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Anorectal malformations in children

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ABSTRACT

Background/Purpose: Anorectal malformations are one of the most common congenital defects. This study was undertaken to study the hospital incidence of anorectal malformations (ARM), frequency of various types of defects, their sex distribution and the spectrum of anomalies associated with ARM. The effect of presence of an associated defect on mortality and morbidity was also studied.

Materials and Methods: One hundred consecutive children attending the pediatric surgery department were included in this study. A detailed history was taken, and examination was performed for the primary as well as the associated defects. Appropriate investigations like invertogram, cologram were done wherever indicated. Management was as per the standard protocol. The data was recorded and analyzed.

Results: Out of the 100 patients, 51 were males and 49 females. One out of every 6.62 admission was for ARM. Twenty percent of the female babies had high, 76% intermediate and 4% had low anomalies, whereas 80.39% males had high, 3.92% intermediate and 15.6% showed low malformations. Ten percent of the patients had pouch colon. Associated anomalies were seen in 33 patients – 20 males and 13 females; 19 in high, 10 in intermediate, 1 in low group and 3 children with cloacal malformations. Associated defects seen were urogenital (17%), cardiovascular (7%), gastrointestinal (9%), genital (5%) and limb defects (7%). There were 8 deaths, and complications were seen in 13 patients. Ten patients had two or more defects associated with ARM.

Conclusions: Anorectal malformations occurred equally in males and females. Females had intermediate defects more frequently, rectovestibular fistula being the commonest. Males were more likely to have high lesions; anorectal agenesis without fistula was the commonest defect. The most common associated defects seen were vesicoureteric reflux and esophageal atresia. Complications were seen more commonly in males with high lesions. There was a significant association between presence of an associated defect and mortality and morbidity.

KEY WORDS: Anorectal malformations, associated defects, cloaca, colostomy, pouch colon

Anorectal malformations (ARM) are one of the most common congenital defects. The usual reported incidence is between 1 per 1500 and 1 per 5000 live births.[1-5] ARM presents with a wide spectrum of defects, ranging from relatively low malformations to very complex cloacal anomalies.[6-15] Forty to seventy percent of these patients have one or more additional defects of other organ systems.[5-11] Urological defects are the commonest anomalies associated with ARM, followed by defects of the spine, extremities and the cardiovascular system.[14,15] The management of ARM in the neonatal period is crucial because it will determine the immediate future of the child.[15-17] The most important decision to be made is whether the patient needs a colostomy and whether a urinary diversion is necessary to prevent sepsis or metabolic acidosis. With better understanding of the anatomy, early diagnosis of ARM and its associated defects and increasing experience in management, better results are now being obtained. Not many studies on this subject are available in India.[1,2,7] Moreover, no definitive data regarding the frequency and distribution of various forms of ARM in the Indian population is available.[1,2,7] This study analyses the relative incidence of various types of ARM and their outcome in a childrens hospital in northern India.

MATERIALS AND METHODS

The prospective study was carried out between February 1999 and January 2000, and it consisted of 100 consecutive children with ARM. The study included children presenting with ARM in the neonatal period, those reporting for corrective surgery and patients for colostomy closure after the elective surgery. A detailed

clinical pro forma was filled up for each patient, incorporating antenatal history, postnatal history, clinical presentation and physical examination for the primary defect as well as associated anomalies. All the defects were classified as per the Wingspread classification. A nasogastric tube was passed to rule out the presence of esophageal atresia (EA), and further clinical examination of the cardiovascular system, abdominal wall and extremities was carried out. Radiological investigations included an invertogram after 18-24 h of life and an anteroposterior view of the abdomen in all newborn babies to look for other gastrointestinal defects such as atresias and rule out congenital pouch colon, a common occurrence with ARM.

Depending upon the location of rectal pouch in male babies, ARM was classified as high, intermediate and low; whereas in female babies, perineal examination confirmed the diagnosis, and radiological examination was done in selected cases.

