Congenital Lower Intestinal Tract Anomalies :Review of Surgical Management of 50 Iraqi Patients in Al –Najaf City

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ABSTRACT:

BACKGROUND:

Congenital lower intestinal tract anomalies are significant cause of morbidity and mortality in children, classified in two groups, major one is anorectal malformation, other one Hirchsprung's disease, usually associated with other anomalies and should be treated as early as possible.

OBJECTIVE:

To show the prevalence and our experience with congenital lower intestinal tract anomalies in AL-Najaf city.

PATIENTS AND METHODS:

A total 50 patients had various types of congenital lower intestinal tract anomalies , 22 of them high types (anorectal) treated by 3 stages operations(preliminary colostomy, nearly new definitive surgery infracoccygeal approach and closure colostomy), minor surgery for low types (12) and for Hirchsprung's disease treated by usual pull through operation(Swenson's).

RESULTS:

The anorectal anomalies (A.R.M) are the most common anomalies of lower intestinal tract , high type cases were 25 , low type were 12 cases and Hirchsprung's disease were13 cases . Male were 30 cases and female were 20 cases , age of presentation between first day of life and 18 years of life .

CONCLUSION:

Infracoccygeal approach was the safe and non complicated operation in dealing with high type of anorectal malformation and the good results obtained when the operation was done early period of life (around one year).

KEY WORDS: anorectal malformation, infracoccygeal approach, , Hirchsprung's disease.

INTRODUCTION:

Congenital lower intestinal tract anomalies (C.L.I.T.A) are significant cause of morbidity and mortality in children; these abnormalities include developmental obstructive defects of the anorectal region and colonic aganglionosis of large bowel. [1,2]

Anorectal Malformations (A.R.M) are one of the most common large bowel birth defect [2], with this defect, the anus and rectum do not develop properly, which occur during fifth to seventh week of fetal development, (A.R.M) affect 1 in 5,000 babies and is slightly more common in males[3,4] most of the time, the cause is unknown, in some cases environmental factors or drug exposure during pregnancy may play a role, but this is still unclear.

Up to one third of children who have genetic syndromes and chromosomal abnormalities, or other congenital defects like, VACTERL association (a syndrome in which there are Vertebral, Anal, Cardiac, Tracheal, Esophageal, Renal and Limb abnormalities) [5].

Associated congenital anomalies occurred in 50-60% of the infants with A.R.M. Urological defects are the commonest anomalies associated with ARM which presents with a wide spectrum of defects, ranging from relatively simple malformations to very complex cloacal anomalies (a cloaca is a confluence of the rectum ,vagina and urethra which open into a single common cavity) occur 1 per 20000 live births .It's occurred exclusively in girls . When a baby is born, the pediatrician performs a thorough physical examination that includes the anal examination [6,7].

Low and high type depend on relation to levator ani muscle of lower rectal end, if above it's high type, if below it's low type(diagnosed during surgery) and preoperatively by lateral abdominal invert gram (upside down) with metal marker on anal dimple for one minute 16 hrs after birth if the distance >2.5cm is high type , also to know if there are abnormalities of

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the spine and sacrum, other diagnostic tests , abdominal ultrasound and intravenous urography, to show the urinary tract abnormalities , echocardiogram, this test is performed to determine if there are heart defects and magnatic resonance imagining -- In selected cases, is necessary to make a definitive diagnosis of tethered cord or other spinal abnormalities. It is also used to define the anatomy of pelvic muscles and structures, the anal sphincter is usually not well developed in high type of A.R.M. [8]
The type, number and time of operations necessary depends on the type of abnormality . Home treatment including repeated anal dilatation and high fiber diet. [9,10]

