TO ASSESS THE MORBIDITY, MORTALITY AND SHORT-TERM OUTCOME OF PRIMARY PSARP IN MALE BABIES WITH HIGH AND INTERMEDIATE ANORECTAL MALFORMATIONS

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ABSTRACT

BACKGROUND
The aim of the study is to assess morbidity, mortality and short-term outcomes of Primary Posterior Sagittal Anorectoplasty (PSARP) in male neonates.

MATERIALS AND METHODS
Design - Prospective Experimental case study.
Setting - Paediatric Surgery Department of a Tertiary Care Hospital.
Study Period - January 2010 to December 2011.
Participants - Neonates and Infants with Anorectal Malformations.

RESULTS
20 babies with intermediate and high anorectal anomalies which had under gone primary PSARP without colostomy were included in the study. Mean operational time was 1 hour. Meconuria was present in 35%. 75% had intermediate type and 25% high fistula. Associate anomalies were present in 60% cases. Post-operative early complications were seen in 30% and late complications in 28.5% cases. One (5%) died due to septicaemia and possible cardiac anomaly. No one suffered from true incontinence after operation.

CONCLUSION
Primary PSARP is an ideal treatment in male babies with high and intermediate ARMs.

KEYWORDS
ARM (Anorectal Malformations), PSARP (Posterior Sagittal Anorectoplasty), VACTREL Anomalies.


BACKGROUND
Anorectal malformations (ARMs) are frequently encountered anomalies that represent an important component of paediatric surgical practice. In some children these defects are associated with lifelong sequelae such as faecal and urinary incontinence. During the last two decades significant advances have occurred in the management of ARM with the pendulum swinging towards good quality of life following definitive repair.

The overall incidence of ARM is 1 in every 2500-5000 live births and the risk of recurrence in future pregnancy is 1%. Chattjee has cited an incidence of 1 in 1862 live births in Calcutta. Males are affected more commonly (57%) than females (43%). The exact aetiology of ARM is not known. The most common associated malformation is of genitourinary system with an incidence of 21-61%. As many as 45% of the patients had sacral abnormalities, other abnormalities are GI anomalies and Cardiac defects. ARM may be part of VACTREL (Vertebral defect, Anal atresia, Cardiac defect, Tracheo-oesophageal fistula, Renal anomalies and Limb abnormalities) anomalies.

Penna and deVries 1st introduced Posterior Sagittal approach in 1982. Prior to Posterior Sagittal approach, the intrinsic anatomy of these defects was not known, and the surgical repair involved several blind steps. 90% of male patients with ARM can be repaired through Posterior Sagittal approach without opening the abdomen. Laparoscopy is an advantage for 10% of patient who suffer from Recto-bladder neck fistula.

The current goals in the management of these defects are (a) to anatomically reconstruct all malformation; (b) to recognize and treat any associated defects that may be life threatening and (c) to treat the functional sequelae of the...
malformations, in order to provide good quality of life. Patients with perineal fistula (Low ARMs) are repaired primarily in newborn period by Anoplasty without colostomy. The standard care for intermediate and High ARMs involved staged procedure consisting of diverting colostomy soon after birth, Posterior sagittal anorectoplasty (PSARP) at around 3 months of age and colostomy closure subsequently.

In recent year, based on observations of Freeman, Thomas Moore, there is tendency among Paediatric Surgeons to repair the defects earlier in life without a protecting colostomy. According to Freeman, anorectum must be in a position as early as possible so that appropriate stimulation can reach the cortex during the critical stage of development of synaptic connections and this can be achieved by performing the definitive surgery as soon after birth as possible. Moore has described that the “Single stage operation at birth relieves Gastro Intestinal tract Obstruction, eliminate urinary tract contamination through fistula, established anorectal continuity, provides maximum potential for defecation reflex at birth and achieves all these in one rather multiple operations.”

Other advantages of primary PSARP in neonatal period are:

- Elimination of colostomy associated complications i.e. diarrhoea/ dehydration, skin excoriation, prolapsed, stenosis, retraction, malfunction, parastomal hernia, intestinal obstruction, wound dehiscence, necrotizing enterocolitis, infection and chronic blood loss leading to anaemia, malnutrition and growth retardation.
- Only one operation and anaesthesia instead of three.
- Reduction in hospital stay and visits.
- Reduction of economic burden of the patient’s family and health care system.
- Decreased psychological burden to the family
- No abdominal scar.
- No need for tapering of rectal pouch.