Spinal and sacral defects found on clinical examination were evaluated with plain radiograph. Ultrasonography was done for all newborn babies and for children admitted for definitive surgery. Indications for a voiding cystourethrogram were presence of hydroureter, hydronephrosis, distended trabeculated bladder and posterior urethral dilatation on ultrasonography. An echocardiogram to evaluate the cardiovascular system was done whenever clinical examination revealed central cyanosis or a cardiac murmur. A genitogram was performed in patients with cloaca.

All male babies presenting with a low type of defect underwent a V-Y anoplasty in the neonatal period. Rest of the male babies with high or intermediate types of defects underwent a divided sigmoid colostomy. In female babies with anorectal defects with fistula, diverting colostomy was performed at the age of 3-6 months.

Definitive surgery was usually performed at the age of 1 year when the child was gaining weight, thriving well and having hemoglobin of at least 10 gm%. Before subjecting a male baby to definitive surgery, a distal cologram was performed under fluoroscopic control. All male patients with intermediate and high types of defects underwent definitive surgery in the form of a posterior sagittal anorectoplasty (PSARP).[17,18] Females with low types of defects and fistulas underwent a limited form of PSARP. In patients with a very high rectal pouch, an abdominal component was added to the repair in addition to posterior sagittal dissection. The technique of cloacal defect repair was based upon the length of the common channel. The technique employed for repair of rectovaginal fistula was same as that for vestibular malformations, the only difference being the length of the incision and the amount of dissection required to bring the rectum down to the perineum.

Postoperatively, all patients received broad-spectrum antibiotics. Oral feeds were started on the day of surgery followed by a regular diet. However, the patients who underwent a laparotomy in addition to a pull-through procedure required a period of fasting for 1-2 days along with nasogastric tube drainage.

Two weeks postoperatively, anal calibration was started with Hegars dilators, followed by a program of regular anal dilation till the desired size of anus was reached. The colostomy closure was usually done 2-3 months after the definitive surgery in most of the cases.

Functional evaluation of the patients who had completed all stages of surgery was done by interviewing the parents for bowel movement pattern, episodes of soiling, constipation and social activities as compared to the peers.

Associated defects were managed as per the standard methods.

RESULTS

Out of the 100 patients, 51% were males and 49% females. One out of every sixth child was attending the hospital for ARM. In males, high defects were more common (80.39%), whereas intermediate defects (65.30%) were commonest in females [Figure 1].

Various patients presented in different stages of treatment [Table 1]. Distribution of various types of ARM in male and female patients as per the Wingspreads classification has been shown in Tables 2 and 3.

Out of 45 neonates, 8 males underwent anoplasty for low ARM, whereas remaining 37 patients required colostomy. None of the female neonates showed low ARM without fistula. Ten patients presented with pouch colon (Type I-3, II-3, III-2 and IV-2).

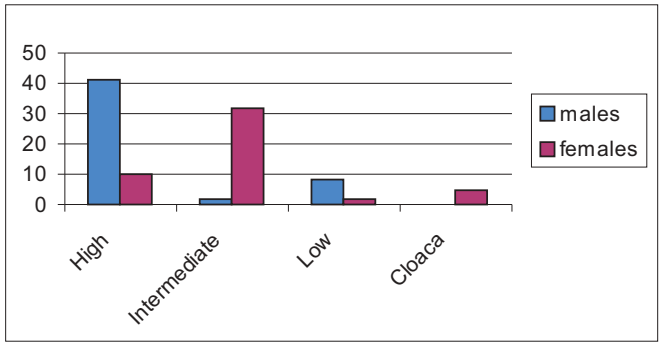

Figure 1: Distribution of defects

Table 1: Profile of patients

Category	Males	Females	Total
New born	32	13	45
For PSARP	9	12	21
For colostomy closure	8	13	21
Redo surgery (PSARP)	2	3	5
Recto and anovestibular fistula for colostomy beyond neonatal age	0	8	8
Total	51	49	100

PSARP - Posterior sagittal anorectoplasty

Table 2: Distribution of defects in males

Type of defect	No. (%)
Anorectal agenesis without fistula	25 (49.01)
Anorectal agenesis with rectoprostatic fistula	6 (11.8)
Pouch colon	7 (13.7)
Rectovesical fistula	3 (5.9)
Rectobulbar fistula	2 (3.9)
Anocutaneous fistula	8 (15.7)

Table 3: Distribution of defects in females

Type of defect	No. (%)
Anorectal agenesis without fistula	7 (14.3)
Pouch colon	3 (6.1)
Rectovestibular fistula	30 (61.2)
Rectovaginal fistula	2 (4)
Anovestibular fistula	2 (4)
Cloaca	5 (10)

Associated defects were seen in 33 patients (20 males and 13 females). Nineteen of 51 patients with high ARM (37.25%) and 60% of patients (3 out of 5) with cloaca had associated anomalies as compared to 25% association with intermediate and low combined (11 out of 44). Ten patients had more than one associated anomaly.

