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STUDY OF CONGENITAL POUCH COLON ANOMALY IN AHMEDABAD REGION

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ABSTRACT

Background: Congenital pouch colon is an unusual abnormality of like dilation of shortened colon, associated with Ano-rectal malformation. The clinical and radiological details of this condition have been described in detail and are well established, but there is a lack of consensus regarding management protocol.

Objective: To study the epidemiology of pouch colon anomaly, analyze various modes of presentation. To see various anomalies associated and to evaluate the outcome of various surgical modalities.

Methods: A prospective study of patients of pouch colon anomaly, who were admitted in our Hospital from April 2004 to February 2007, was done. Cases ware studied as per a fixed pro-forma.

Observation and Discussion: 11.4% of all male (149) and 8.5% of all female (130) Ano-rectal malformation patients had pouch colon anomaly. They were investigated by X-ray abdomen, USG abdomen, x-ray spine and 2-D echo. Associated anomalies with pouch colon are observed, patients were managed surgically by 3 stage procedure and outcome recorded.

Conclusion: All patients except one were full term delivered. Antenatal ultrasonography could not suggest any prominent shadow in lower abdomen of fetus along with normal small bowel shadow may be antenataly diagnostic of pouch colon in a fetus. Pouch colon anomaly made 10% of total Ano-rectal malformation cases (279) and 13.46% of total high Ano-rectal malformation cases (205) who were admitted in our hospital during same study duration. The preferred surgical treatment in our study was Colorrhaphy and abdominperineal pull through with proximal loop ileostomy.

INTRODUCTION

Congenital pouch colon is an unusual abnormality of like dilation of shortened colon, associated with Ano-rectal malformation (Luzzatto *et al*., 1990; Narismharao *et al*., ? and Sharma and Harjai, 1977). Congenital pouch colon is a variant of ARM, characterized by replacement of a variable length of normal colon by a pouch like structure associated with short length of the total colon along with fistulous communication with the genitourinary system. The pouch usually terminates in a fistulous communication with genitourinary tract (Chaddha, 2004 and Buddhiraja *et al*., ?). The condition may range from complete absence of normal colon with ileum opening directly in pouch to presence of nearly normal colon with only rectum or recto sigmoid being affected (Gopal, 1978; Herman *et al*., 2000 and Kalani and Sogani, 1984). Interestingly, the entity has a special predilection for the northern areas of India and most of the literature regarding its embryology, etiology, anatomy, and management has been reported from the same region. Sporadic cases or case series have been documented from rest of the world (Chaddha *et al*., 2004; Chiba *et al*., 1976 and Shafie, 1978). The clinical and radiological details of this condition have been described in detail and are well established, but there is a lack of consensus regarding management protocol.

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Objective

To study the epidemiology of pouch colon anomaly, to analyze various modes of presentation, to study various anomalies associated with pouch colon anomaly, and to evaluate the outcome of various surgical modalities used in present study.

MATERIALS AND METHODS

This is a Prospective study of patients of pouch colon anomaly, who were admitted to B J Medical College and Civil Hospital Ahmedabad from April 2004 to February 2007, Cases ware studied as per a fixed pro-forma. The patients details were recorded using the pro-forma for demographic information, clinical features, Investigations performed operative and preoperative events and outcome of surgery. Sufficient permissions and consents were procured and clearance from the Institutional Ethical committee was obtained in advance. Data thus obtained was statistically analyzed.

