Original Article

Congenital pouch colon: A rare presentation of Anorectal malformation

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Abstract:

Background: Congenital pouch colon is an usual abnormality associated with anorectal malformation in which the colon is partially or totally replaced by a pouch like dilatation connected to the genitourinary tract by a fistula(colovesical).The condition varies from complete absence of normal colon with the ileum opening into colonic pouch to the presence of nearly normal colon with only the rectum or recto sigmoid being attached to a pouch which in turn connected to bladder by a band or fistula.

Objective: is to evaluate the methods of diagnosis and management of(C.P.C) with studying environmental factors especially pesticides and its prevalence.

Patients and Methods: A total number of seven cases of(C.P.C) associated with high type of imperforate anus seen in Al-Emamain Al-Kadhemain Medical City and Al-Kadhemia Pediatric Hospital from October 2004 to October 2014.

Results: Incidence of (C.P.C) was (2.9%) of all cases of high type of imperforate anus with male predominant (2.5:1).Most of patients were from Baghdad's periphery with suspicioun of relationship of pesticides and its prevalence.Preoperative erect abdominal X-ray was diagnostic in (71%) especially type I and II .The most common type of (C.P.C)was type (II) (34%) followed by type (I) (28%). Abdomino-perineal pull-through and anoplasty were performed at mean age sixth month for most patients with resection or tabularizing of the pouch.Fecal continence was(8o%) .

Conclusions: Erect abdominal X-ray have to be done routinely for every case of high type ARM in order not to miss any case of (C.P.C).It is better to excise the pouch completely whatever type was because histopathological exam of the pouch revealed abnormal muscle coat with failure to produce propulsive movement.

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Introduction:

Congenital pouch colon is a variant of anorectal malformation, 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 earliest report of(C.P.C) can be attributed to Spriggs in 1912 in London Hospital Museum who described a pathology specimen with imperforate anus and absence of left colon .(1) In 1977,Singh reported ten cases of(C.P.C) and described its anatomy in details.(2) About 90% of the cases were from India with few cases reported in Japan, China, Turkey ,Iran, Europe, Canada and U.S.A.(3). This malformation is sporadic in distribution with no evidence of familial inheritance or religious,or socioeconomic status difference.There is a suspension of environmental) pesticides (and dietary) vitamin B and Iodine deficiencies (factors to be related to the appearance of this malformation in these communities .There are many studies and theories suggesting the possible etiology for this deformity .Obliteration of inferior mesenteric artery early in fetal life is one of the proposed theories (1).Wakhlu et al have postulated that) C.P.C(represents a stage in the development of cloacal exstrophy and is the combined effect of the defective development of the splanchic layer of the caudal fold (4).Narasimharao et al had classified)C.P.C (.to four subtypes according to the length of normal colon proximal to the pouch.(5).Grossly the pouch has usually short and poorly developed mesentry,with illdefined or absence taenia coli,hausterations,and appendices epiploicae.(2) The pouch is supplied by a leash of vessels originating from branches of superior mesenteric artery.

Inferior mesenteric artery has been reported to be absent with type II and III)C.P.C. (3,5)(The pouch wall has normal ganglion cells though few have found reduced and very small ganglion cells (5).Mucosa is flattened and widened submucosa,while the muscle coat is not differentiated to inner)circular(and outer)longitudinal, (and the muscles are arranged in a decussating and arborhizing manner(6).

