

MANAGEMENT OF ANORECTAL MALFORMATION

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Received date: 26 April 2023

Revised date: 16 May 2023

Accepted date: 06 June 2023

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ABSTRACT

Background: The treatment of anorectal abnormalities is difficult and extremely complicated. Careful preoperative and surgical planning must be carried out in order to produce the desired results in children with this diagnosis. Additionally, bowel control and lifelong care are important for patient success.

Objective: To study the management of imperforate anus and the proper surgical procedure for each type.

Patients and Methods: A prospective study of 80 case of imperforate anus. Setting: All of our patients managed at Al-Khansaa Hospital between September 2010 and September 2012. **Results:** Prenatal history shows positive polyhydramnios in 27.5%. Fifty one percent presented with acute intestinal obstruction in the first week of life. Of these 80 cases 47.5% were high lesion, 18.7% intermediate lesion and 30.0% low lesion. Thirty one percent of male lesions were recto urethral fistula as the commonest male lesion while recto vestibular fistula represents the commonest female lesion (12.5%). Forty percent has associated anomalies involving different systems. Cross table radiograph used to determine the level of the lesion shows 80.0% true results, while the remaining 20.0% failed to predict the level of the lesion. Distal colostogram used for identification of urinary fistula and it was true positive in 40.0% while in the remaining 60.0% fistulae were detected per operatively. Low lesions managed by one stage perineal approach using cruciateanoplasty in 14 patients, anal transposition in 7 patients and cut back operation in 2 patients, five patients with anal agenesis successfully treated by one stage cruciateanoplasty. Three stage operations using posterior sagittal anorectoplasty were the standard technique for the remaining lesions in 92.5% and in the remaining 7.5% an abdominal approach was added. **Conclusions:** High lesion carries the highest incidence of associated anomalies which is responsible for majority of the deaths, and genitor-urinary anomalies are the commonest. Cross table radiograph and distal colostogram carry a significant percent of false results. Low lesions and anal agenesis can be managed through perineal approach without colostomy. Posterior sagittal anorectoplasty is the optimal surgical procedure for high lesions and rectovestibular fistula.

KEYWORDS: Anorectal Malformation, Anorectoplasty, Colostomy.

INTRODUCTION

Malformation of anogenital region have always aroused feeling of horror, this is well expressed on a stone slab from the library of King Assurbanipal of Nineveh.

The anal canal is surrounded by intertwining involuntary and voluntary sphincters that provide a fine degree of control of the passage of flatus and stool. The internal sphincter is an extension of the inner circular layer of the bowel wall; it extends just beneath the mucosa of the bowel distal to the pectinate line. This muscle is the most important factor in anorectal resistance to defecation.

The internal sphincter relaxes in response to rectal distension to initiate the act of defecation.^[1,2]

In 1970 an international classification was suggested by an Ad-Hoc committee with its modification into: low (trans levator) type, intermediate type, high (supra levator) type and miscellaneous, with its sub-classification.^[3]

Stephens has proposed complete and detailed classification, although very accurate, it has proved difficult for other surgeons to use. Ladd and Gross

classification has been used by some surgeons. The key point for this classification is the distinction made between those lesions that have descended below the puborectalis muscle and those that have stopped above this muscle.^[4] A new international classification lists different types of anorectal malformation. In high anomalies the bowel terminates above the pelvic floor and there is often an associated fistula into the urethra in male and into vagina in female. In low anomalies the bowel traverses the pelvic floor but fails to develop a normal anus. In male there is often a narrow fistula in the perineum or scrotum leading back to a normally sited covered anus. In females an opening below the hymen confirm the presence of low anomalies. Anomalies that do not fit into either of these groups are termed "intermediate anomalies" and are usually best managed as high lesions.^[5]

Imperforate anus occurred in 1 in 5000 to 1 in 20000 live births, affecting males and females almost equally.^[6] Fistulas were present in 75.0% of the cases 85.0% of the female and 66.0% of the male has such connection. A familial history is rare and apparently there is no racial predilection.^[7]

In most of these anomalies, the born baby presents with intestinal obstruction.^[8] Failure to pull the buttocks a part in the examination of the newborn baby often causes anorectal anomalies to be missed. Most of the low type malformation can be completely diagnosed with certainty by simple inspection.

