Asian Journal of Health and Medical Research (AJHMR)
Volume 3, Issue 1, December, Page No.01-02, 2017

ORIGINAL ARTICLE

CONGENITAL POUCH COLON WITHOUT FISTULA: OUR EXPERIENCES AND LESSONS LEARNED

Dr. Vaibhav Pandey*[1], Dr. Pranay Panigrahi[2], Dr. Rakesh Kumar[3]

[1]M.B.B.S, M.S, M.Ch., Assistant Professor, Department of Paediatric Surgery, Institute of Medical Sciences, Banaras Hindu University, Varanasi, U.P. Email address: sunny.imsbhu@gmail.com
[2]M.B.B.S, M.S, M.Ch., Senior Resident, Department of Paediatric Surgery, Institute of Medical Sciences, Banaras Hindu University, Varanasi, U.P. Email address: docpranaya@gmail.com
[3]M.B.B.S, M.S, M.Ch., Senior Resident, Department of Paediatric Surgery, Institute of Medical Sciences, Banaras Hindu University, Varanasi, U.P. Email address: drrkumar2611@gmail.com

ABSTRACT

Congenital Pouch colon without fistula: *Our experiences and lessons learned*

Introduction: *In anorectal malformations (ARM), usually the bowel ends as fistula into the uro-genital tract. ARM without fistula implies that the rectum ends blindly at the level of the bulbar urethra without a fistula. Pouch colon is another rare variant associated with ARM. The combination of these two rare variants is still rarer with unique variations.*

Material and method: *The retrospective study was conducted after approval by institutional ethical committee and case records of patients with pouch colon without fistula were assessed for gender, associated anomalies, Intra-operative complications and continence outcome. Data were coded and summarized using Statistical Package for Social Sciences (SPSS) version 16.0 for windows.*

Results: *Four cases with pouch colon without fistula presented during the study period. All patients were male. Mean age of presentation was 2.34 ±0.45 days. One patient was having Down's syndrome and 50% (2 cases) had cardiac lesion. 50% (2 cases) had pneumoperitoneum at time of presentation and presented on 3rd day of life. Invertogram showed high type of Anorectal malformation. These were managed with three staged repair. 50% (2 cases) were managed with primary Abdominoperineal pull through. 75% had Type 2 pouch and 25% had type 1 pouch colon [Figure 1-2]. All patients underwent regular dilatation. At age of three years 75 % had good continence according to Kelly's score and 25% had fair continence.*

Conclusion: *This extremely rare entity is associated with higher incidence of congenital anomalies and pneumoperitoneum with overall good prognosis.*

Keywords: *Anorectal malformation, Pouch colon, ARM without fistula, APPT.*

INTRODUCTION

Anorectal malformation (ARM) is one of most common paediatric surgical entity [1]. Usually the bowel ends as fistula into the uro-genital tract. ARM without fistula implies that the rectum ends blindly at the level of the bulbar urethra without a fistula. This is a rare entity with challenging preoperative diagnosis and intra-operative identification of the problem [2]. Pouch colon is another rare variant associated with ARM. It is defined as a sac-like dilation of the shortened colon, leading to a massively dilated distal "pouch" which fistulates into the genitourinary tract [3-4]. We are presenting our experience of management of ARM with pouch colon without fistula.

MATERIAL AND METHOD

The retrospective study was conducted in the Department of Paediatric Surgery after approval by institutional ethical committee. Case records of patients with pouch colon without fistula were assed for gender, associated anomalies, intra-operative complications and continence outcome. Data were coded and summarized using Statistical Package for Social Sciences (SPSS) version 16.0 for windows. Student t test / Mann-Whitney U test was used to compare mean/median in quantitative data whereas for qualitative data Chi- square test was used. P value <0.05 was taken as significant.

RESULTS

Overall 4 cases with pouch colon without fistula presented during the study period. All patients were male. Mean age of presentation was 2.34 ±0.45 days. All the patients had abdominal distension at time of presentation. Two patients had positive sepsis screen at presentation. One patient was having Down's syndrome and 50% (2 cases) had cardiac lesion. 50% (2 cases) had pneumoperitoneum at time of presentation and presented on 3rd day of life. Invertogram showed high type of Anorectal malformation. These were managed with three staged repair. 50% (2 cases) were managed with primary Abdominoperineal pull through. 75% had Type 2 pouch and 25% had type 1 pouch colon [Figure 1-2]. There were no intraoperative complications. All patients underwent regular dilatation. At age of three years 75 % had good continence according to Kelly's score and 25% had fair continence.