Genitourinary anomalies were the commonest (17 anomalies). Vesicoureteric reflux (VUR) (6); renal agenesis (3); ectopic kidney, urethral stenosis, polycystic kidney (one each) were the urological anomalies; whereas genital defects were vaginal agenesis (2), hypospadias (1), bifidus phallus (1) and duplicate penis (1). Gastrointestinal anomalies were esophageal atresia and tracheoesophageal fistula (6), duodenal atresia (2), omphalocele (2) and Meckel's diverticulum (1). Cardiovascular defects included ventricular septal defects (3), dextrocardia (2) and patent ductus arteriosus (2). Vertebral and spinal defects (sacral agenesis-3, spina bifida-1, hemivertebra-1, sacrococcygeal teratoma-1), limb defects (talipes equinovarus-1, radial agenesis-2, syndactyly-2, supernumerary digits-1), facial clefts (2), tongue-tie (1) and subglottic stenosis (1) were the other associated anomalies.

Overall mortality rate was 8%, and three-fourths of the patients had associated anomalies (urological-4, gastrointestinal-1, cardiovascular-1). All the patients who died were neonates undergoing colostomy (7) and anoplasty (1). Amongst the survivors, wound infection (3), colostomy prolapse (3), postoperative fever (2), wound dehiscence (2), revision of colostomy stoma (2) and anal stenosis (1) were the complications seen. Sixty-one percent of the patients having a complication had an additional defect.

Out of 21 patients having complications 13 (62%) were of high ARM and 6 patients had intermediate ARM. Twenty-one patients (8 males, 13 females) completed all the stages of treatment. Eighteen children (85.7%) have normal sensations of bowel movements and are able to keep themselves clean with bowel management program, which essentially consisted of saline enemas, dietary modifications and toilet training. Six patients (2 males, 4 females) had one to two episodes of soiling in 24 h, which according to parents was not hampering their social activities, whereas 3 male children were having more than two episodes of soiling per day.

DISCUSSION

ARMs occur quite commonly.[1-5] Many systems of classification have been suggested; however, the terms high and low have been broadly recognized and applied worldwide. Pena reports the most common lesion to be ARM with recto urethral fistula in males and cloacae amongst females[13]; however, most commonly seen defect in the Liverpool series was anocutaneous fistula.[10] In our series, a high lesion without a fistula was most common in males, whereas in females rectovestibular fistula was the commonest. The level of lesion will obviously have a bearing on the outcome and quality of life.

We found that 33% of our patients had one or more associated malformation, whereas reported incidence varies from 36-67%; highest incidence of associated defects was seen in cloaca followed by high lesions. We found urogenital defects to be the commonest; however, a variable incidence has been reported (20-85.7%) in literature.[13-15]

VUR was the commonest association; it has got long-term implications as well.[13-15] Four neonates, who expired, had associated genitourinary malformations in the initial screening examination.

Incidence of esophageal and duodenal atresia was similar to other reported studies.[1,10] One patient with multiple gastrointestinal anomalies, i.e., duodenal atresia, esophageal atresia and ARM died. Cardiovascular defects were seen less commonly in our patients.[1,2,17] Postoperative wound infection occurred in 3% of the patients, major source being contamination during colostomy as reported by others also.[7,10,18]

This report gives an outline of the epidemiological profile of the ARM patients from a center in northern India, and the babies with associated anomalies undergoing colostomy constituted the group having poor outcome. There was no mortality amongst the older children undergoing definitive surgery. Every attempt should be made to perform the definitive surgery so as to ensure a good outcome, integrating these children into the mainstream.

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