RESULTS AND DISCUSSION

A total 28 cases of congenital pouch colon anomaly were studied in our Hospital from April 2004 to February 2007. Out of these, 5 cases had undergone some surgical procedure in some other institute before coming to our hospital. The patients of our study belonged to different state Gujarat: 19, Uttar Pradesh: 4, Madhya Pradesh: 1. During same duration, total 279 cased of Ano-rectal malformation (high: 205, low:74) were admitted in our hospital; thus pouch colon anomaly made 10% of total Ano-rectal malformation cases, which is comparable to the series of Chaddha *et al*. (1994) (Singh and Pathak, 1972; Spencer, 1965 and Springs, 1912).

term except one case delivered. Antenatal ultra sonography was performed in 7 cases and was reported normal in 6 and abnormal oligo-hydramnios in one case. Presentation varied in accordance to the sex of the child. Our analysis included history and records of 5 male patients, who presented in some other hospital for first stage surgery. Abdominal distension: It was predominant feature in 17 cases (61%). It was earlier and more common in male patient (15 out of 17 males: 88 %) than in female patients (2 out of 11: 18%). This was in contrast to observation of Chaddha *et al*. (1994), who reported gross abdominal distension in 50 % of their male patients and 66% of their female patients. Vomiting: History of bilious vomiting after birth was positive in 12 cases (43%). As with abdominal distension, this feature was more common in male children 10 (59%), female: 2(18%). Meconium in urine: out of all 17 male patients, 10 (59%) had passed meconium in their urine. In series of Chaddha *et al*. (1994) 56% of male patients had passed meconium in urine. Stool from Introitus : all female patients presented with complaint of absent anal opening and passage of stool through Introitus, which was noticed at variable times after birth. The two exceptions (18%) that had presented on 2nd and 5th day of life, respectively and had not passed meconium till then. Anal dimple: It was well developed and pigmented in 15 patients (53%). Sex wise, a significant difference was seen as 14 out of 17 male patients (82%) had well developed anal dimple, while only 1 out of 11 female (9%) had such prominent anal dimple.

Table 1. Incidence

	Our series	Chaddha et al[3]	Chaddha et al[4]	Gangopadhyaya et al [24]
Number of cases	28	41	39	200
Duration	2yrs.11mths	1986-1990	1991-1997	1991-2003
% of total arm	10%	9%	6.5%	15.1%
% of total arm	13.46%	15.2%	-	19.5-%

Sex ratio

In our study, 17 patients were male and 11 were females, making a male to female ratio of 1.55:1 which was comparable with the series of Puri *et al*. (Archana Puri, 2006). During same study duration, 149 male and 130 female Ano-rectal malformation patients were admitted; thus pouch colon anomaly was seen in 11.4% of all male and 8.5% of all female Ano-rectal malformation patients.

Age at presentation

Age at presentation to hospital for first surgical intervention, including those patients whose first stage surgery was performed in some other institute, mostly depended on the sex of the patient.

Table 2. Age at presentation

Age at presentation	Male	Female
0-7 days	14	3
7-28 days	2	0
1month-1year	1	5
More than 1 year	0	3

Birth history

All patients included in our study were delivered at B J Medical College and Civil Hospital Ahmedabad, were full

Table 3. Birth weight

Birth weight	No of patients
More than 2.5 k.g.	3
2 to 2.5 k.g.	11
Less than 2 kg	3
Not known to parents	11

