Original Article

Congenital Pouch Colon from Al-Ahsa Region of Saudi Arabia – A Changing Demography?

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ABSTRACT

Objective: To study the demography, anatomy and management of congenital pouch colon (CPC) in the Al - Ahsa region of the Kingdom of Saudi Arabia (KSA)
Design: Retrospective descriptive analysis of CPCs was made with regard to patient demography, pre- and intra-operative diagnostic features and initial neonatal management.
Setting: Maternity and Children Hospital, Al-Ahsa region, KSA
Subjects and Methods: Records of all children with anorectal malformations (ARM) treated between August 2004 and August 2007 were reviewed. Out of the 37cases of ARMs, seven were diagnosed as CPCs.
Interventions: Records were analysed for perineal findings, X-ray abdomen and invertogram (when indicated), abdominal ultrasound, type of pouch, division of any urinary fistula, and surgical management.
Results: Six out of seven CPCs were among Saudi nationals belonging to Al-Ahsa region. The male to female ratio was 4:3. Six were type IV and one was type III pouch. Preoperative diagnoses were made in two cases by radiology and all cases showed classic anatomical features of CPC intra-operatively. 71% had associated anomalies. Excision of pouch and end colostomy was done in three, loop stoma was done in two and excision of pouch with neonatal pull-through was done in two cases.
Conclusion: CPCs are recently being increasingly reported in Saudi nationals. Adequate awareness about radiological, anatomical and histological features would lead to correct diagnosis and reporting. Appropriate neonatal management would prevent pouch related morbidity. Collective data from multiple centers across the Arab peninsula will help define the demographic pattern of this entity in the region.

KEY WORDS: anatomy, congenital pouch colon, demography, etiology, Saudi Arabia

INTRODUCTION

The condition of Congenital Pouch Colon (CPC) has been associated with ano-rectal agenesis, particularly seen in Asia and is defined as an anomaly in which whole or part of the colon is replaced by a pouch like dilatation, which mostly communicates distally with the urogenital tract by a fistula[1]. Recently the Krickenbeck classification for anorectal malformations (ARM) has classified this entity as a rare anomaly[2]. This condition is seen much more frequently in the northern, northwestern and central part of India and neighboring nations like Pakistan, Bangladesh and Nepal, accounting for more than 90% of the reported cases. Only a few reports have originated from China, Japan, Sweden, United Kingdom and other parts of world[1,3,4] but none from the Arab peninsula.

The cause of this unique geographical distribution has not yet been ascertained. We report a series of seven cases of CPCs from Al Ahsa region of Eastern Province of Kingdom of Saudi Arabia (KSA) treated at the Maternity and Children's Hospital (MCH). To our knowledge, this is the first reported series of CPCs from KSA and the Arab world.

SUBJECTS AND METHODS

Between August 2004 and August 2007, 37 cases of ARMs were treated at MCH, Al Ahsa region of the Eastern Province of KSA. Seven of these were detected to have CPC. The patient demographics are given in Table 1. We have made a descriptive analysis of the patient characteristics, anatomical features, and initial management at presentation and discussed the etiology of this rare anomaly.

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Table 1: Patient demographics

Patient No.	Sex	Type of pouch and ARM	Associated anomalies	Surgical procedure
1	M	Type IV with pouch-vesical fistula	Lumbo sacral hemivertebrae	Excision of pouch, division of fistula and end colostomy
2	M	Type IV without urinary fistula	Cleft palate and facial dysmorphism	Excision of pouch and end colostomy
3	M	Type III with pouch urethral fistula	Esophageal atresia and tracheo-esophageal fistula(TEF), VSD (VACTREL association)	Loop colostomy and repair of TEF by right thoracotomy
4	F	Type IV and rectovestibular fistula	None	Excision of pouch and neonatal pull-through
5	F	Type IV and rectovaginal fistula	Tetralogy of Fallot	Loop colostomy
6	F	Type IV and rectovestibular fistula	None	Excision of pouch and neonatal pull-through
7	M	Type IV and pouch vesical fistula	Sacral agenesis	Excision of pouch, division of fistula and end colostomy

This study was approved by the ethical committee of the hospital.