The incidence of colostomy related complications ranges from 28% to 74%. Formation of colostomy for ARMs is not a minor procedure. A study from Nigeria showed that only 38.5% and 47.4% of male and female patient respectively lived to have definitive operations after preliminary colostomies.

Short term results of Primary PSARP are good and even long-term outcome is shown to be encouraging, but further studies are required before considering PSARP as a standard procedure for Intermediate and High ARMS. The scenario in India is comparable to that of Nigeria. In our country the relative incidence of ARMS is more, and the majority of population are from low income group and from remote places. Colostomy is socially unacceptable and even when performed is associated with high mortality and morbidity, because most parents are illiterate and cannot manage the stoma and colostomy bags are not available easily. There is high incidence of diarrhoea in children in our country and patients with colostomy in situ are very fragile and deteriorate rapidly if treatment is delayed. Significant numbers of patients are lost to follow up (around 40%).

Perhaps because of mortality (colostomy related) or due to unaffordability of a second operation.

Primary PSARP can effectively solve the problems, cut down the cost and stress on the family with a single hospital admission rather than three. It is a milestone in management of ARMS especially in developing countries like India. However, experience in Primary PSARP in India are limited. This study is being carried out to assess the morbidity, mortality and short-term outcome of Primary PSARP in male babies with high and intermediate ARMS.

Aims and Objectives
To assess the morbidity, mortality and short-term outcome of primary PSARP in male babies with high and intermediate anorectal malformations.

MATERIALS AND METHODS
The study was a prospective Experimental study conducted over a period of two years at the Department of Paediatric Surgery, S.V.P.P.G.I.P, S.C.B. Medical College, Cuttack, Odisha. Twenty cases were included for the study. Duration of study from Jan 2010 to Dec 2011.

Subjects to Study
All male patients with high and intermediate ARMS as per Wingspread classification (i.e. lower limit of rectal pouch seen above the PC line or lying between PC line and Ischial point in prone cross- table lateral X-Ray of Pelvis) were enrolled for the study.

Preoperative Assessment
All the patients were subjected to detailed history and a through clinical examination. Attention was given to find out the associated anomalies and rule out low ARMS.

All the patients were kept nil orally with nasogastric decompression, intravenous fluids and intravenous antibiotics (3rd generation cephalosporin + metronidazole + netilmicin).

Laboratory tests like Hb%, Serum electrolytes, Serum bilirubin, Abdominal X-ray AP view, Prone cross-table lateral X-ray of Pelvis, USG of abdomen and Pelvis, were routinely done.

Inclusion Criteria
All full-term male patients with high and intermediate anorectal malformation (ARM) having birth weight 2.5 kg and above were included in the study.

Exclusion Criteria
Patients who were critically ill or having pouch colon association and presence of associated anomalies like oesophageal atresia were excluded from the study. In addition, patients with very high pouch (pouch above sacral promontory) on cross table prone lateral X-ray were also excluded from the study.
Plan of Surgery
All the patients underwent Primary PSARP of DeVries and Pena\(^6\) without colostomy under general anaesthesia. Fistula ligation was done as per the recommendation of DeVries and Pena. Rectal pouch was well mobilized, and a tension free anastomosis with anal skin was done. Intra operative complications were noted.

Postoperative Care
Postoperatively all the Babies were nursed in lateral position and kept on IV fluids and nil orally for 24–48 hours. Same intravenous antibiotics were continued till post-operative day 7. Once the baby started passing meconium, wound was flushed with povidone iodine solution and povidone iodine ointment was applied locally. Per urethral catheter was kept for 7-10\(^{th}\) postoperative day. Early postoperative complications like perineal excoriation, wound infection, wound dehiscence, neurogenic bladder were noted.

Follow up Assessment
Study subjects were assessed at 3, 6, and 12 months Postoperatively with a minimum follow up of 3 months. Assessment was made for complications (stenosis of neoanus, prolapsed of the pulled through bowel), appearance of neoanus, constipation and the bowel movement. Assessment MRI of pelvis was made in few cases to note the quality of muscle complex and levator ani and its relation to pull through bowel. Duration of hospital stay, expenditure and classification. Out of 20 cases, 12 cases (60%) had associated anomalies other than ARM. Eight (40%) had vertebral anomalies, i.e. absence of one or two pieces of sacral bones. Six cases (30%) had genitourinary anomalies. Two had bifid scrotum and one had non-hypospadias, two had hypospadia and one had non-visualization of left kidney. Two patients had both genito-urinary and vertebral anomalies associated with ARM, i.e. VACTREL association (at least 3 components). In follow up, hydronephrosis resolved in both the cases. Incidences of associated anomalies in relation to type of ARM was all 5 cases (100%) of high ARM had associated anomalies and 7 out of 15 cases (46.66%) of intermediate ARM had associated anomalies.