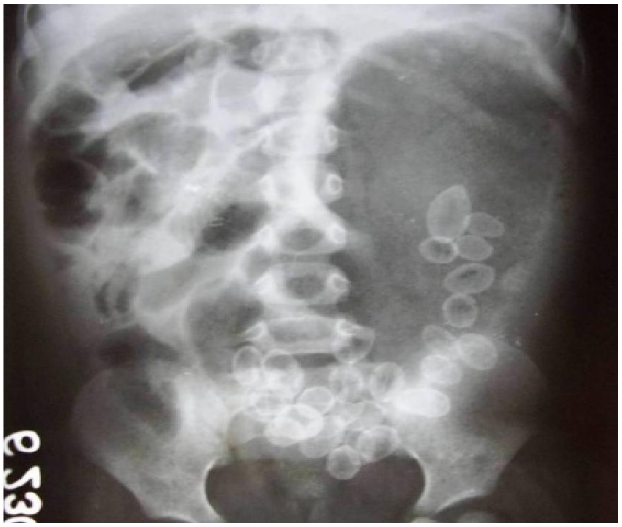

Fig. 1. Xray showing shadow of Pouch colon

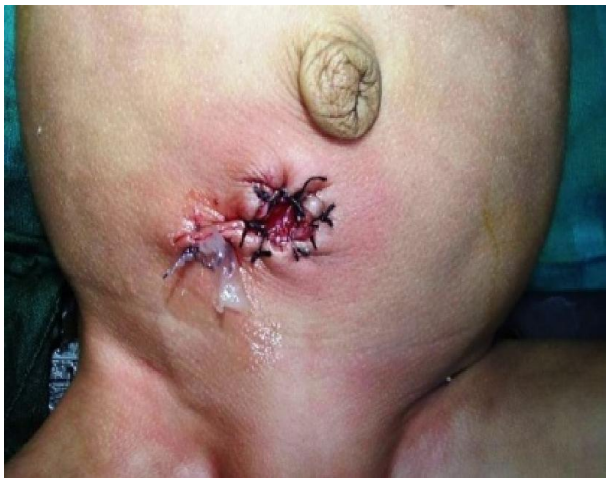

Fig. 2. Pouchostomy

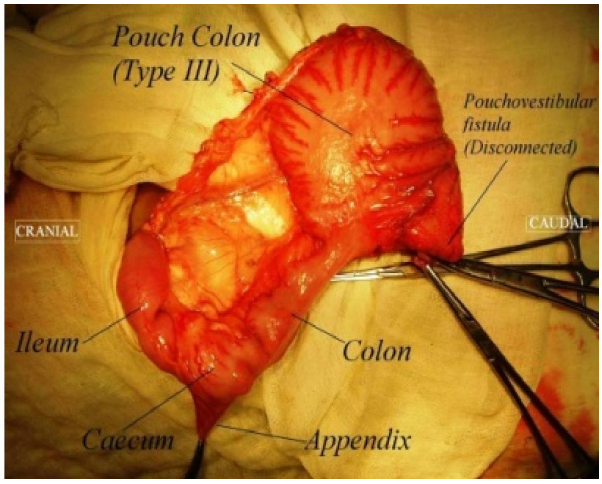

Fig. 3. Pouch Colon Type III

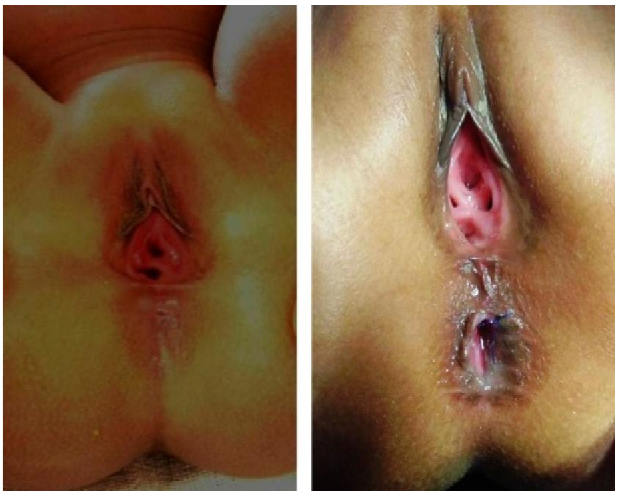

Fig. 4. Before and after pull through procedure

External genitalia: Male out of 17, five (29%) male patients had hypo spadiac penis; (glandular: 1, coronal: 3, distal penile: 1). Six (21%) male patients had undescended testes; unilateral in 4 and bilateral in 2 patients. Females: external genitalia examination showed cloaca in 7, recto-vestibular fistula in 2 and vulval anus in 2 patients.

Management

Out of 23 patients whose first stage surgery was done in our hospital, pouch colon anomaly was diagnosed preoperatively in 11 cases (48%) and per operatively in remaining 12 cases (52%). In series of Gangopadhyaya *et al*. (2005); Trusler *et al*. (1959); Vaezzedeh *et al*. (1982) and Wakhlu *et al*. (1982), preoperative diagnosis was made in 85 % cases. Commonest reason to miss preoperatively was the absence of typical radiological feature, especially when preoperative assessment was done by some junior consultant.