Anal stenosis can be recognized by heaping up of the epithelium around the normal anal area but by careful probing an opening can be found. Membranous imperforate anus diagnosed when a thin membrane is seen to block the passage of meconium^[4], so the diagnosis is not difficult. The location of the fistula site may be a problem. Beading of mucus or meconium along the median raphee of perineum and scrotum denotes a low imperforate anus in the male. Voiding of gas or meconium per urethra suggest urinary fistula of the high imperforate anus in the male. In the female, careful examination of the perineum, forchette and vestibule of the vagina will locate the fistula in most cases. A high fistula in the female may terminate in the vagina and it will be harder to demonstrate. If single perineal opening is seen in the female a cloacal abnormalities is present vagina and rectum opening into a common urogenital sinus.^[8, 9]

There are many intermediate cases, where the anus is located slightly forward of its position that are unrecognized and cause constipation in both males and females they are often referred with a diagnosis of a suspected Hirschsprung's disease.^[9]

Cross table lateral radiograph which done by placing a roll beneath the hips of the infant to elevate the buttocks and allow air to migrate superiorly to the end of the

rectum. In boys retrograde urethrography demonstrated the fistula and its location in two thirds of the cases with a doubtful clinical examination.^[10]

Some cases of anorectal atresia can be suspected on prenatal sonograms' by demonstration of dilated colon. Distinguishing anorectal atresia from other causes of fetal bowel dilatation is important because of the frequency of concurrent anomalies associated with anorectal atresia.^[11] High frequency real time ultrasound provides a non invasive method of accurately determining the location of the distal rectal pouch and readily detects associated renal anomalies knowledge that the pouch perineum distance is 1.5cm is useful in identifying a lesion terminating below the bases of the bladder as "low" and conversely a pouch below the base of the bladder and 1.5cm or more from the perineal surface should be considered "high" until proved otherwise.^[12] CT scan and MRI recently has been suggested as an effective tool in the evaluation of patients with imperforate anus for either primary or secondary operation with poor results and predicting the outcome by providing information about the pelvic musculature, especially in secondary cases.^[13, 14]

A satisfactory result of any operation for the correction of anorectal "anomalies" were depending on the achievement of an efficient continence mechanism. This indicates the presence of the afferent impulses from intact sensory receptors, efferent nerve pathway from higher and lower centers to the muscles concerned with continence and defecation, and adequate muscular system including puborectalis sling, an internal sphincter and the external sphincter.^[15, 16] For the low lesions in male, a perineal anoplasty is performed in boys when there is definite evidence of a fistulous tract either at the anal dimple or forward in the perineum or if there is a bulging anal membrane, anal stenosis simply treated by anal dilatation to produce an adequate sized anus, but severe anal stenosis is opened by a V-Y plasty while anal membrane is opened with cruciate incision with skin flaps sutured up into the anal canal^[1,4,5] While in females, the minimal degree of anterior displacement of the anus in girls requires no treatment if the 5th finger can be inserted into the anal canal more anterior displacement or anal stenosis can be treated with a V-Y operation.^[1,7] Sometimes simple "cut back" of posterior sagittal anoplasty may be required to repair this defect with a good functional result but cosmetically less desirable. Dilatation required afterward.^[1, 2] Anovestibular fistula is too far forward and too close to the vagina for cut back operation and it is essential to distinguish it from rectovestibular fistula, which requires colostomy and later repair.

Intermediate and high lesions were cured with a perineal anoplasty during the neonatal period in lithotomy position with urethral catheter in male and nerve stimulator to identify external sphincter muscle. If the pouch is not found within 1.5cm either the diagnosis or

the dissection is wrong.^[1, 4] The remaining types and the cloacal malformation were treated by colostomy performed in the neonatal period and the definitive operation delayed until the patient is about 6-12 months of age or sometime even less than that" at 4-8 weeks of life".^[1,2,5,17]

Imperforate anus surgery is not only concerned with survival but also with the problem of incontinence. The ideal long term result must be fecal continence without constipation or fecal soiling. This should be achieved before the child starts school.^[1, 3, 18] Many approaches are used for repairing an intermediate and high type of imperforate anus but mainly anterior permeal operation, sacro perineal approach, endorectal pull through, anterior Abdomino-perineal approach, posterior sagittal anorectoplasty (PSARP).

Aim of the study

To study the imperforate anus, its epidemiology and mode of presentation with special emphases on the predisposing factors, associated anomalies and its effect on outcome, role of radiology in determining the level of the lesion and fistula in comparison to operative findings, and the appropriate surgical technique for each type with special concern on the role and short term results of the posterior sagittal anorectoplasty.

PATIENTS AND METHODS

A prospective study of 80 patients with anorectal malformations admitted to pediatric surgery unit in AL-Khansaa Hospital from September 2010 to the end of September 2012. Most of these cases came from Nineveh, the rest were referred to from different areas in the north of Iraq.

Some cases presented with neonatal intestinal obstruction and were operated upon urgent operations, the rest were referred to the Outpatient Department complaining from chronic constipation, straining on defecation or passage of stool in abnormal opening noted by the parents or incidentally by the doctor.