- In low abnormalities(membranous,stenosed,ectopic and covered anus) , stenosed one means small anal orifice, covered anus means there is bridge of skin cover, treatment usually , including minor surgery under local anesthesia,followed by repeated anal dilatation by the family for 3-6 months, but for high abnormalities(anorectal agenesis, rectal atresia and cloaca) present a very difficult problem, the definitive surgery at body weight around 8-10 kg(8 months and above) and each case must be considered on it merits , the possibilities are, [11,12].
- laparotomy, division of rectourethral fistula and transverse colostomy .
- laparotomy, division of fistula and pull-through operation in one stage .
- Division of fistula and rectal pull-down operation through the perineum.
- Presacral (infracoccygeal approach).
- Perminant Colostomy for cloacal variety.
- Concerning Hirchsprung'sdisease(aganglionic megacolon) is a birth defect in which ganglion cells in the wall of the large intestine do not develop. This clinical condition usually becomes apparent shortly after birth when the infant passes very little meconium and the abdomen is distended. In older patients, severe constipation and recurrent fecal impactions are more common. The defect occurs in approximately one of each 5000 live births and is familial. It is much more common in males (male to female ratio 2:1). There is an 8% association with Down's syndrome and a variety of other anomalies. It accounts for 20 to 25 percent of the cases of neonatal bowel obstruction. Most often rectosigmoid and sigmoid are involved. [13]

Clinical features of Hirchsprung's disease vary depending on the child's age[14]

1- In a newborn a delay in passing the first stool (meconium)
2- In an infant Constipation and distented abdomen.
3- In a child Constipation and poor growth with attacks of diarrhea enterocolitis(non bacterial, non viral).

To confirm the diagnosis, we need rectal biopsy and barium enema, rectal biopsy either suction biopsy include mucosa and submucosa by special capsule or full thickness biopsy from posterior wall of rectum under general anesthesia include mucosa and muscularis mucosa to show absence of ganglion with hypertrophy of nerve fiber and histochemistry staining for cholinesterase . Barium enema is normal during neonate, so, postponed after neonatal period to show contracted (aganglionic segment) and transitional dilated normal segment, their types are { long , short , ultrashort (ring type) and total colon type} which diagnosed during surgery, rarely Hirchsprung's disease involve small intestine. [15]
In most cases, surgery is done shortly after birth. Two or three stages surgery usually done, depend on the age of patient , the length of the involved segment , severity of symptoms and the present of enterocolitis, after initial transverse loop colostomy for virable period, the choice of definitive surgical procedure as follow:

1- Ultrashort or short segment disease may respond to an extended myectomy.
2- Long or total segment disease treated by one of the 4 operations below, the definitive operations are:

- Swenson's operation (pull through operation) .
- Soave's procedure(mucosectomy).
- Duhamel operation(retrorectal space coloanal anastomosis).
- Ileo-anal anastomosis for total aganglionosis.

The definitive operation is delayed when the baby received and treated at neonatal period until the child weight approximately 8-10 kg when the pelvis is still shallow but wide enough to give good access, but when received at older age group, the definitive procedure postponed for 3-6 months to allow the distended hypertrophied segment return to normal.[16,17,18]
Major complications from surgery include anastomotic leaks, scar tissue formation (stricture) and Ischemic enterocolitis. [19,20]

PATIENTS AND METHODS:

This is a prospective study has been carried out between Nov. 2001 to May 2008, it included (50) cases of C.L.I.T.A .

A detailed clinical information were taken from each patient, including short antenatal and postnatal history, clinical presentation and physical examination (including general and regional examination for the primary defect as well as associated anomalies). This study includes 37 children presenting with ARM and 13 children with Hirchsprung's disease, all treated by corrective surgery (definitive).

Data were collected from public and private in AL-Najaf city . All cases with C.L.I.T.A had routine investigations including hemoglobin level, white blood cell count, general urine examination, urine culture and random blood sugar. Invert gram abdominal x-ray and ultrasonography (U.S) were done for all patients at different times of presentation, intravenous urography and barium enema were done for some patients accordingly, all cases of Hirchsprung's disease were diagnosed by full thickness rectal biopsy under general anesthesia .

Even those presented to an emergency unit the operation postponed next day for investigation and resuscitation.

For low type A.R.M (12) patients were treated as follow:

In case of membranous anus treated under local anesthesia in outpatient clinic by crucuiate incision, for ectopic anus under general anesthesia by plastic cut back operation with mobilization of lower end posteriorly and suturing by 2/0 rounded needle silk suture, the above 2 types followed by repeated daily anal dilatation at home for about 3-6 months with good results under supervision by the same surgeon, for stenosed anus only treated by repeated daily anal dilatation at home.

