ANORECTAL MALFORMATIONS: FUNCTIONAL OUTCOME OF POSTERIOR SAGITTAL ANORECTOPLASTY

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ABSTRACT

Objective: *To assess the functional outcome of patients with anorectal malformations treated with Posterior Sagittal Anorectoplasty (PSARP).*

Material and Methods: *This study was conducted on 355 patients with anorectal malformations at Lady Reading Hospital, Peshawar from July 1998 to July 2004.*

Distal loopogram was performed routinely in all patients except the female patients with rectogenital tract fistula. PSARP was performed in all the patients with intermediate, high type and cloacal malformations. Operative and postoperative mortality and morbidity was recorded. Regular anal dilatation was advised after discharge, which was continued for 3-6 months after the PSARP procedure. Patients were followed up for 6-12 months.

Results: *Study included 355 patients (245 boys and 110 girls) ranging in age from 12-36 months. PSARP was performed in all the patients. There were 195 boys with rectourethral fistula and 95 girls with genitourinary tract fistula while 5 girls had cloacal malformations. Operative and postoperative mortality was 9/355 (2.5%). Early functional results were good in 30%, fair in 45% and poor in 25% patients. Chronic constipation and anal stenosis was found in 99 and 35 patients respectively. Mucosal prolapse with perineal itching was present in 60 patients. Recurrent UTI and orchitis was found in 5 and 3 patients respectively. Urethral stricture and urethral diverticulum was found in two cases each, while redo surgery was performed in two patients.*

Conclusion: *The incidence of high and intermediate anorectal anomalies was more in male babies. The PSARP procedure is safe with good functional results in terms of faecal continence.*

Key Words: *Anorectal Malformation, Anorectal Anomalies, Posterior Sagittal Anorectoplasty, Pull Through, Faecal Continence, Constipation.*

INTRODUCTION

Anorectal anomalies are congenital malformations in which the distal part of the hindgut fails to develop or partially develops leading to variable types of anomalies of the anorectal region[1] Different types of the anorectal anomalies are grouped together and an international classification was proposed by Stephan which was revised by Smith[2]. Winspread classification in 1985 proposed a working formulation in which anorectatal anomalies were grouped into low, intermediate, high and rare types. Sex of the baby and the presence or absence of the fistula to the genitourinary tract is considered to determine the type of anomaly[3]. Babies with low type imperforate anus are treated with perineal surgery in the neonatal period while other patients (high, intermediate and rare type) are managed in stages i.e first colostomy and then Posterior Sagittal Anorectoplasty (PSARP) followed by closure of colostomy.[4,5] A number of procedures have been adopted by various authors in the past but now-a-days the Pena's procedure (PSARP) is the well established and the gold standard procedure for the definitive treatment of patients with anorectal malformation.[5-10]

The aim of this study was to assess the functional outcome of patients with anorectal malformations treated with Posterior Sagittal Anorectoplasty.

MATERIAL AND METHODS

A total of 355 patients were studied

SEX WISE DISTRIBUTION OF DIFFERENT TYPES OF ANORECTAL MALFORMATIONS (ARM)

Types of ARM	Sex		Total
	Boys = 245(69%)	Girls = 110(31%)	
with fistula	290	195	95
without fistula	50	50	10
Cloaca	05	00	05
Total	**355**	**245**	**110**

Table 1

including both sexes during a period of 6 years from July 1988 to July2004, who were admitted through OPD to Paediatric Surgery Unit, PGMI, Lady Reading Hospital. Age of the patients ranged from 12 - 36 months. Proper clinical record was maintained including history and physical examination of the patients to assess the type of anomaly, any other associated anomaly and assess the fitness for the procedure. Distal lopogram was performed in all patients except the female babies with fistula to the genital tract. The distribution of anorectal anomalies was based on the Wingspread classification. In all the patients with, intermediate, high and cloacal anomalies, PSARP was performed. Any mortality, and morbidity related to the procedure was recorded postoperatively. Results were recorded and analysed after the assessment of these patients in the OPD during their follow up visits. The duration of follow up ranged form 6-12 months. Defecation history of the child and examination of the anus was the routine in follow up examination. Every patient was advised regular anal dilation with an appropriate size anal dilator for a period of 3-6 months after the PSARP.