A special data collecting form has been used including: name, age, sex, weight, gestational age, residency, family history, prenatal history, presentation, investigations, associated anomalies, type of lesion, operation, operative findings, postoperative complications, secondary operations, follow up and death.

Diagnosis was based on history and proper clinical examination aided by radiological examination and urinalysis. Ultrasound examination was used mainly for the diagnosis of some associated anomalies. Other investigations were done accordingly.

The patients were classified according to the international classification into low, intermediate, high, cloaca (in female patients) and rare malformation.

Associated anomalies involving other systems were diagnosed and assessed during the period of follow up.

The presence of fistulous connection was searched clinically and radiologically using distal colostogram, and the presence of these fistulae especially to urinary system was compared with the operative findings.

Management of low lesion, all male intermediate lesions and perineal canal (anovestibular) were done through one stage perineal approach. Female intermediate lesions, high and cloaca managed through staged operation (colostomy-definitive pull through surgery-closure colostomy). Pull through operations used were mainly posterior sagittal anorectoplasty and in some patients through an abdominoperineal approach. Preoperative assessment for condition of anal dimple, external sphincter and puborectalis muscle was done. The distal pouch sutured to the site of proposed anus by absorbable suture material, drain were used. Post operative complication are identified and treated accordingly.

RESULTS

The sample consisted of 80 patients; 56(70.0%) were males and 24(30.0%) were females. Full term patients were 76 (95.0%) and only 4 patients (5.0%) were pre term. The presenting age ranging from 1 day to 2 years old, according to that 2 groups have been identified. Positive family History was found in only five patients (6.25%).

Polyhydramnios was positive in mothers of 22 patients (27.5%), history of drugs ingestion by mothers in the first trimester were positive in 9 patients (11.2%). Infections were present in mothers of 8 patients (10.0%). Radiation was positive in one patient (1.5%).

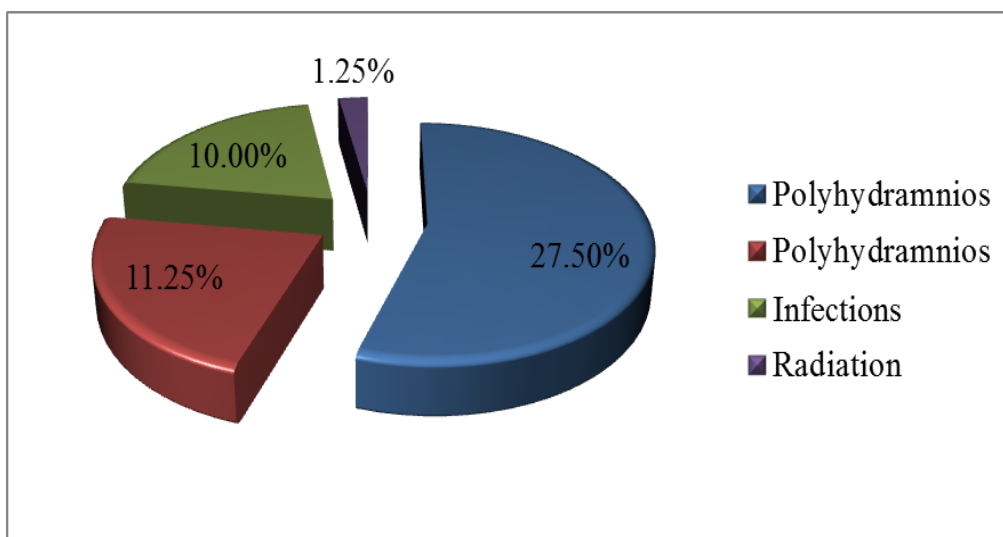


Figure (1): Prenatal history and predisposing factors.

Mode of presentation was demonstrated in table (1) and showed that in group 1, Fifty-one patients (63.8%) presented in the first week of life with acute Intestinal obstruction. ten patients with low type, 8 patients with intermediate type, 31 patients with high type and two patients with cloaca. Forty two patients presented in the first 3 days of the life 20 of them in the second day of life. In group 2, twenty nine patients (36.2%) with age

ranging between 2 weeks - 24 months, 3 of them presented at age of 2 weeks- 1 month and 11 at age between 1-2 months. This group presented with colostomy referred from other centers, chronic constipation, straining on defecation, or passage of stool from vagina, scrotum, and abnormal perineal opening or even from urethral. These are discovered either by parents or incidentally during examination.

Table (1): Mode of presentation.