The remaining 38 patients:

3 patient were referred to our department with temporary sigmoid colostomy, the other 35 cases(both high type and Hirchsprung's disease) treated by temporary transverse loop colostomy near hepatic flexure through right upper transverse supraumbilical incision, preoperative preparation including nil by mouth, intravenous fluid and antibiotic cover 24 hours before by Cefotaxime 50 mg\kg and metronidazole 21 mg\kg intravenously, (chemical preparation) , postoperatively we advised and taught the family how to take care about colostomy.

The definitive surgery was done at the age of 10 months and above with chemical preparation as above but mechanical preparation by manual faecal evacuation and normal saline bowel washing through colostomy opening, during surgery (22) cases of them were treated by nearly new infracoccygeal (presacral) approach

The other 3 cases were started with presacral approach as above, but we didn't found the rectal pouch , then an immediate explorative lower midline laparotomy done, the finding as follows:

The first case was a male patient with 3 previous operations for removal of vesical stones only as family said, but I found multiple vesical stones, whitish (faeces and barium in nature) with wide rectovesical fistula, I did separation and manipulation of rectal end, removal of stones, closure of bladder and closed the abdomen with drain, then continue as above in presacral approach.

The second case was female, I found rectouterine fistula and uterovesical fistula with no external urethral orifice, so introduce a catheter through vagina, separation done for rectouterine fistula, I found very small uterovesical fistula, so withdrawn the Foley's catheter through the uterus into bladder, then continue as above in presacral approach.

The third case was a female with no evidence of bladder and uterus; I found a common single cavity (cloaca), so left with permanent colostomy.

These cases stayed for some days postoperatively in hospital according to their stages of operation, after discharge, I advised the family to do 2 -3 times daily frequent anal dilatation by Hegar dilator with gradual increasing size under my supervision (after second stage).

For Hirchsprung's disease(13) cases, pull through operation (Swenson) by a lower midline laparotomy incision at about 10 months of age and in addition dulcolox suppositories 24-48 hours before and bowel preparation before surgery did as above according to their presentation and initial treatment

The above 38 cases (high type and Hirchsprung's disease) followed by repeated anal dilatation for about 3-6 months.

1-2 months later after definitive surgery of the above cases closure colostomy by double layers were done, and continue by regular doctor checking and proper bowel toilet and education to make accepted bowel control .They continued daily anal dilatation to complete their period of dilatation as mention above.

RESULT:

The most common C.L.I.T.A. are the A.R.M. which are 37 and the Hirchsprung's disease are 13 and more common in female as shown in table 1.

Table1: Show the classification and sex distribution C.L.I.T.A. of studied group.

Type of anomaly	male	female	total	%
A.R.M	22	15	37	74
Hirchsprung's disease	8	5	13	26

C.L.I.T.A. most commonly presented during first 3 days of about 66% of cases and presented as abdominal distension as shown in table 2.

Table 2: Show age distribution &type of C.L.I.T.A. presentation.

age of presentation	No.	%	Clinical presentation
(1-3) days	33	66	Abdominal distension with no or little meconium passage
4 days-1year	9	18	Difficulty in passing motion , repeated vomiting, failure to thrive with badly smelled urine in addition to abdominal distension
(1-6) years	5	10	repeated vomiting , abdominal distension with history of recurrent U.T.I
6 years and more	3	6	Abdominal distension with chronic constipation and attacks of diarrhea
total	50	100	

Most cases (22 out of 25) of A.R.M. (high type) were treated by safe and uncomplicated infracoccygeal approach , and only 3 by laparotomy as shown in table 3.

Table 3 :Show type of surgical treatment of studied patients

surgical approach	Male	Female	total	%
Infracoccygeal	15	7	22	88
laparotomy	1	2	3	12
total	16	9	25	100

Table 4: Show sub classification of C.L.I.T.A. of studied patients

High type	No.	%	Low type	No.	%
Anorectal agenesis	20	80	membranous	5	41.5
Rectal atrasia	4	16	ectopic	5	41.5
cloaca	1	4	stenosed	2	17
total	25	100	total	12	100

The most common form of A.R.M.(high type) are anorectal agenesis 20 cases and least was cloaca as shown in table 4.