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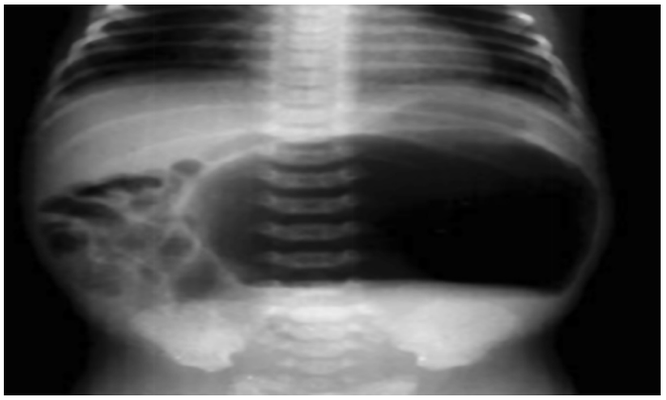
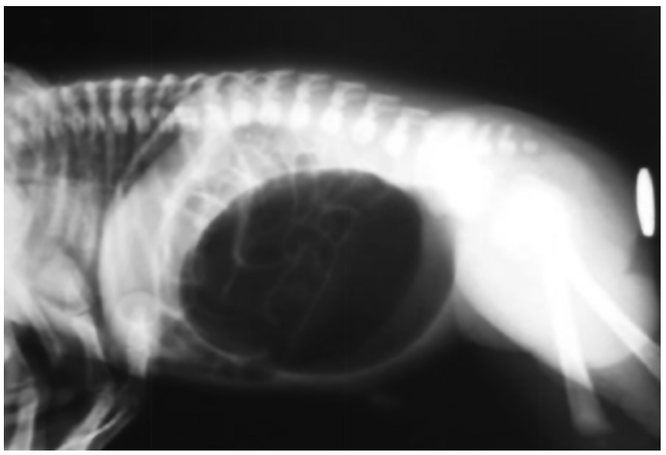
The majority of patients of) C.P.C (can be diagnosed by an erect plain abdomen with a typical feature of a single enormous air fluid level on the left side of abdomen occupying more than half of the width of abdomen(3,5,6).The primary procedure of management) C.P.C (is fistula ligation,coloplasty,and abdomino-trasanal endorectal pull-through and coloanal anastomosis(7,8).The voluntary musculature of the pelvis and perineum is usually well developed(9).

Diagram (1): Anatomic morphological classification of congenital pouch colon.

Type of CPCs	Classification
Type I	Normal colon absent and ileum open into the pouch colon.
Type II	Ileum open into normal cecum which opens into the pouch colon.
Type III	Normal ascending colon & transverse colon which open into the pouch colon.
Type IV	Normal colon with rectosigmoid pouch.



A single erect plain abdominal radiograph could be the most valuable single investigation to diagnose and classify) C.P.C(with preoperative plan to the surgical strategy without the necessity of cross table x-ray.



Cross- table x-ray with marker on anal dimple showing high anorectal malformation.

Patients and Methods:

A total number of seven cases of congenital pouch colon have been reported in AL-Emamain AL-Kadhemain Medical City and AL-Kadhemia Pediatric Hospital from October 2004 to October 2014. During the period of ten years, we have seen only seven cases of(C.P.C) associated with imperforate anus and managed thoroughly in pediatric surgery department in both hospitals. A standardized data was prepared for collection of complete information including ,the child age ,sex,body weight,perinatal history ,mother illness, drugs ingestion during pregnancy , exposure to radiation ,family occupation ,residence ,socio-economic state and family history of(ARM). .All the patients were from the peripheries of Baghdad .As a routine all patients with (I.A) were admitted to the pediatric surgery ward for full clinical examination and associated anomalies. Erect plain abdomen and invertogram were performed for all patients. Abdominal ultrasonic exam was performed to diagnose other associated anomalies in gastro-intestinal and genitourinary systems. (C.T) Scan and(M.R.I) to the abdomen were done for very few cases. Other investigations were also performed like cystourethrography,echocardiography .All the patients have been evaluated carefully and prepared for operation on the second or third day after replacement of fluid ,electrolytes and covered by broad spectrum antibiotics with nothing by mouth and nasogastric tube induction to decompress the bowel. The pouch attached to the colon was either completely resected (type II,III ,IV) and sent for histopathological exam or performed coloplasty of the pouch(type I) with end colostomy or ileostomy for subsequent pull through and anoplasty . Colo-vesical fistuletomy was performed for all patients .The patients have been categorized into four types according to modified Narasimaharo classification of C.P.C.