DISCUSSION

The incidence of anorectal malformations (ARM) is reported as 2.0–2.5 per 10,000 live births [1]. The pouch colon variant of ARM more commonly occurs in northern India 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 [2-4]. 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 [5] and Dickinson [6]. In North India, congenital pouch colon comprises 4.38% to 8.3% of all ARMS and 10% to 26% of high ARMs. The pouch colon in most cases presents as rectovesical fistula. High imperforate anus without fistula is defined as rectum ending blindly at the level of the bulbar urethra without a fistulous communication. ARM without fistula occurs in 5% equally in both males and females and is an uncommon presentation in the normal population. However, 50–95% of patients have Trisomy 21 and the others tend to suffer from syndromes such as Apert [7-8]. In our series only one case had Down's syndrome. Other important finding in our series was high percentage of patients with pneumoperitoneum. As pouch colon is usually associated with rectovesical fistula, the gut decompression is quite good and these patients present even quite late without much distension and rarely perforation. But in cases without fistula gut decompression is not at all present so 50% of cases presented with perforation even on second day of life. The importance of this article is rarity of the disease, and such case has not been reported previously.

CONCLUSION

Pouch colon syndrome in ARM without fistula is an extremely rare entity. The condition has challenging preoperative diagnosis and higher incidence of associated anomalies and pneumoperitoneum. The extremely rare nature of disorder and challanging management makes every case worth to be reported in literature.

Acknowledgment: Nil

Illustrations:

Figure 1: Pouch colon without fistula

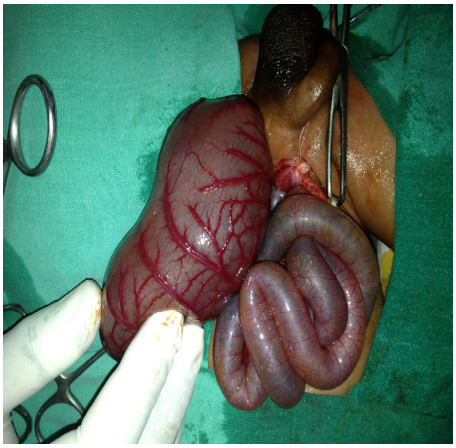
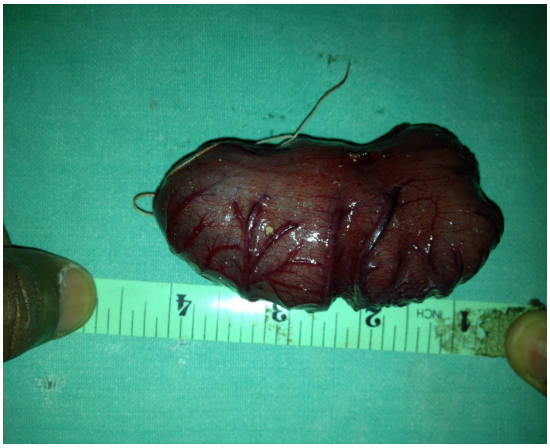


Figure 2: Excised pouch colon without fistula.



REFERENCES

1. Pena A. Anorectal Malformations. Semin Pediatr Surg 1995;4:35-47.
2. Torres R, Levitt MA, Tovilla JM, Rodriguez G, Peña A. Anorectal malformations and Down's syndrome. J Pediatr Surg 1998;33:194–197
3. Narasimharao KL, Yadav K, Mitra SK. Congenital short colon with imperforate anus (pouch colon syndrome). Ann Paediatr Surg. 1984;1:159-67.
4. Chadha R. Congenital pouch colon associated with anorectal agenesis. Pediatr Surg Int 2004;20:393–401
5. Bourdelat D, Labbe F, Pillet J. A study in organogenesis: the arterial supply of the anorectal region in the human embryo and fetus. Surg Radiol Anat. 1988;10:37-51.
6. Dickinson SJ. Agenesis of the descending colon with imperforate anus. Am J Surg. 1967;113:279-81.
7. Rao KL, Menon P. Congenital pouch colon associated with anorectal agenesis (pouch colon syndrome). Pediatr Surg Int 2005;21:125–126
8. Gangopadhyay AN, Shilpa S, Mohan TV, Gopal SC. Single-stage management of all pouch colon (anorectal malformation) in newborns. J Pediatric Surg. 2005;40;1151-5.