Table 5 :Show radiological finding of Hirchsprungs diseased patients

Type of cases	No.	%
Short segment	10	77
Ultra short segment	2	15.5
Long segment	1	7.5
total	13	100

Short segment type of Hirchsprung's disease demonstrated by barium enema was the commonest type (10 out 13)of Hirchsprung's disease of studied group.

Table 6: Show associated anomalies with C.L.I.T.A. of studied patients.

Type of anomaly	associated anomalies	No.	%
High type	Rectourethral	15	30
	rectovaginal fistula		
	uterovesical fistula	2	4
	rectouterine fistula		
	Rectovesical fistula	1	2
	Limb anomaly		
	Down syndrome	1	2
	Right unilateral hypertrophy of labia majora	1	2
		1	2
		1	2
		2	4

The most common associated anomalies of A.R.M. (high type) was rectourethral fistula 15 casses as shown in table 6.

Table 7: Show complications of C.L.I.T.A. surgery.

Complication	No.	%
Temporary Perianal skin eruption and little mucous and faecal incontinence	50	100
Colostomy prolapsed	12	31.5
Post colostomy incisional hernia	2	.25
Parastomal hernia	3	9.3
Anal stenosis	4	12.4
Segmental stenosis for Hirchsprung's disease	2	15.3
Laparotomy wound infection	9	28.1
Presacral wound infection	2	21.8
Abdominal incisional hernia	1	3.1
Rectocutaneous fistula	1	3.1
Vaginocutanous fistula	1	3.1

The major complication of C.L.I.T.A. was temporary perianal skin eruption and little mucus and faecal incontinence especially of A.R.M. (high type) , all cases were affected as shown in table7.

DISCUSSION:

Most of our cases were presented by the first 3 days of life especially for those were delivered in hospitals because they received surgical advices from doctors and nursing staff but the other cases were presented later because of low family education, presentation of associated fistula and midwives delivery. I found that majority of cases of C.L.I.T.A. presented early 1-3 days of life which similar to the study in 1996 by Wakhlu AK, [1]

I found that most common type of Hirchsprung's disease were short segment which unlike the study done in 1991 by chatterjee . [21] This study gives an outline of a profile of the ARM and Hirchsprung's disease patients in a center of Al-Najaf .Many systems of classification have been suggested , however, the terms high and low have been broadly recognized and applied worldwide, the most common associated lesion were recto urethral fistula in males which similar to other study in Oxford University, 1991 by Chatterjee SK.[21] Our cases of A.R.M. of high type were nearly similar to other study done in Chicago 1986 by Templeton JM,O'Neil JA jr.[22]

I found that majority of our patients had one or more associated malformations which similar to other study done by Hassink EA, 1996. [6]

The main approach (infra-coccygeal) was done for majority of my cases (high type), which unlike that mentioned by (Norman S. Wiliams, 2004) [13]where laparotomy was the main approach butsimilar to study done in Indianby Pathak IC, 1996. [9] and arlins, RW.[14]

In 3 cases presented to our Najaf center with sigmoid loop colostomy were done by other surgeons, so I found difficulties during definitive surgery in dealing with affected loop, 2 cases of them were of Hirchsprung's disease but the third case of A.R.M(high type) regarding suturing of rectal pouch without tension, so I change a sigmoid loop colostomy into transverse loop colostomy as in my cases.

Most common type of complications were perianal skin eruption which unlike the study in 1975 by Swenson O, et al…., were segmental stenosis, and in study in 1986 by Teich S, et al….., were ischemic enterocolitis. [19,20]

Previously sigmoid loop colostomy where mainly done, but nowadays mainly transverse loop colostomy near hepatic flexture to decrease incidence of prolapse, and a way from operative site and the diseased area especially in Hirchsprung's disease.[15]

CONCLUSION:

Transverse loop colostomy near hepatic flexure is better than sigmoid loop colostomy regarding to prolapsed, during definitive surgery, laparotomy incision and length of affected segment in case of Hirchsprung's disease. Swenson's operation regarded as good procedure where end with good results as shown by my study.