Type of lesion	Group 1	Group 2
	Intestinal obstruction	Other presentation*
Low	10	14
Intermediate	8	7
High	31	7
Cloaca	2	-
Rare malformation	-	1
Total	51(63.8%)	29(36.2%)

Classification and incidence of types was demonstrated in table (2) and showed that 24 patients (30.0%) with low lesion; 4(5.0%) of them with anal stenosis, 5 (6.25%) patients with anterior ectopic anus, 9(11.25%) with anocutaneous fistula, 1(1.25%) with imperforate anal membrane and five patients (6.25%) patients with anovestibular fistula. Fifteen (18.7%) patients with

intermediate lesion, 5(6.25%) of them with anal agenesis and 10(12.5%) with rectovestibular fistula. Thirty eight (47.5%) with high lesion; 25 of them (31.25%) with rectourethral fistula, 11(16.75%) with rectoanalagenesis with no fistula, 2(2.5%) with rectovesical fistula. Two (2.5%) female patients with cloaca. One (1.25%) patient with rare malformations (cloacal exstrophy).

Table (2): Classification and incidence of types.

Type of lesion	Male	Female	Total	%	
Low (n=24 (30.0%))	Anal stenosis	-	4		
	Anterior ectopic anus	-	5		
	Anocutaneous fistula	9	-	9	
	Imperforate anal membrane	1	-	1	
	Anovestibular fistula	-	5	5	
Intermediate (n=15 (18.75%))	Anal agenesis	-	5		
	Rectovestibular fistula	-	10	10	
High (n=38(47.5%))	Recto urethral fistula	-	25	25	
	Recto anal agenesis-no fistula	10	1	11	
	Recto vesical fistula	2	-	2	
Cloaca	-	2	2	2.5	

Rare malformations (Cloacal exstrophy)	-	1	1	1.25
Total	56	24	80	100

Results of invertogram in relation to operative finding were demonstrated in table (3), in which a total number of 35 patients with cross table radiograph (excluding those with evident fistula) 28(80.0%) of them with correct diagnosis radiologically and operatively,

7(20.0%) patients with false result in which 4(11.4%) of them with radiological evidence of low type but operatively high type and 3(5.4%) patient with high lesion radiologically but proved to be low lesion operatively.

Table (3): Results of invertogram in relation to operative finding.

Finding (Invertogram / operative)	No.	%
True results	28	80
False results	7	20
Invertogram-low lesion / operative high lesion	4	11.4
Invertogram-high lesion/operative low-lesion	3	5.4
Total	35	100

Results of distal colostogram were demonstrated in table (4) and showed that it was done for 20 patients with high

lesion; 8(40.0%) of them shows urinary fistula and in 12(60.0%) patients no connection visualized.

Table (4): Results of distal colostogram.

Finding (Fistula)	No. of patient	%
Positive	8	40
Negative	12	60
Total	20	100

Operative findings-condition of anal dimple was demonstrated in table (5), which elicited those 20 (25.0%) patients with low type, 7(8.75%) patients with intermediate type, and 8 (10.0%) patients with high type. Flat perineum found in 2(2.5%) patients with low type,

8(10.0%) patients with intermediate type, 32(40.0%) patients with high type including cloaca, and 1(1.25%) patient with cloacal exstrophy. Bucket handles deformity is present 1(1.25%) patients with anal stenosis. Only one patient (1.25%) had normal anal opening.

Table (5): Operative findings-condition of anal dimple.

Condition of anal dimple	Low	Intermediate	High & cloaca	Rare malformation	Total	%
Well performed	20	7	8	-	35	45
Flat	2	8	32	1	43	52.5
Bucket handle	1	-	-	-	1	1.25
Normal anus	1	-	-	-	1	1.25
Total	24	15	40	1	80	100

Types of operations was demonstrated in table (6), in which among the males, for the low lesions, the Cruciate anoplasty was done on 14 patients, 4 of them were with severe anal stenosis, 9 with anocutaneous fistula and 1 with anal membrane. For the intermediate lesion, all 5 male patients with anal agenesis underwent single stage cruciate anoplasty. For the high lesion, staged operation was the standard method for management. Thirty seven patients underwent first stage colostomy operation, second stage pull through done 6-12 month later on in 29 patients; all of them underwent closure of colostomy 2 months later. Among the female; for the low lesions, anal transposition done on 9 patients with anterior ectopic anus and anovistibular fistula, cut back surgery in 2 patients with anovestibular fistula and limited one stage posterior sagittal anoplasty in 5 patients with ectopic anus and 2 patient with anovestibular fistula. One patient with anterior ectopic anus put on regular dilatation.

Concerning the intermediate lesion, staged operation used as in those with high lesion. First stage colostomy operation done in 10 patients followed by pull through operation 2-4 months later in 8 patients to whom colostomy closed 2 months later. For the high lesion and cloaca, first stage colostomy done on 1 patient with high lesion and 2 patients with cloaca. Second stage pull through operation done for 2 patients with cloaca and 1 patient with rectoanal agenesis with no fistula followed by closure of colostomy 2 months later.