Results:

Congenital pouch colon accounted for 2.9% of all cases of high type of (I.A) and ano-rectal malformation (n;240). All patients who had (C.P.C) associated with (I.A) were from the peripheries of Baghdad (Radhwania,Abo-ghraib,Al-Taji and Yosofia) that are well known in rural .This phenomenon makes us suspect that there is a relationship between the appearance of these cases in these areas and the usage of pesticides in these agricultural districts.Prenetal history revealed presence of polyhydramnia in(5) patients(71%). Five patients were male and two were female with ratio 2.5:1 (male predominant). Six patients were full term with normal body weight except one was premature with low

birth weight. All patients had absent anal opening and hugely abdominal distention even the female patient due to absent fistula neither to vestibule nor to vagina (cloaca). Only one male (20%) passed meconium with urine. The two female patients had cloaca with single perineal opening one with colo-uterine fistula with vaginal agenesis, and the other one had blind rectum without fistula. Two patients presented in the first 24 hours after birth, while four of them within 48 hours and the seventh presented after 72 hours (unfortunately died second postoperative day due to sepsis) . Associated anomalies have been seen in(5)patients (71%) .Table (1)shows details.

Table (1) : Associated anomalies with (C.P.C)

Congenital anomalies	No. of cases	Percentage %
Hypospadias	3	42.8%
Right orchidism	3	42.8%
Bilateral vesico-ureteric reflux	2	28.5%
Renal agenesis	1	14%
Cyanotic congenital heart disease	2	28.5%
Absent appendix	1	14%

The first patient diagnosed(C.P.C) was male with (I.A) .During routine left lower oblique abdominal incision for sigmoid colostomy, we faced an unusual mass occupying most of the abdominal cavity. It was difficult to deliver the mass out ,so we was compelled to extend the incision down ward and transversely .A big pouch was noticed attached to a very short segment of colon(caecum,appendix and about five centimeters of the ascending colon) while other end of the pouch was connected to the bladder with a big fistula (colo-vesical fistula) .This was (type II)(C.P.C) according to Narasimaharo classification. Diagram (no.1) shows details.Fistulectomy ,coloplasty of the pouch with end colostomy was performed .Biopsies from the pouch were taken for histopathological exam .Abdomino-perineal pull through and(P.S.A.R.P.)had been performed when the patient reached 6th month old age. After appearance of this first case of(C.P.C.) we started to do erect plain abdomen routinely in addition to invertogram and cross table prone view for every case of(I.A) to exclude presence of associated(C.P.C) .Erect plain abdominal x-ray findings were highly suggestive for diagnosis of five cases(71%) of (C.P.C) especially type I,II,III by showing a single large air-fluid level in the left side of the abdomen occupying more than half of the abdominal width with deviation of the small bowel to the right side. During laparotomy two patients(28%) had type(I), three (34%) had type(II) while one patient(14%) had type(III) and the other one (14%) had type(IV).Table (2)shows details.

Table (2): Types of congenital pouch colon in our study according to Narasimaharo classification.

Type of C.P.C	No. of cases	Percentage %
Type I	2	28%
Type II	3	42%
Type III	1	14%
Type IV	1	14%
Total	7	100%

In all cases of type (I) and type(II) the inferior mesenteric artery was absent . The mesentery was very short. The only single branch from superior mesenteric artery ended in leash of vessels was supplying the pouch. The pouches as a whole were devoid from hausterations ,taenia coli and appendices epiploica.The wall of the pouches were thick in all patients except one patient with type(IV)(C.P.C) because the pouch was very dilated and ended blindly with a fibrous band to the vesical wall.Colovesical fistula was present in five patients(71%) except one patient(14%) the pouch was ended with a fibrous band to the vesical wall. Coloplasty with end colostomy were performed for (2)patients with type (I) while complete resection of the pouch and end colostomy were done for all patients with type (II,III,IV).Biopsies were taken from the pouch of five patients(71%) with(C.P.C) and sent for histopathological exam in our hospital and the results were normal mucosa and submucosa with presence of mature ganglion cells. The muscle coat was arranged in a decussating manner without differentiation of inner and outer muscle fibres. .Abdomino-perineal pull through and(P.S.A.R.P.) have been performed at mean age 5th month for the five cases of (C.P.C) after resection of the pouch in type(II) and (III) while pull through of the tabularizing coloplasty for two cases of type (I) .Follow up of the patients till now is continuous and four of them are continent except one patient who seems to have undeveloped muscle complex .Two patients(28.5%) died ,one in second postoperative day due to sepsis and the other one died after one month postoperatively due to sudden development of very severe jaundice leading to hepato-renal failure.