Intra-operatively muscle complex were assessed according the appearance and construction on electrical stimulation by AIIMS muscle stimulator. Among 20 cases, 10 cases (50%) had good muscle complex (well defined vertical and parasagittal fibers with good contraction), 8(40%) had fair and 2 (10%) had poor muscle complex (Only few muscle fibers with weak contractions). In two patients per urethral placed catheter was lying in the rectal pouch and faced difficulties in negotiating the catheter through fistula proximally into the bladder but possible by rail-road method. Intra-operatively two cases (10%) had urethral injuries while separation of common wall and were repaired with Vicryl. In those cases, per urethral catheter was removed on post-operative day 14, and both the patients passed urine
constipated cases become symptom free with dilatation. of the 12 cases (85.71%) cases used to pass stools normally. 4(28.57%) cases soiling only during stress, and 2 (14.29%) times or more per day.

All the parents were asked for the number of stools baby was passing per day, and noted. 6(42.86%) cases were looking neo for perineal appearance of neo and perineal scarring. 10(71.42%) cases had normal cardiac anomaly.

The reason for prolonged hospital stay was wound gaping in 3, transient neurogenic bladder in one. Duration of hospital stay ranged from 2 to 24 days with a mean of 10.28 days. One patient expired on Post-operative day 2, 4 patients stayed more than 2 weeks in the hospital.

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Kiesewetter & Chang functional scoring at each follow-up showed that there was an increasing trend of improvement in faecal continence with time.

In addition to assessment by functional score, MRI of pelvis was done in four cases to note the relation of pulled through bowel with muscle complex and levator ani. In all four cases, it was noted that quality of muscle complex and levator ani was good and bowel was properly pulled through it.

Faecal continence was assessed according to Kiesewetter & Chang scoring. Among 14 cases with a minimum of 3 months follow up Kiesewetter & Chang scoring was good in 8 (57.14%), fair in 6(42.86%). No patient scored as poor.

<table>
<thead>
<tr>
<th>KCFS</th>
<th>At 3 months</th>
<th>At 6 months</th>
<th>At 1 year</th>
</tr>
</thead>
<tbody>
<tr>
<td>Good</td>
<td>8(57.14%)</td>
<td>8(72.72%)</td>
<td>5(83.34%)</td>
</tr>
<tr>
<td>Fair</td>
<td>6(42.86%)</td>
<td>3(27.28%)</td>
<td>1(16.66%)</td>
</tr>
<tr>
<td>Poor</td>
<td>Nil</td>
<td>Nil</td>
<td>Nil</td>
</tr>
<tr>
<td>Numbers</td>
<td>14</td>
<td>11</td>
<td>6</td>
</tr>
</tbody>
</table>

Table 1. Kiesewetter and Chang Functional Scoring at Each Followup

Discussion
Previously mortality rate was very high because of non-availability of advanced care units, experienced anaesthetists, better anaesthetic drugs and paediatric surgical care. Now the scenario is different due to availability of all the above. Primary PSARP has many advantages over staged procedure. Inspite of all its advantages, this procedure cannot be carried out in all babies.