RESULTS

Out of 37 cases of ARM, seven cases were diagnosed to have CPC. There were four male and three female babies. Six out of seven cases were Saudi nationals and one was an Indian expatriate. In two of the male babies a large air fluid level occupying more than half of the width of the abdomen on plain skiagram helped in the preoperative diagnosis of CPC (Fig. 1). In rest of five cases CPC was diagnosed intra-operatively. The anatomical features of pouches in our series included thick walled abrupt change of tubular caliber of distal colon into spherical pouch without any transition zone (Fig. 2). There were absent or ill defined taenia coli, absent haustrations and appendices epiploicae. The mesentery was somewhat short and vessels were arising from superior mesenteric artery or hypoplastic inferior mesenteric artery forming a leash of blood vessels around the pouch. The wall of the pouch was thick, muscular, lined with hypertrophied mucosa. Histopathologically there was thickening of the submucosal layer and criss-cross pattern of decussating fibres in the muscle coat. The normal longitudinal and circular pattern was lost.

We categorized our CPCs according to the popular classification by Rao *et al* [5], as shown in

Table 2: Types of pouch colon (Rao *et al*)[5]

Pouch colon	Diagnosis of CPC
Type I	Absent normal colon, ileum opens directly into colonic pouch
Type II	Cecum and short segment of ascending colon opens into colonic pouch
Type III	Cecum, ascending colon and part of transverse colon opens into colonic pouch
Type IV	Most of the colon is normal, part of sigmoid and rectum are pouch like

CPC = Congenital pouch colon

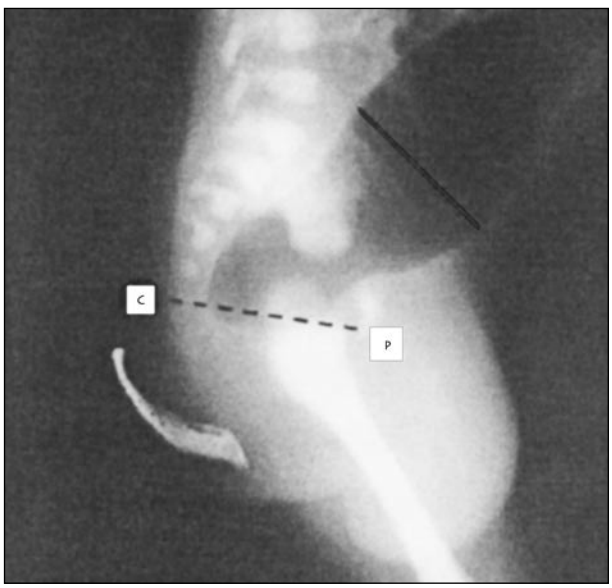

Fig. 1: Invertogram showing colonic air shadow greater than 50% of abdominal width (solid line) and intermediate ARM (dotted line denotes pubo-coccygeal line).

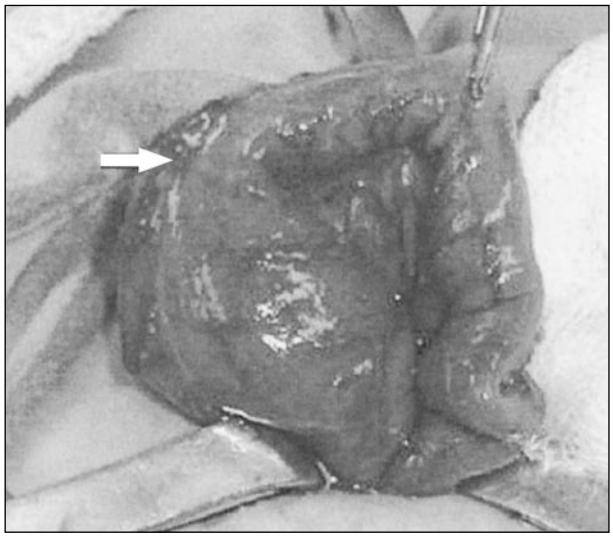

Fig. 2: Intraoperative feature of pouch colon. Block arrow shows sudden transition of tubular colon into a spherical pouch.

Table 3: Working classification of congenital pouch colon (Gupta *et al*)[1]

1. Complete Congenital pouch colon – If there is no or little normal colon left that is not enough for performing the pull-through. A coloplasty procedure would be required to retain only 15 cm length of pouch colon in the form of a tube and this is 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 colon is adequate 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 definite pull-through later. A single stage neonatal pull-through can also be undertaken if the condition of the baby permits.