In follow up visits emphasis was given to the pattern of defecation, constipation, perineal soiling, position, size and shape of the anus and also the general health of the patient was assessed. Faecal continence was the aim to be achieved with efferts to preserve the components of the sphincteric mechanism. The results of continence were assessed and graded as good (continent), fair (occasional soiling), and poor (incontinent). Other functional results were also assessed and recorded.

RESULTS

A total of 355 patients were studied during 6 years period from July 1998 to July 2004.

There were 245 boys and 110 girls. Age ranged form 12-36 months, provided the child is otherwise in good health and fit for surgery. All the patients with intermediate, high anomalies and cloacal malformation needed posterior sagittal anorectoplasty (PSARP). There were 195 boys with rectourethral fistula and 50 without fistula. Ninety five girls had rectovaginal or rectovalvar fistula while 10 girls had no evidence of any fistula. There were 5 girls with cloacal malformation (table 1). In 295 patients isolated PSARP was performed while in 60 patients this procedure was combined with abdominal approach. There were 5(3 male + 2 female) deaths after the procedure (isolated PSARP) while 4(3 male + 1 female) patients died postoperatively who were treated with combined abdominal approach (table 2). Early functional results were observed which was good in 30% fair 45% and poor in 25% of patients (table 3). There were chronic constipation in 99 patients. mucosal prolapse in 60, anal stenosis needing regular dilatation in 35 patients, urethral stricture and diverticulum each two cases. Rec-UTI after the procedure was found in 5 while 3 patients presented with orchitis (one side). Redo surgery was done in two cases one male and one female, who improved later on after the procedure (table 3).

DISCUSSION

The long term functional outcome of the repair of anorectal anomalies remains incomplete which leads to significant anorectal malfunction in childhood and beyond that age. Although there is tremendous improvement in the understanding of the development and surgical anatomy of the anorectal malformations, it is still unknown, which is the optimal procedure to treat these complex anomalies. For the last 15-20 years PSARP procedure has been the gold standard of all the procedures (pull through) performed in the past for the treatment of anorectal malformations. The short term complications in patients treated with this procedure are less as compared to the old-fashioned classical pull through procedures, which involved abdominal approach for the mobilization of the distal gut[5,11,12].

We performed PSARP in 355 patients with good and fair results in majority of the patients (75%) in terms of faecal continence. Mortality was high in patients where the PSARP was combined with abdominal approach i.e 4 out of 60(6.6%) as compared to isolated

MORTALITY RATE AND ITS RELATION WITH THE TYPE OF PROCEDURE

TYPE OF PROCEDURE	Mortality		Total
	Male	Female	
PSARP (Isolated) (n=295)	03	02	05(1.7%)
PSARP Abdominal approach (n=60)	03	01	04(6.6%)
Total (n=355)	**06**	**03**	**09(2.5%)**

Table 2

PSARP i.e 5 out of 295 (1.7%). The overall mortality is in comparison with the results mentioned in the literature(2.5%)[5,6,13,14]. But most of the mortality is because of the extensive nature of the abdominal approach of PSARP procedure (6.66%), which is acceptable in our setup. This can be reduced by improving the postoperative care and the advent of paediatric intensive care facilities.[15,16]

PSARP was the procedure in all patients irrespective of the type of malformation. Anal continence was the main aim of the whole exercise in the management of anorectal anomalies. Preservation of the voluntary sphincters is the major component of the continence mechanism which is achieved by PSARP but there are other factors which lead to poor results in some of the patients as there was no selection of cases for this procedure and was performed randomly in all cases with high and intermediate anorectal malformations. Sacral anomalies and flat bottom patients were also offered the some procedure because the alternative option (permanent stoma) was not acceptable to the parents which caused increase in the number of patients with poor results. The good and fair results combined together (75%) were acceptable by the parents in early period of the study which may improve with age when the child grows older. As the follow up is short and irregular, exact results cannot be determined. Symptoms may improve when the children grow to adolescence age. More or less the same results have been observed in other studies.[17-21]