Table (6): Managing-Types of operations.

Type of lesion	Type of procedure	Male	Female	Total
Low	Cruciate ano plasty	14	-	14
	Cut back surgery	-	2	2
	Transplantation	-	7	7
	Dilatation	-	1	1
Intermediate	- Single stage operation	5	-	5
	* Cruciate ano plasty	5	-	5
	- Stage operation	-	10	10
	* Colostomy	-	10	10
	* Pull through	-	8	8
	* Closure colostomy	-	8	8
	- Dilatation	-	-	-
High and cloaca	-Stage operation	37	3	40
	* Colostomy	37	3	40
	* Pull through	29	3	32
	* Closure colostomy	29	3	32

Type of Colostomy was showed in table (7) and revealed that transverse colostomy has been done on 25(50.0%) patients and pelvic colostomy 25(50.0%) patients.

Table (7): Types of colostomy.

Types of stoma	No.	%
Transverse colostomy	25	50
Pelvic colostomy	25	50
Total	50	100

Types of pull through were demonstrated in table (8) and showed that the main pull through operation used was the posterior sagittal anorectoplasty (PSARP) which has

been used in 37 (92.5%) patients, while abdominoperineal pull through has been used in 3 patients(7.5%).

Table (8): Types of pull through operations.

Types of procedure	Intermediate	High	Colaca	Rare	Total	%
Abdomino-perineal	-	2	1	-	3	7.5
P.S.A.R.P.	8	28	1	-	37	92.5
Total	8	30	2	-	40	100

DISCUSSION

Anorectal malformations are probably the most common congenital cause of intestinal obstruction in the newborn and perhaps the most common congenital anomalies in the gastrointestinal tract encountered in any part of the world. Most of the cases of imperforate anus should be discovered at birth during the initial physical examination. Delivery of an infant by a midwife failed to reveal the anomaly during the cursory examination and further delayed the diagnosis.^[19, 20]

In most instances the lesion will be noticed immediately after birth by simple inspection but the perineum may look remarkably normal to casual attendant and the abdominal distension of low intestinal obstruction may in these circumstances be the presenting feature. If the fistula is of considerable size the bowel may however decompress itself adequately for a considerable period of time.^[21]

In the current study, the main presenting feature was acute intestinal obstruction (group 1) in 63.8% of cases;

31 of them with high type. Forty two (52.5%) of this group presented to us in the first 3 days of life and the remaining 9 patients are of high lesion with fistula has got delay in their presentation up to seven days: the presenting features for (group 2) were delayed in diagnosis from one month to two years in some cases, majority of them were of low and intermediate type. In all these cases there was a communication of the rectum to perineum or urethra. These findings reflect poor neonatal assessment whether by birth attendant or the family, which leads to delay in diagnosis of most of our patients with increasing morbidity and mortality. At the same time some patients with high lesions presented in the second week of life with mild form of intestinal obstruction or passage of meconium per urethra or vagina. This sustained that wide fistulous connection to urinary system may deflate the bowel for a period of time. Five percent of the present patients were premature and this suggests that the prematurity has nothing to do with increase incidence of anorectal malformation.

A survey of the surgical section of American Academy of pediatrics reviewed 1142 children. Of 616 males, 328 (50%) had supra levator lesion in contrast to 90 (19%) of 381 females.^[1] According to this study supralelevator lesion represent about 69% this is nearly similar to what we record in present study in which supra levator lesions (high and intermediate) represent 68.7%.

In all reported series including Liverpool series the most common defect in boys is rectourethral fistula.^[2, 21] This is similar to present findings in which rectourethral fistula represent (31.2%) which are the commonest male lesion. The second common male lesion is perineal fistula^[2] which does not fit to our finding where anorectal agenesis without fistula is the second common male lesion with an incidence equal to (12.5%), this could be attributed to high percent of Down's syndrome in whom anorectal agenesis without fistula is the commonest on the other hand rectovestibular fistula is still the most common lesion in girls^[2, 19] which is consistent with our study with an incidence equal to (12.5%).

Most cases appear sporadically with no evidence of an increase risk in sibling.^[21] A familial history of anorectal anomalies is unusual. The risk of a family having subsequent infant with the same anomaly is about (1.0%).^[1,2] In the present study family history was positive in (6.25%) which is higher than the above percent, but this cannot exclude that genetic factor has no role in etiology since most studies point to an autosomal recessive type of inheritance with a risk of recurrence of about (25.0%). At the same time an autosomal dominant syndrome affecting anus, hands and ears has been described by Townes and Brockes.^[21]

Hydramnios during pregnancy is a sign of an abnormal fetus. In one series of 74 mothers with polyhydramnios during pregnancy, 27.0% of the infants had severe congenital anomalies, 8.0% had gastrointestinal tract obstruction.^[1] In the current series polyhydramnios were positive in mothers of 22 patients (27.5%), most of these infants have no perineal connection. This suggest that hydromnios is an important sign for gastrointestinal malformations including anorectal malformations that should alert us to assess and investigate fetuses of mothers with polyhydramnios, and to assess and examine the baby soon after delivery so that problems of delay diagnosis will be prevented or reduced.