Indications for primary PSARP in male babies are any newborn with intermediate and high anorectal malformation. Absolute contra indications are presence of a fatal associated anomaly, pouch colon, and a very high pouch. Relative contraindications are very sick babies, sacral agenesis, low birth weight and prematurity. Cases with very high pouch, very sick babies, low birth weight (<2.5 kg), premature babies were treated by neonatal colostomy (staged procedure) at same time period. Pouch colons were done by excision of pouch, tuboplasty and end colostomy. Comparison of different one stage PSARP studies
In this study, minor complications were noted in 30% with superficial wound gaping being the commonest. In other studies, postoperative complication rates varied from 6.25 to 70%. All superficial wound gaping healed with flushing the wound with povidone Iodine Solution and local application of povidone iodine ointment. One patient with deep wound gaping also responded to conservative treatment and doing well with 2 to 3 stools per day and no soiling and even the perineum has healed without scarring. In 1 case with perianal excoriation Zinc Oxide cream was applied liberally and improved with time. The perianal excoriation is caused by frequent passage of loose stool with suture line, resulted in lysis of suture material (Vicryl) and wound gaping. Intra-operative injuries (urethral injuries) occurred in 2(10%) cases in this study. Mishra B N et al documented real incontinence in 4.14% cases. Xiao Song Xu et al have documented soiling in 6.25% (3/48). In the present study, patients were not advised for routine anal defecation under anaesthesia. In this study, patients were not advised for routine anal defecation under anaesthesia, with the conception that by doing primary PSARP, early establishment of anocutaneous neuro-circuitry defection reflex (Moore’s hypothesis) and stool passing through the neo anus itself act as dilator and by doing tension free anastomosis to the perianal skin. In this study among 14 cases, one developed neo anus stenosis and responded well to anal dilatation.

Incidence of constipation in the present study was in 14.29% (2/14), while Liu et al documented constipation in 44.61% (29/65) and Xiao Song Xu et al have documented constipation in 6.25% (3/48). In the current study, it had been noticed that both cases who had constipation had superficial wound gaping leading to fibrosis. Incidence of soiling in this study was 42.86% (6/14), mucosal prolapse in 21.43% (3/14) and no case suffered from real incontinence. Liu et al have documented soiling in 50.8% (33/65) cases, rectal prolapse in 7.6% (5/65) and 6.15% (4/65) had real incontinence. KLN Rao et al have documented real incontinence in 4.14% cases. Xiao Song Xu et al documented soiling in 8.33% (4/48), mucosal prolapsed in 4.16% (2/48) and none of their patient suffered from real incontinence. In the present study, among 6 cases those who had soiling, 3 had wound dehiscence, 3 had fair muscle complex and mucosal prolapse. From the observation of the current study, it can be inferred that incidence of soiling can be reduced by preventing wound dehiscence. Another factor contributing to soiling and mucosal prolapse was fair muscle complex which is non-modifiable.

In the present study, faecal continence was assessed according to Kiesewetter & Chang functional scoring (KFPS) with a minimum period of 3 months follow up. The continence at last follow up among 14 cases were good in 8 cases (57.14%), fair in 6 cases (42.86%) and no case suffered poor continence. There was improvement in the continence rates with time. The achievement of good continence score of 83.34% and fair in 16.66% patients at

<table>
<thead>
<tr>
<th>Study</th>
<th>No. of Cases</th>
<th>Age Group in Days</th>
<th>Weight in kg.</th>
<th>Complications (%)</th>
<th>Mortality (%)</th>
<th>Intra-op Injury (%)</th>
<th>Hospital Stays in Days</th>
<th>Functional Score</th>
</tr>
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<tbody>
<tr>
<td>Moore TC et al (1990)</td>
<td>4</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NIL</td>
<td>NA</td>
<td>NA</td>
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<tr>
<td>Goon HK et al (1990)</td>
<td>32</td>
<td>4</td>
<td>2.1-3.8</td>
<td>57.58</td>
<td>Nil</td>
<td>Nil</td>
<td>14-28</td>
<td>90.9</td>
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<tr>
<td>Albases CT et al (1999)</td>
<td>5</td>
<td>&lt;2</td>
<td>2.7-4.4</td>
<td>20</td>
<td>Nil</td>
<td>Nil</td>
<td>7</td>
<td>NA</td>
</tr>
<tr>
<td>Adeniran JO et al (2005)</td>
<td>13</td>
<td>2-4</td>
<td>NA</td>
<td>30.77</td>
<td>NA</td>
<td>NIL</td>
<td>19</td>
<td>NA</td>
</tr>
<tr>
<td>Gangopadhyay AN et al (2006)</td>
<td>429</td>
<td>3.1</td>
<td>NA</td>
<td>17.35</td>
<td>4.5</td>
<td>2.65</td>
<td>11.2</td>
<td>68</td>
</tr>
<tr>
<td>KLN Rao (2007)</td>
<td>30</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>10</td>
<td>-</td>
<td>8.33</td>
<td>79.12</td>
</tr>
<tr>
<td>Xiao Song Xu et al (2010)</td>
<td>48</td>
<td>NA</td>
<td>NA</td>
<td>6.25</td>
<td>2.08</td>
<td>NA</td>
<td>NA</td>
<td>100</td>
</tr>
<tr>
<td>Present Study</td>
<td>20</td>
<td>1-7</td>
<td>2.5-3.7</td>
<td>30</td>
<td>5</td>
<td>10</td>
<td>10.28</td>
<td>83.34</td>
</tr>
</tbody>
</table>