Table (3): Comparison of incidence of (C.P.C) types associated with(I.A) of our study with other authors.

Author	Type I	Type II	Type III	Type IV
Narasimaharo et al. (25) n=56	32	21	2	1
Wardhan et al. (41) n=18	8	10	-	-
Chadha et al. (4) n=41	21	11	7	2
Budhiraja et al. (3) n=27	8	7	3	9
Our study (7) n=240	2	3	1	1

Discussion:

The incidence of (C.P.C) associated with(I.A) in our study was(2.9%).It was very low in comparison to other studies especially in India .Kalawati Soran (6.5%)(13) ,Avtar Kishan (7%)(10), Devendera K .Gupta (10%)(11) R.K. Ghritlahary(11%)(12),Rajeendra K (13.71%)(14) . Male predominance was apparent which is like other authors(12),but differs from Sarin YK ,(M:F=1.6:1)(15). .Most of the patient were from peripheries of Baghdad which is famous in agriculture and widely using pesticides . There may be a relationship between pesticides and appearance of (C.P.C) in these instances. Most of our patients were full term with normal body weight. Associated anomalies were mainly in genito-urinary system .From our study there was no family history of (C.P.C) inheritance which was like Rajiv and Devenda(11). Preoperative radiological diagnosis of(C.P.C) was significant in (71%) through showing a left sided single large air-fluid level occupying more than (50%) of abdominal width with deviation of small bowel to the right side in most cases of C.P.C especially type I , II and III .Vivek Gharpure had considered it diagnostic in(89%)(17) Type (2) (C.P.C)(34%) was the most common in our study. It was like Wardhan study(18). Type(1) was encountered most common in Narasimaharo study (47%)(5). Each type (3) and(4) were (14%) which were again different from Narasimaharo (type(3)=3.5%)and (type (4)=1.8%). Budhiraga study obtained type (4)=33.5%.(16) .During laparotomy all male patients of type(1)and(2) (C.P.C) had colovesical fistula (71%) except one(14%) ended by a fibrous band with the vesical wall.Rajive had reported the incidence (92.7%)(19),while Deepuk (85%)(20) and ending by a fibrous band (15%).Inferior mesenteric artery was absent in all cases of type (1)and(2) of (C.P.C) which was like study of Devendra K.,while Rajive repoted absence of I.M.A from all patients with type(I) (C.P.C)(11,19) In our study the pouch was devoid from hausterations ,taenia coli and appendices epiploica .Rajive and Devendra also had the same observation as we have seen.(11,19)After abdomino-perineal pull through of the five cases of (C.P.C),four of them(80%) were fecal continent which was like Rajive(19). Two cases type (I) developed constipation for a long time and needed frequent ano-rectal dilation which means that pull through of the tubularized colon (pouch) in definitive operation is no longer rewarded because of failure of propulsive movement of the pulled tubularized pouch segment due to abnormal muscle coat pattern. High incidence of redilatation had been seen by other authors also Rajive (78%) ,Deepak(28.5%) ,Avater(20%) and Wakhalu (11%). (10,11,19,,21).

Conclusions:

Although the incidence of (C.P.C.) associated with imperforate anus is low (2.9%) in our hospitals ,there may be other centers have seen or under estimated cases of (C.P.C) .Erect plain abdominal X-ray is diagnostic for most cases of (C.P.C) especially type (I) and (II), so it must be done routinely for every case of imperforate anus.Because of the abnormal histopathological pattern of the muscle coat and improper propulsive movement of the pouch after pull-through of the tubularized pouch ,it is better to excise the pouch completely with performing ileo- anal anastomosis.