Maternal ingestion of drugs like thalidomide, oral contraceptive has been implicated in the etiology and theoretically folic acid inhibitors or folic acid deficiency could cause multiple system anomalies including anorectal anomalies. Mechanical factor has also been implicated.^[21] These findings suggest an environmental factors including drugs that might predispose to anorectal malformations, 11.2% of the present patients have positive history of drug ingestion by their mothers during the first trimester, most of it were antibiotics and some of

it were antihypertensive and contraceptive and only one patient with cloacal exstrophy his mother had received chemotherapy and radiotherapy because of malignancy. 10.0% of the present patients their mothers gave history of infection in the first trimester in the form of local genital or systemic infection. These findings may support the environmental factor in predisposition to anorectal malformations but further study is necessary.

In many cases only clinical means have been used and the lesion will have been noticed immediately after birth by simple perineal inspection while in others adequate use of technical studies are necessary.^[21-23]

The present policy for diagnosis of imperforate anus and its level depends first on proper clinical examination, which has been found to be sufficient in most patients in (group 2). Difficulty in differentiating anovestibular and reovestibular fistula has been overcome by probing the fistula under anesthesia and noticing the direction of the probe.^[1] On the other hand difficulties in guessing or confirming the level of the lesion in group (1) reduced by radiological examination using the classical cross table lateral radiograph which gives us diagnosis in accordance with the clinical and operative findings in 28 patients (80.0%)

Ultrasonography provides excellent information about the location of rectal pouch.^[1] Oppenheimer *et al.*,^[12] and Donaldson *et al.*,^[24] found that ultrasound correctly predicted the level of the distal pouch and can readily detect associated renal anomalies. This has been tried but the unavailability of high resolution ultrasonographic machine and lack of a well trained operator forces us to restrict the use of ultrasound for the detection of associated renal anomalies.

Accurate demonstration of the anatomy of any associated fistula between urinary tract and rectum is essential for optimal surgical management, a simple contrast medium through distal colostomy has been used by us but too often failed to demonstrate a urinary fistula because in 20 of the present patients with distal colostogram, only 8(40.0%) of them show positive fistula and the remaining 12(60.0%) patients show no fistula radiologically but positive fistula operatively, such finding is similar to that done by Lerhau *et al.*,^[15] who found that none of his five patients with simple distal colostogram demonstrate urinary fistula as a result of that an augmented pressure distal colostogram is recommended to fulfill this requirement and to confirm the level of the anomaly prior to definitive surgical repair.^[25, 26]

It is generally agreed that there are few problems in the treatment of low anorectal malformations. There is however a controversy in the treatment of high deformities for which several different operative methods is favored by different surgeons.^[27]

Among the present study sample, the first step during the operation is the assessment for a condition of anal dimple and found that 83.3% the patients with low lesion and 1/2 of intermediate lesions have well formed anal dimple while more than 3/4 of the patients with high lesion have flat perineum. This was very helpful as a supportive clinical finding to the judgment in determining the level of the lesion, since a well formed anal dimple suggests the presence of a very low defect, while patients with high lesions tend to have high incidence of flat perineum.^[1, 2]

Through clinical, radiological and operative assessment fistulous termination to various sites namely urinary, genital and perineum were found in 63.7% of the present sampled patients, about 29.4% in female and 71.8% in males. This is similar to the series of the surgical section of the American Academy of Pediatrics with regard to the male, but not the female, in which fistulous connection found in 72% of males and 90.0% of females.^[1] The majority of urinary fistula (70.4%) among patients in the current study were not evident clinically or radiologically and proved to be positive during pull through operation.

All patients with low lesions were managed by single stage perineal operation. The repair of cutaneous fistula is by opening the fistula till reaching the anal canal then skin and mucosa approximated by absorbable suture but in severe anal stenosis V-Y plasty is used or regular anal dilatation^[1,4,5,21] while in complete anal membrane anoplasty is preferred. None of current patients underwent V-Y plasty but cruciate anoplasty form the bulk of the present surgical procedure with very good, and satisfactory results in 14 of the present patients, namely anal stenosis, anocutaneous fistula and anal membrane. Minimal anterior ectopic anus in female required no treatment or simple dilatation otherwise V-Y plasty is done^[1] or minimal mobilization of the intestine sufficient for it to be transposed and placed within the limit of the external sphincter.^[2] While 9 of the patients with anterior ectopic anus and anovestibular fistula were treated by transplantation of the ectopic opening within external sphincter muscle fibers and those with mild degree of ectopic anus put on regular dilatation only.