Chronic constipation was commonly observed in this study (99 cases). Although we use the terminal part of the gut which theoretically has component of the internal anal sphincters. Probably this is the reason that internal splinter saving procedure has high incidence of chronic constipation[22]. Rectal pouch insensitivity or lack of activity in the rectal pouch can be another possibility which may lead to rectal inertia and chronic constipation[23]. Most of the time the part of the fistula is taken along with the rectum and is utilized in the anorectoplasty which is usually devoid of the ganglion cells and lacking peristaltic activities. Ischemia of the distal end of the rectum will damage the ganglia in the rectum again that segment acts as an achalasia of the gut at that point. Sometimes congenital. aganglionosis may be associated with anorectal malformation which has been mentioned by some observers and can be the cause of chronic constipation[24]. We have treated all of our patients who had constipation with laxatives, kleen enemas, regular anal dilation with satisfactory improvement in their symptoms. The exact cause of constipations could not be determined in our cases but a combination of the above mentioned factors could be responsible as observed in many other studies[24,25]. Anal stenosis was seen in 35 patients needing repeated anal dilatation under general anesthesia to an adequate size. One patient had severe anal stenosis who ended up with revision of the procedure through perineal anoplasty.

Perineal soiling (160 cases) and mucosal prolapse (60 cases) were troublesome problems which were of concern for the parents. Social stigma of perineal soiling at odd times of the day compelled the parents to seek advice. Initially the condition was refractory to treatment but with increasing age it was observed to achieve improvement in symptoms with regular toilet training. Mucosal prolapse was not a problem and was acceptable to many of the parents. Some of the parents complained of perineal itching and excoriation but got better with local treatment. In 2 patients redundant mucosa was excised which relieved the symptoms. Urethral stricture, urethral diverticulum and recurrent UTI was seen in 10 patients which is comparable with the observation of many other studies[11,12,25]. Long-term

	No. of Patients (n=346)	Percentage
Perineal Soiling	160	45%
Clean	105	30%
Ch. Constipation	99	28%
Feacal incontinence	81	25%
Mucosal prolapse	60	17%
Anal stenosis	35	10%
UTI- recurrent	05	1.5%
Orchitis	03	1.9%
Urethral stricture	02	0.5%
Urethral diverticulum	02	0.5%
Redo Surgery	02	0.5%

Table 3

antibiotic prophylaxis were advised with which they improved and were followed up and further evaluated for other urological problems.

Redo surgery is rarely needed for various reasons. There were 2 cases which needed redo-surgery. One girl developed severe anal stenosis and stricture of the anorectal canal which was resistant to regular anal dilatation. In this case abdominal approach was done for the correction of the problem. The other patient was a boy whose anus was incontinent which was placed wrongly outside the muscle complex. (anterior to the sphincter). This was corrected by redo-surgery only through PSARP who got better and doing well after surgery. The results of redo-surgery is difficult to assess because long term results will need prolonged follow up which is poor in our setup.

This initial report of a series of patients with anorectal malformation gives us tremendous encouragement to continue PSARP procedure for the treatment of all these patients. The results of our study are more or less the same as reported by Pena and other workers[8,9,25,26].

Mortality was observed in 9 patients, which was more (4 out of 60) in patients with abdominal approach than the patients with isolated PSARP (5 out of 295). Mortality should be low with isolated PSARP, which is also observed in our study and is in comparison with the literature. The existing mortality is mostly due to the associated anomalies especially cardiac and renal anomalies, however nothing such problem was observed to be the cause of deaths in our study. Aspiration pneumonia was responsible for the mortality observed in these patients as apposed to other causes seen in other studies. We treated all these patients in conventional three-staged procedure which is safe in our setup, however some authors have recommended anterior perineal anorectoplasty and one stage repair of the anomaly with good short-term results[4,11,25,26].

CONCLUSION

Male patients predominate in this study, which shows that the incidence of high and intermediate type of anorectal anomalies is more in male babies than the female babies. Mortality and morbidity is more in patients in whom abdominal approach was adopted along with PSARP as compared to isolated PSARP. Over all the procedure is safe with good functional results in our setup which are more or less comparable to the ones reported by various other studies in the world literature so we recommend this procedure to be the gold standard and most appropriate in all patients with high and intermediate anorectal malformation.

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