Table 2
one year of age is comparable with Ong NT and Beasley SW\textsuperscript{23} & KLN Rao.\textsuperscript{21}

The incidence of continence widely varies from one study to another, Goon HK et al\textsuperscript{16} reported good faecal continence in 90.1%, fair in 9.9% and poor in nil, because among 32 cases only 12 were male and rest were females. As continence rates are better in females because of more low variety of anomalies which has been reported by Ong NT et al.\textsuperscript{21} Gangopadhyay et al\textsuperscript{10} reported a good score in 68%, fair in 22% and poor in 10% cases inspite of excluding patient with sacral agenesis and meningomyelocele. KLN Rao\textsuperscript{21} reported good score in 79.1%, fair in 16.67% and poor in 4.16%.

The present study recorded more number of good and fair scoring in average and none of the cases suffered from real incontinence which is far better when compared to other studies. In addition to functional scoring, MRI of pelvis is performed in three cases. In all three cases muscle complex and levator ani were good and bowel was properly pulled through it.

In this study, the average duration hospital stay was 10.28 days. The average duration of hospital stay reported by Moore TC\textsuperscript{10} et al was 7 days, by Adeniran JO et al\textsuperscript{16} was 19 days, Gangopadhyay AN et al\textsuperscript{10} was 11.2 days, Goon HK et al\textsuperscript{17} was 2 to 4 weeks and KLN Rao\textsuperscript{21} was 8.33 days. In this study 6 cases stayed for more than 2 weeks, because of intra-operative urethral injury in 2 cases (urethral catheter was kept for 2 weeks), wound gaping in 3 cases and transient neurogenic bladder in one case. Other cases were discharged in 7 days.

Primary PSARP is better than conventional staged procedure in all means. Scientifically it is based on concept of Freeman NV and Bulut M.\textsuperscript{9} According to Freeman, anorectum must be in position as early as possible so that appropriate stimulation can reach the cortex during the critical stage of development of synaptic connections and this can be achieved by performing the definitive surgery as soon after birth as possible. Moore has described that the "Single stage operation at birth relieves gastro Intestinal tract obstruction, eliminates urinary tract contamination through fistula, establishes anorectal continuity, provides maximum potential for defeaction reflex at birth and achieves all these in one rather multiple operations". In this study, mortality was low; most of the complications were managed effectively without any residual effects with a good and fair continence score. None of the patient suffered from real incontinence.

CONCLUSION
Primary PSARP is the ideal treatment option in male babies with high and intermediate ARM. It is safe and feasible with superior results in term of mortality, morbidity, continence and cost.

- Twenty male babies with intermediate and high anorectal anomalies had undergone primary PSARP without colostomy in a prospective study over 2 years.
- Birth weight ranged from 2.5 kg to 3.7 kg in 90% case, in two cases those presented late i.e. one at 2 months weighed 4.2 kg and other at 5 months weighed 4.5 kg.
- Eighteen cases (90%) underwent surgery within 24 hours of admission. With a mean operating time of one hour.
- Meconuria was seen in only 7 cases (35%) out of 20 cases.
- Out of 20 cases 15 cases (75%) had intermediate type and 5 cases (25%) had high fistula.
- Out of 20 cases, 19 cases had recto-urethral fistula (7 recto-prostatic, 12 recto-bulbar) and one had no fistula.
- Associated anomalies were seen in 12 (60%) cases.
- Post-operative early complications were seen in 6 cases (30%). Superficial wound gaping (15%) was the common early complications.
- Late complications were seen in 4(28.5%) cases. Partial mucosal prolapse (18.75%) was the common complication.
- Mortality in primary PSARP was 5%.
- Among 14 cases of regular follow-up, 42.86% cases had 1-3 stools per day, more than half of cases have not suffered from faecal soiling and none suffered from urinary incontinence.
- Faecal incontinence was good in 8(57.14%), fair in 6(42.86%) and none had true incontinence.
- Mean duration of hospital stay was 10.28 days.

REFERENCES

Original Research Article