Anal agenesis can be managed in neonatal period through perineal anoplasty and as the radiological examination cannot give a final correct diagnosis of this lesion. It was used to aspirate meconium through the skin of the anal dimple under general anesthesia and measuring the actual distance between the skin and the blind bowel^[4] and by this the diagnosis could be confirmed. Stevenson *et al.*,^[28] has developed a technique to facilitate localization and dissection of the rectal pouch for perineal anoplasty in patients with infralevator anorectal malformation who do not have a perineal fistula present through fluoroscopic percutaneous transperineal placement of Fogarty embolectomy catheter through the centre of the anal wink with excellent results.

All the present sampled patients with anal agenesis underwent preoperative pouch localization through meconium aspiration and perineal cruciate anoplasty were done for all of them with uncomplicated postoperative period. Surgery for rectovestibular fistula has been the subject of controversy most pediatric surgeons use cutback, fistula transposition with or without colostomy and lately posterior anorectoplasty with colostomy. Demirbilek *et al.*,^[29] prefer to perform anal transposition without colostomy in 47 patients when the rectovaginal septum is amenable to dissection (width >2mm). However as cutback surgery produces cosmetically ugly perineum and psychological upset to the parents, it has been prohibited by us and although Demirbilek prefers anal transposition with a good result, put these patients on regular fistulous dilatation was preferred and defer the operation till 5-6 months of age at which 3-stages operation performed using posterior sagittal anorectoplasty as advocated by Pena and by this, the risk of infection, dehiscence and recurrence of fistula were obviated.

The recent advances in the management of anorectal malformations have yield good results in the short term. In 1982 Pena and Devries introduced the PSARP, which has been used in about (92.5%) of patients of the present study who underwent definitive pull through operation and in the remaining 7.5% a high rectal pouch that cannot be reached by posterior sagittal approach, combined abdominal and posterior sagittal approach was needed. This is similar to what have been found by Pena in which 90% of the malformations can be repaired by a posterior sagittal approach and the remaining 10.0% an abdominal approach is added.^[8] All female intermediate lesions, all high lesions and cloaca treated by stage operation started by colostomy in the form of transverse colostomy in 50.0% and pelvic colostomy in 50.0%.

Recently Albanes *et al.*,^[30] found that one stage repair of high imperforate anus in the male neonate is feasible, but it is not agreed upon yet whether it is preferable compared with delayed (2 or 3 stages) repair. This depends on ultimate long term anorectal function which cannot be assessed for several years. Until that time, the surgeons continue on the standard rule of deferring pull through operation till 6-12 months of age^[1], although it can be done earlier than that.^[2] It was found that PSARP is very suitable for all supralevator lesions with good exposure of the rectum, fistula and pelvic musculature plus minimal dissection, and recently Ismail-A reported that the muscle component are divided in PSARP and this still does not affect continence, assuming it is carried out in exactly the midline.^[31]

Tapering was done in few cases and the rectum sutured to skin by absorbable material, drain were used and removed 2 days later. Foly's catheter left for a week in cases of urethral fistula.

CONCLUSIONS

High lesion carries the highest incidence of associated anomalies which is responsible for majority of the deaths, and genitor-urinary anomalies are the commonest. Cross table radiograph and distal colostogram carry a significant percent of false results. Low lesions and anal agenesis can be managed through perineal approach without colostomy. Posterior sagittal anorectoplasty is the optimal surgical procedure for high lesions and rectovestibular fistula.

Recommendations

1. Every mother with hydromnios, history of drug ingestion or infection in the first trimester should be screened for the presence of anorectal anomalies.
2. All birth attendants should be instructed to examine the newborn baby perfectly before discharge to obviate the problem of delay diagnosis.
3. Regular checkups are crucial for determining the long term result of continence.
4. Regular postoperative dilatation is an important part of the treatment and parents must be taught how to perform dilatation as their rule in this is of great importance.