Table 4. Presentation (history and examination)

Presentation		Male	Female	Total
Abdominal pain		15	2	17
Vomiting		10	2	12
Meconium in urine		17	0	17
Stool from Introitus		0	9	9
Anal dimple		14	1	15
	Hypospadias	5		
	Unilateral testes	4		4
	Bilateral testes	2		2
External genitalia anomaly	Recto vestibular fistula		2	2
	Vulval anus		2	2
	Cloaca		7	7
	Pouch colon with cloacal malformation		7	7

Table 5. Pouch colon with cloacal malformation in Females

	Our series n=11	Chaddha et al [3] n=9	Chaddha et al [4] n=12
Cloacal malformation	7(63%)	3(33%)	4(33%)

Table 6. Investigations

Investigation	Findings	Total
X-ray abdomen-	With 1 fluid level	10
Gas shadow	Multiple fluid level	4
	No prominent gas shadow	2
X-ray lumbosacral spine	Spina bifida	1
Usg abdomen kub	Hydroureteric Nephrotic kidney	1
2-d echocardiogram	Asd/Vsd/Pah	6
Mcu	B/l vu relux	3

Such cases that lacked typical radiological feature were either cases of type iv pouch colon or were the cases with wide fistula with bladder/ vestibule/cloaca. Additionally, in case of female patients, significance of one constant and prominent gas shadow in left lower abdomen was gradually better perceived as our study progressed and whenever needed, such case were subjected to further investigations like contrast enema before any surgical intervention.

Type of pouch colon

Detailed comment on type of pouch colon was made on 25 cases where laparotomy was performed in first or second stage. Three cases, in which only pouchostomy was done, were excluded from classification till second stage.

Bowel prolapse: Dehiscence of pouchostomy and colostomy stitches and prolapse of bowel loops occurred in two cases and was managed by re-suturing.

Malnutrition: One patient suffered from malnutrition after pouchostomy and later, died. After colorrhaphy and pull through: Anal stenosis: One patients had defaulted and reported back after one year with anal stenosis confirmed by distal loopogram. Re-do p.s.a.r.p. was performed as stricture was too long to respond do anal dilatation. Anterior retraction of neo-anus Occurred in one female patients in whom pouch excision and pull through of colon was done. Re-do p.s.a.r.p. was performed.

Table 7. Types of pouch colon

Type of pouch colon	Our series (n=25)	Chaddha et al[3] n=41	Chaddha et al[4] n=39	Gangopadhyaya et al [24] n=105	Puri et al [23] n=22
I	8(32%)	21(51%)	13(34%)	18(17%)	7(32%)
II	6(24%)	11(27%)	18(46%)	44(42%)	10(45%)
III	8(32%)	7(17%)	2(5%)	24(23%)	2(10%)
IV	3(6%)	2(5%)	6(15%)	19(18%)	3(13%)

All patients were managed by staged surgery

First stage

Puchostomy: It was done in 9 male and 6 female patients on initial presentation. Pouchostomy was placed 1.5 below and towards right of umbilicus. Regular pouch washes were continued at home postoperatively. Proximal diversion: Proximal loop transverse colostomy was done in three male patients with type IV pouch colon. Primary colorrhaphy and abdominoperineal pull through with ileostomy: This was performed in first stage in four female patients proximally, loop ileostomy was done. Out of these four, one female was operated in neonatal period. Pouch excision and abdominoperineal pull through of colon with ileostomy: This was performed in one female patient. Proximal diversion was made in form of ileostomy.

Second stage

Colorrhaphy and abdominoperineal pull through with proximal loop ileostomy: This was performed in second stage in 9 male and 1 female patients. Regular distal loop washes were been given at home and distal loopogram was performed before ileostomy closure.

Third stage

Ileostomy closure 6 patients in our study had undergone all surgical stages, so far.

Complications

Stomal stenosis: 15cases (26%) occurred in four patients female: 3 male: 1, this was managed by regular dilatation and washes.