I recommend to obtain pediatric operative theatre with proper temperature and humidity control, trained anesthetic staff for pediatric age group, pediatric surgical and anesthetic instruments, incubators. expert pediatric nursing staff for pre and postoperative follow up, pediatric intravenous fluid sets, colostomy bag of pediatric age group and a frozen biopsy procedure to asses the proper levels of resection in case of Hirchsprung's disease during surgery . All these to obtain near normal bowel control and to prevent and treat complication as early as possible.

I recommend that all C.L.I.T.A. cases should be referred to pediatric surgeon or a general surgeon with experience in pediatric surgery to obtain best result and less complication

REFRENCES:

1. Chadha R, Bagga D, Malhotra CJ, Mohta A, Dhar A, Kumar A. The embryology and management of congenital pouch colon associated with anorectal agenesis. J Pediatr Surg 1994;29,439-46.
2. Gangopadhyay AN, Shilpa S, Mohan TV, Gopal SC. Single-stage management of all pouch colon (anorectal malformation) in newborns. J Pediatr Surg 2005;40,1151-5.
3. Rao KL, Menon P. Congenital pouch colon associated with anorectal agenesis (pouch colon syndrome). Pediatr Surg Int 2005;21,125-6.
4. Kumar A, Agarwala S, Srinivas M, Bajpai M, Bhatnagar V, Gupta DK, et al . Anorectal malformations and their impact on survival. Indian J Pediatr 2005;72,1039-42
5. Metts JC 3rd, Kothin L, Kasper S, Shyr Y, Adams MC, Brock JW 3rd.Genital malformations and coexistent urinary tract and spinal anomalies in patients with imperforate anus. J Urol 1997;158,1298-300.
6. Hassink EA, Rieu PN, Hamal BC, Severijnen RS, vd Staak FH, Festen C. Additional congenital defects in Anorectal malformations. Eur J Pediatr 1996;155,477-82.
7. Gupta DK, Charles AR, Srinavas M. Pediatric Surgery in India- A specialty come of age. Pediatr Surg Int 2002;18,649-52.
8. Saaull DB, HARRISON EA. Classification of Anorectal malformations –Initial approach ,diagnostic tests and colostomy. Semin Pediatr Surg 1997;16,187-95.
9. Pathak IC, Saifullah S.Congenital Anorectal malformations: An experience based on 50cases. Indian J Pediatr 996;36,370-9.
10. Sharma S, Gupta DK, Bhatnagar V, Bajpai M, Agarwala S. Management of congenital pouch colon in association with ARM. J Indian Assoc Pediatr Surg 2005;10,S-22.
11. Pena A. Anorectal Malformations. Semin Pediatr Surg 1995;4,35-47.
12. Pena A, Devries PA. Posterior sagittal anorectoplasty: Important technical considerations and new applications. J Pediatr Surg 1982;17,796-881.
13. Norman S. Wiliams,2004 ;chapter 76 ,1247 .
14. arlins, RW. In 2000 ;chapter:79,367, pediatric text book of surgery .
15. Harrison MW, Diets DM, et al: Diagnosis and management of Hirschsprung's disease .Am J Surg 1986;152,44.
16. Martin LW: Surgical management of total colonic aganglionsis. Ann Surg 1972;176,343.
17. Polley TZ, Coran AG, et al : A ten year experience with ninty-two cases of Hirschsprung's disease. Ann Surg 1985;202,349 .
18. Soave F: Hirschsprung's disease: A new surgical technique. Arch Dis Child 1964;39,116.
19. Swenson O, Sherman JO, et al: Treatment and postoperative complications of congenital megacolon: A 25 year follow- up. Ann Surg 1975;182,266.
20. Teich S, Schisgall RM, et al: Ischemic enterocolitis as a complication of Hirschsprung's disease. J Pediatr Surg 1986;21,143.
21. Chatterjee SK. Anorectal malformations. A Surgeon's experience. Oxford University Press: Delhi; 1991.
22. Templeton JM, O'Neil JA Jr. Anorectal malformations. *In* : Welch KJ, Randolph JG, Ravitch M (editors). Pediatric Surgery. 2nd Year Book Medical: Chicago; 1986,1022-37.