REFERENCES

1. Raffensperger JG. Swenson's Pediatric Surgery. Fifth ed. USA: Printice Hall International., 1990; 587-623.
2. Spitz L and Goran AG. Rob and smith's Operative Pediatric Surgery. Vol.1.Fifth ed. London: Printice Chapman and Hall Medical., 1995; 423-458.
3. Adkins JC and Kieswetter WB. Imperforate anus. *Surg clin North Am.* 1976; 56(2): 379-394.
4. Holder TM and Ashcraft KW. Pediatric Surgery. First ed. USA: W.B. Saunders Company. 1980; 401-417.
5. Kiely EM. Imperforate anus. *Surgery.* 1986; 1(39): 927-929.
6. Schwartz SI. Principles of surgery. Vol.2. Sixth ed. USA: McGraw Hill International. 1979; 1681-1719.
7. Woolley MM. Mark Ravitch Pediatric Surgery, Vol.2. Third ed. USA: Printice Hall International. 1979; 1060-1072.
8. Sbobat SJ. Nelson Text book of Pediatrics. Fifteenth ed: USA: W.B. Saunders Company. 1996; 1075-1078.
9. Hardy hendren W. Constipation caused by anterior location of the anus and its surgical correction. *J pediatr surg.*, 1978; 13(6): 505-512.
10. Claus D, Wese FX, Kay umbi J, Otte JB. Initial diagnosis of anorectal malformations. *Acta Chir Belg.*, 1983; 82(3): 163-177.
11. Harris RD, Nyberg DA, Mack LA, Weinberger E. Anorectal atresia: Prenatal sonographic diagnosis. *AJR Am Roentgenol.*, 1987; 149 (2): 395-400.
12. Oppenheimer DA, Carroll BA, Shochat SJ. Sonography of imperforate anus. *Radiology.* 1983; 148 (1): 127-128.
13. Tam PK, Chan FL, Saing H. Direct sagittal CT scan: anew diagnostic approachfor surgical neonates. *J pediatr surg.* 1987; 22 (5): 397-400.
14. Sachs TM, Applebaum H, Touran T. Use of MRI in evaluation of anorectal anomalies. *J pediatr surg.*, 1990; 15(7): 817-821.
15. Lernau OZ, Janca J, Nissan S. Demonstration of rectourinary fistula by pressure Gastrografin enema. *J pediatr surg.*, 1978; 13(6): 497-498.
16. Aleem AA, Mira K, Elbanna I, Eltabawy N, Aburayh G, Gaber A. A new transrectal pull through operation for high anorectal anomalies. *J pediatr surg.*, 1980; 15(5): 620-622.
17. Adeyemi-SD and de-Rocha-Afodu N. Management of imperforate anus at Lagos university teaching hospital, Nigeria: a review of ten years experience. *Prog pediatr surg.*, 1982; 15: 187-194.
18. Leenders E. Imperforate anus. Principles and methods of surgical treatment. *Acta Chir Belg.*, 1983; 82(3): 183-190.
19. AL-Salem A, Qaissaruddin S, Kvarma K. Anorectal malformations experience from pediatric surgery unit in Saudia Arabia. *Saudi Med J.*, 1997; 18(4): 353-355.
20. Acosta-Frina DJr, Ortiz-Interian CJ, Acosta-vasquez CESr. Imperforate anus delayed presentation in a 7-year-old girl. *J pediatr surg.*, 1993; 28(7): 962-964.
21. Rickham PP, Lister J, Irving IM. Neonatal Surgery. Second ed. UK: Butter worth and Co(publishers). 1978; 475-481.
22. de-Espanosa H. Anorectal malformations: their diagnosis and the initial decisions. *Rev Med panama.* 1994; 19(2): 79-83.
23. Young DG and Martin EJ. Baby surgery. Nursing management and care. Second ed. UK: HM+M publisher. 1979; 65-68.
24. Donalson JS, Black CT, Reynolds M, Sheran JO, Shkolnik A. Ultrasound of the distal pouch in infants with imperforate anus. *J pediatr surg.*, 1989; 24(5): 465-468.
25. Gross GW, Wolveson PJ, Pena A. Augmented-pressure colostomy in imperforate anus with fistula. *Pediatr Radiol.*, 1991; 21(8): 560-562.
26. Wang C, Lin J, Lim K. The use of augmented pressure colostography in imperforate anus. *Pediatr surg Int.*, 1997; 12(5-6): 383-385.
27. Ito Y, Yokoyama J, Hayashi A, Ihara N, Katsunata K. Reappraisal of endorectal pull through procedure: Anorectal malformation. *J pediatr Surg.*, 1981; 16(4): 476-483.
28. Stevenson RJ, Sheldon C, Ildstad ST. Percutaneous trans perineal pouch localization in low imperforate anus: a new approach. *J pediatr Surg.*, 1990; 25(2): 273-275.
29. Demirbilek S and Atayurt JF. Anal transposition without colostomy: functional result and complication. *Pediatr Surg Int.*, 1999; 15(3-4): 221-223.
30. Albanes CT, Jennings RW, Lopoo JB, Bratton BJ, Harrison MR. One stage correction of high

- imperforate anus in the male neonate. *J pediatr-Surg.*, 1999; 34(5): 834-836.
31. Ismail A. Reappraisal of the anatomy of the anorectal musculature. *Saudi Med J.*, 1999; 20(7): 501-503.