Pouch prolapsed: (13%) Prolapse of pouch was seen in two patients of our series. Wakhly *et al*. (1996) has reported 25% rate of combined prolapse and stenosis in his series.

Enterocolitis: One female patient suffered from repeated attacks of enterocolitis for first 6 months after ileostomy closure. Attacks were managed by bowel washes and antibiotics. Episodes gradually decreased in frequency and severity. Another patient suffered from and died of enterocolitis at home.

Histopathology: Examination of excised pouch tissue was done in 9 cases. Normal colonic mucosa was seen in 6 cases, while features of some degree of colitis were present in 5 cases. Besides, muscular thickening and hyetrophic nerve bundles were seen in 2 cases. Ganglion cells were normal in all specimens. Chaddha *et al*. (1998) and Wakhlu *et al*. (1996); Wakhlu *et al*. (1996) and Yuejie *et al*. (1990) have reported pouch colon tissue histologically normal in their series.

Contrast enema: Postoperative contrast enema was performed in 5 patients, which did not show excessive dilatation of tube in any case.

Outcome

Despite of all the possible attempts of communication personal, postal, telephonic, 6 patients from study group lost from regular follow up. Out of remaining 22 patients, 8 died after various stages at hospital or home confirmed by communication with parents. Out of these, two patients died of ventricular tachycardia during second stage surgery. One of these two had associated Atrial septal defect, diagnosed by pre-operative 2d echocardiography

Table 8. Outcome

Out come	Number of cases
Lost from follow up	6
Died	8
Waiting for next surgical stage	9
All stages completed	5
Total	28

In our Study, mortality after pouchostomy was 20% (3 out of 15 cases) and overall mortality was 28.6%. Other persons have reported mortality after first stage as 27% and 19% in series of Chaddha *et al*. (1994) and Wakhlu *et al*. (1996) respectively. Out of 28 patients, all stage of surgical management was completed in 6 patients. Out of these, one patient died of enterocolitis at home. Remaining 5 patients have survived.

Conclusion

28 cases of pouch colon anomaly were studied 17 patients were male and 11 were females, making a male to female ratio 1.55:1, Pouch colon anomaly made 10% of total Ano-rectal malformation cases. 11.4% of all male (149) and 8.5% of all female (130) Ano-rectal malformation patients had pouch colon anomaly. Antenatal ultra-sonography could not suggest any prominent shadow in lower abdomen of fetus along with normal small bowel shadow may be ante-nataly diagnostic of pouch colon in a fetus. The preferred surgical treatment in our study was colorrhaphy and abdominperineal pull through with proximal loop ileostomy. This was performed following pouchostomy/ colostomy in 10 case, while in 4 cases, primary colorrhaphy and abdominoperineal pull through with ileostomy was performed. Out of these 4cases, 2 cases were diagnosed preoperatively, while other 2 were diagnosed preoperatively.

Most common complications after pouchostomy are stenosis (26%) and prolapse (13%) of pouch. After colorrhaphy and abdomino-perineal pull through, anal stenosis, anterior retraction of anus and enterocolitis was seen in one patient each. Complications are known to occur with Congenital pouch colon whether managed by multiple stage or single stage procedures. Mortality after pouchostomy was 20% (3 out of 15 cases) and overall mortality was 28.6 %. Out of 14 cases, in whom, colorrhphy and abdominoperineal pull through was performed, 5 patients are healthy and continent and 4 patients are waiting for ileostomy closure. One died preoperatively, two died before patient was lost from follow up. 5 patients who have gone through all surgical procedures were satisfactory in terms of frequency, consistency, sensation and continence. Anal opening is of adequate caliber with healthy peri-anal skin. Variations in anatomy, presentation, initial treatment by different surgeon make it difficult to propose a standard management protocol and every patient needs a procedure appropriate to his/her anatomy. Pharmacotherapy and rigorous bowel management lead to satisfactory social continence. The Crux of the treatment lies on timely diagnosis and planned management. Intensive counseling and motivation is required when these patients reach puberty and adolescence.

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