Table 2 and it included six type IV and one type III pouch. Our initial management of these babies with ARM was based on recent recommendations similar to those by Gupta *et al*[1] as shown in Table 3. We performed excision of pouch with end colostomy in three of these and complete neonatal pull-through in two cases (with vestibular fistulae). Out of two remaining cases, loop stoma was constructed in a male child due to multiple congenital anomalies requiring simultaneous thoracotomy and repair of tracheo-esophageal fistula in order to minimize the operating time. Also in another female child with recto-vaginal fistula, type IV pouch and associated tetralogy of Fallot a proximal loop colostomy was constructed as a quick initial procedure due to anesthetic issues (baby developed hemodynamic instability due to Tetralogy of Fallot). The five neonates with stoma underwent definitive pull-through (abdomino - sagittal anorectoplasty) between 3 - 6 months of age. Five out of seven cases had associated congenital anomalies (2 vertebral, 2 cardiac anomalies including one VACTREL, one cleft palate and facial dysmorphism). Two out of four males had pouch vesical fistulae, one male baby had pouch prostatic urethra fistula and one female had a pouch genital fistula (recto-vaginal fistula).

DISCUSSION

Ever since its first description in 1912 by Spriggs in a London hospital museum specimen with absence of the left half of the colon and rectum[6], various terms have been used to describe congenital pouch colon including pouch colon syndrome[5], extrophia splanchnica[7], congenital absence of colon and rectum[8], colonic reservoir[9] and association of imperforate anus with short colon (AIASC)[10,11].

Interestingly, the sex ratio reported by authors outside Indian subcontinent has been almost equal (M:F = 1.27:1), while the reported incidence in India favors male preponderance (M:F = 3 - 4.3:1)[1] .We had an M:F sex ratio of 1.3:1 in our series. Similar to the recent trend, we had encountered mostly incomplete type of CPCs (six type IV and one type I). Associated anomalies were seen in 71% (5 out of 7) cases, higher than those reported in the literature[1,3,4]. Most of these cases (5 out of 7) were operated in a staged manner; *i.e.* excision of pouch, division of any urogenital fistula in the neonatal period and definitive pull-through at 3-6 months age. In two female babies with type IV pouch colon and pouch vestibular fistula, single stage neonatal repair was done.

The incidence of CPC among all cases of ARM in northern India has been reported to be between five to 10%, in Bangladesh 1.07% and in Pakistan 8-10 %[1]. Only sporadic case reports are from other parts of the world. Recently Donkol *et al*[12] have reported this anomaly in a Saudi Arabian neonate from Western province (Al-Abha region). Our report is the first case series from Saudi Arabia and the whole of Arab peninsula. In another recent series published from Al-Khobar region of KSA, the author has described 17 cases of CPCs. However, all are from northern part of India (none in Saudi nationals)[13].

Since Saudi Arabia has been harboring a significant expatriate population from India, Pakistan, Bangladesh, Nepal and Srilanka, few cases of CPCs could be expected among them. However, we have found six of our seven cases to be Saudi nationals. We believe the lack of awareness among the physicians about this entity would have led to under-reporting among Saudi nationals and expatriates. Many high ARMs, especially those without fistula, have associated dilatation of blind rectal pouch. It is important for the surgeons to

distinguish these dilatations from pouch by the features mentioned above (see results section) which otherwise prompts tapering colorectoplasty during postero saggital anorectoplasty (PSARP) instead of excision of pouch. Retained pouch tissues lead to postoperative constipation and secondary pouch dilatation with suboptimal outcome.

Various authors have tried to explain the embryological basis of this anomaly including preferential hindgut stimulation[10], vascular insult[11], faulty rotation and fixation of gut theory[9]. Recently, Gupta *et al*[1] have incriminated iodine and vitamin B deficiency, use of pesticides and low socio-economic status as possible environmental factors affecting or precipitating the anomaly at a window time after conception when hindgut is developing and differentiating into urinary and intestinal tracts.

Occurrence of CPC among Saudi neonates points towards a possibility of changing demography of this entity that is not simply explainable by population migration from high prevalence areas. The role of genetic predisposition and interplay of multiple environmental factors in the causation of this entity (mentioned above) needs to be explored in the community. Eastern province in particular has a high rate of consanguinity and interplay of above mentioned environmental factors.

CONCLUSIONS

CPC is a rare variety of ano-rectal malformation known to cluster in specific geographical areas. This is recently being increasingly and disproportionately reported in the Saudi Arabian population. Whether this is reflected by under-reporting or under-diagnosing of CPCs elsewhere, or represent a changing demography needs to be confirmed by collective multi-centeric data from the Arab peninsula. More importantly, it is necessary to identify these cases by the characteristic radiological, anatomical and histological features and to differentiate it from simple dilatation of blind rectal pouch. It is extremely important to employ correct initial management in the newborn period to decrease subsequent pouch related morbidity.

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