Authors contributions:

Dr.Ahmed Zubar Zain had two cases of C.P.C with complete details .

Dr.Raghad Jaleel Abolhab had one case of C.P.C with an efficient informations and they collected with me the data about our research .

Iam the correspondence dr. Salah Salman Mahmood .It was my opinion to do this research and I had four cases of C.P.C.

List of abbreviations

(C.P.C) = (congenital pouch colon)

(I.A) =(imperforate anus)

(P.S.A.R.T)=(posterior sagittal anorectoplasty)

ARM: aneorectal malformation

References:

1.Spriggs NJ.Congenital occlusion of the gastrointestinal tract.Guys Hospital.Rep 1912;766:143.

2.Singh A,Singh R,Singh A.Short colon malformation with imperforate anus.Acta Paediatr Scand 1977;66:589_94

3.Chadha R ,Mahajan JK.The embryology and management of congenital pouch colon associated with anorectal agenesis.J Pediatric surg.1994;29:439_46.
4.Wakhl a R(1982) Short colon with anorectal malformation . AK, Tandon RK ,Kalr .Indian J.surg.1982; 44:621_9.
5. Narasimharao KL, Yadav K, Mitra SK, Pathak K IC. Congenital short colon with imperforate anus (pouch colon syndrome).Ann Pediatr Surg 1984;1:159_67
6. . Dickinson SJ .Agenesis of the descending colon with imperforate anus.Am J Surg 1967;113:279_81
7.Sunita Singh, Intezar Ahmed, Ashish Wakhlu. Newer variant of congenital pouch colon with rectal agenesis: management strategy and review of the literature. BMG case report .2011,363. (IVSL)
8.Parelkar S , Oak S, Mishra PK,et al.Congenital pouch colon with rectal atresia; a case report.J Pediatr Surg 2010; 45:639-41.
9.Wakhalu A K. Congenital pouch colon.In:Gupta DK,editor. Textbook of neonatal surgery. New Delhi: Modern Publishers; 2000. P. 240-8.
10.Wakhlu AK, Wakhlu A,Pandey A,Agarwal R,Tandon RK,Kureel SN.Cogenital short colon.World J Surgery.1996;20:107_14
11.Gupta DK, Sharma S .Congenital pouch colon –Then and now, A plea for common agreement. J Indian Assoc Ped Surg 2007;12:110-129.
12.Ghritlaharey R K, Budhwani K S, Shirvastava D K, Gupta G. Experience with 40 cases of congenital pouch colon. J Indian Assoc.Ped. Surg, 2007;12:13-63.
13.Gupta DK, Sharma S. Congenital pouch colon.In: Anorectal Malformations, 1st ed, Hutson J, Holschneider A, editor. Spriger Heidelburg; 2006. Chapter 11: p.211-22.
14.Rajeendra K, Ghritlahary,K S
. Budhwani.Two staged management of all types of congenital pouch colon.African J Ped.Surg 2013;10:1; page 17-23.
15.Sarin YK, Nagdeve N G, Sengarm. Congenital pouch colon in female subject. J Indian Assoc.Ped Surg 2007;12:17-21. (IVSL)
16.Budhiraja S ,Pandit SK, Rattan KN. A report of 27 cases of congenital short colon with an imperforate anus: so –called . pouch colon syndrome,.Trop Doct 1997;217-20.
17.Vivek Gharpure: Our experience in congenital pouch colon.J Indian Assoc.Pedia Surg 2007;12:22-5.
18.Wardhan H, Gangopadhyay AN, Singhal GD,Gopal SC,(1990) Imperforate anus with congenital short colon(pouch colon syndrome).Pediatr Surg Int 5:124-6.
19.Chadha R.Congenital pouch colon revisited. Pediatric Surg Int.2004;20:393_401.
20.Singh S, Pathak I C. Short colon Malformation with imperforate anus.Surgery 1992; 71:781-6.
21.Chadha R, Bagga D, Gupta S,Prasad A. Congenital pouch colon:massive redilatation of the tubularized colonic pouch after pull-through surgery.J .Pedia Surg 2002;37:1376-9.