was 9 months, and age at the time of stoma closure was 15 months. Primary single-stage procedure was done at the median age of 3.7 days. The duration of hospital stay for the initial diversion procedure was 7 days; for definitive procedure was 14 days; for stoma closure 7 days; and for complications (diarrhea, etc), 7 days. In total, the mean period for SP was 35 days and for SSP 12 days. The cost of treatment in SP was around 10000 Rs for initial diversion, around Rs 25000 for definitive procedure, around Rs 16000 for stoma closure, and around Rs 5000 for management of complications.

Table 4: Continence results as assessed by Kelly's method.

	Good (%)	Fair (%)	Poor (%)
SP n = 55	15 (27.2%)	18 (32.7%)	22 (40%)
SSP n = 65	30 (46.1%)	23 (35.3%)	12 (18.4%)

In SSP, it was Rs 20000. Anatomical normalcy was attained in SSP at the age of 7.4 days and at 15 months in SP. Physiological normalcy (near normal bowel frequency of 3-4 stools per day) was attained at the age of 12 -15 months in SSP and beyond 6 to 7 yrs in SP. Mortality was high (approximately 52.93%) in SP; whereasin SSP, it was 16.78% . The difference between the 2 groups was statistically significant at P 0.001. Continence was assessed by Kelly's method. In Staged procedures, (59.9%) 33 /55 cases had good or fair continence. Of the 65 cases of SSP, 53 / 65 (81.4%) had good or fair continence (Table 4). The difference between the 2 groups was statistically significant at $P < .01$ ($z = 2.82$).

DISCUSSION

The main objective in the management of ARMs are relief of intestinal obstruction and restoration of anorectal continuity at birth, with optimal anatomic approximation, sphincteric function, and early postnatal establishment of the brain-defecation reflexes, which is also applicable in the management of congenital pouch colon.[8]

Congenital pouch colon is an unusual type of ARM most common in India, particularly in North India. No exact etiology and embryogenesis could be found. We believe in the vascular ischemia theory proposed by Bourdelat et al and Dickinson et al.[9,10] In North India, congenital pouch colon comprises 4.38% to 8.3% of all ARMS and 10% to 26% of high ARMs. In our study, the incidence was much higher at 30.6% and 44.1% of all high-ARM cases whereas in a study by Gangopadhyay et al it was 15.1% and 19.5% respectively.[11] Male predominance was noted in all the series (2.25:1) and even in our series it was 9:1. Associated malformations commonly involve gastrointestinal and genitourinary system in the form of double appendix, hydronephrosis and vesicoureteric reflux; in our series it was more of cardiac (VSD, ASD, TOF) with rest same gastrointestinal and genitourinary anomalies but incidence being slightly less. Gatrointestinal and genitourinary complications were seen more frequently in study by Gangopadhyay et al.[11]

The diagnosis of pouch colon clinically requires high grade of suspicion and experience on the part of the surgeon and should be suspected in cases of disproportionate tense abdominal distension of the abdomen, especially on the left side, within 24 hours.

Majority of pouch colon cases were diagnosed only after radiological investigations, which showed the presence of semicircular pouch occupying more than half of the abdomen except in the type IV variety, which was diagnosed many times preoperatively similar to our study.

Diversion procedures have so many complications such as stenosis, prolapse, diarrhea, anemia, bleeding, skin excoriation, malnutrition, and psychological trauma. Enterostomy is a potentially morbid condition in neonates and is prone to complications with incidence of 28% to 74%.[12,13] There is much physical and psychological stress to the child, parents, and family members and even to the treating doctor in managing diversion complications, to make the child fit for the next surgery by improving his/her hematological parameters. The child loses an important period while waiting for the definitive procedure, which leads to improper continence results because the optimum and vital period for the development of the higher center control is lost. This is clearly evident in our series where definitive procedure was done at 9 months of age on average, which led to less continence results than in the other series with SP. In SSP, the definitive procedure was done very early, so that anorectal continuity could be achieved early, which led to better continence results. Similar continence results were shown in a study by Gangopadhyay et al.[11]

The Quality of life, including somatic, social, participation in group activities, relationship with siblings and psychological, is terrible, owing to poor fecal continence after surgery.[14,15] The children will have problems in peer relationships, school absentees and behavioral problems. This led us to evolve single-stage management of all pouch colon cases to reduce diversion-related complications, readmissions, and the cost of multiple operations.

As described by Moore et al, single stage operation at birth relieves alimentary tract obstruction, eliminates urinary tract contamination through fistulas, establishes anorectal continuity, maximum potential for normal defecation reflexes at birth achieves all these in one rather than multiple operations.[9] Because of the long follow-up period in SP, the families cannot bear the financial burden in a country like ours, where medical facilities are not available in each nuke and corner to treat colostomy complications, the delay of which results in so many preventable deaths. This is evident as many cases could not come for follow-up and complete the treatment. Majority of the remaining patients might have died as a result of colostomy complications, and some of them will still be living with colostomy. Methods of measuring bowel function such as rectal manometry, electromyography, MRI of pelvis, and anal endosonography only evaluate the isolated aspects of a very complicated physiological mechanism, and therefore the results of these objective evaluations often poorly correlate with the quality of life. The clinical scoring methods were devised by Kelly, Templeton et al, Kiesewetter and Chang, and Stephens and Smith according to the degree of continence and the quality of life after management.[16] We used Kelly's method to assess continence in our series, as strength of puborectalis action on digital examination is also included in this.

Abdominoperineal pull-through cases showed lower continence results than abdomino-PSARP cases; as in later procedure, direct visualization of the striated muscle complex could assist in the correct positioning of the anorectal tube at the exact anatomical position with the support of a levator, internal sphincter, and external sphincter.[17] In abdominoperineal pull-through cases, the rectum was pulled inadvertently most of the time through a space between the urethra and the anterior wall of the muscle complex. In our series, we found there is improvement in continence with age as observed by Templeton and Ditesheim, but it is not clear whether it was because of improvement in compromised sphincter function or because of voluntary exercises, daily enemas, and alteration of bowel habits, although Kiely and Pena do not believe that fecal control improves with time.[1] Primary single-stage procedure is all the more important in developing countries where colostomy is socially unacceptable. It is now thought that neuronal framework for normal bladder and bowel function exists at birth, but there is a learning or training period in which long-lasting, activity-driven, neuronal changes take place during neuronal circuitry development. By delaying the repair of anorectal anomalies, critical time may be lost in which neuronal networks and synopses would have formed resulting in normal or near normal rectal function.[20] So the earlier the definitive surgery the better will be the chances of patients achieving continence.

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