Primary Repair of High and Intermediate Anorectal Malformations in the Neonates

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Background/Purpose: The standard treatment of high and intermediate anorectal malformation (ARM) is the staged approach. A growing interest in one-stage correction of ARM was noted recently. The aim of this study was to examine the feasibility, safety, and short-term outcome of primary repair of high and intermediate ARM in neonates.

Materials and Methods: This prospective study included 38 full-term infants (22 females and 16 males) with either high (n=9) or intermediate (n=29) ARM treated during a 6-year period. All patients underwent posterior sagittal anorectoplasty (PSARP) without colostomy. The exclusion criteria included: prematurity, associated major congenital anomalies, bad general condition, persistent cloaca in females and unfavorable circumstances. Preoperative imaging using ultrasonography and magnetic resonance imaging (MRI) was performed to assess the severity of ARM and any associated anomalies. All patients were evaluated as regard to operative details, postoperative complications, and bowel function on short and long-term follow up, which ranged from 2 to 62 months.

Results: All patients were treated entirely through the posterior sagittal approach except one male patient with rectovesical fistula, who required laparotomy. Intraoperative complications included: opening the posterior wall of the vagina (n=6), injury of the seminal vesicle (n=1). Postoperative complications included: wound infection in 9 patients, one of them required colostomy and 2 required secondary sutures. Postoperative anal stenosis occurred in five patients, all treated with anal dilatation. Twenty-five patients defecate spontaneously, 9 require oral medication or rectal stimulation or enemas occasionally, and three are maintained on bowel program. Eight of the 10 females older than 3 years of age are continent, while only 5 of the 8 male patients older than 3 years are continent with infrequent soiling. One patient had anesthetic complications and died 5 days after surgery.

Conclusion: 1. One-stage repair of intermediate and high ARM both in male and female neonates is technically feasible; 2. The safety of this approach depends on adherence to strict inclusion criteria; 3. The early postoperative complications are acceptable and can be managed successfully; 4. The functional results are comparable to those reported in patients undergone the standard staged technique.

Index Word: Anorectal Malformations, Primary Repair, Neonates

INTRODUCTION

The standard treatment of high and intermediate anorectal malformation (ARM) is the staged approach, which entails performing colostomy shortly after birth followed by posterior sagittal anorectoplasty (PSARP) few weeks or months later, and finally colostomy take down as a third stage.1,2 PSARP procedure has the advantage of direct visualization of the striated muscle complex that could assist in the correct positioning of the anorectal tube at the exact anatomical position.3
Many authors recommended the formation of initial colostomy for those children not only for colon decompression, but also to diminish the frequency of infection and breakdown in the subsequent definitive operation and to allow better radiologic study of the malformation. Recently, many surgeons have argued for a definitive repair in the neonatal period without colostomy.

The aim of this study was to evaluate the feasibility, safety and outcome of one-stage PSARP in cases of high and intermediate ARM in neonates.

**MATERIALS AND METHODS**

Thirty eight full-term infants with high (n=9) or intermediate (n=29) ARM were included in this prospective study, which was conducted at the department of pediatric surgery in Tanta University Hospital, Ain Shams University Hospital and affiliated hospitals during the period from October 2000 through February 2006. This group of patients included 22 (57.9%) females and 16 (42.1%) males. Nine of the 16 males were of high type, while 7 were of the intermediate type. All females were of the intermediate type, 21 of them had rectovestibular fistula, while one had imperforate anus without fistula. The types of anomalies were classified according to the Wingspread International Classification of Anorectal Malformation. The exclusion criteria included: associated major congenital anomalies, bad general condition, prematurity, some complex anomalies as persistent cloaca or cloacal extrophy.

All patients undergone complete physical examination for evaluation of the buttock contour, contraction of the anal dimple, the presence of fistula, and presence of other major congenital anomalies.

An invertogram was performed to show the level of the rectal pouch and any sacral anomalies. Transperineal and abdominal ultrasonography were used for determining the level of rectal pouch and for exclusion of associated congenital anomalies (fig.1). Other imaging studies such as MRI was performed whenever feasible for better anatomical visualization of the site of the fistula, the sphincter complex and the level of the rectal pouch (fig.2).

**Operative technique:**

Following preoperative assessment and preparation, all patients had undergone the standard PSARP approach. Preoperative bowel preparation was carried out through the fistula in females with rectovestibular fistula; otherwise the meconium was evacuated during operation in all other patients. Cephalosporin and metronidazol were given before induction of anesthesia and continued postoperatively for 5 days. The details of operative technique were described by De Vries and Pena. During operation all patients were catheterized and put in prone position. Posterior sagittal incision was done (fig.3). Rectal pouch was opened between stay sutures. Gas and feces were suctioned as much as possible. The rectal pouch was cleaned thoroughly with betadine solution (fig.4). The rectal pouch was not tapered. The perineum was closed and anoplasty was performed around size 6 Hegar’s dilator. The operation was accomplished entirely through a posterior sagittal approach in all patients except one with high-located rectum ending at posterior wall of the urinary bladder (Fig 5 A&B), who required combined posterior sagittal and abdominal approach.

Breast feeding or artificial feeding was allowed on the 2nd postoperative day. Infants with urinary tract fistula had a urinary catheter left for 5-7 days.

All patients had regular anal dilatation up to size 9 Hegar’s dilator starting 10 days after surgery. They were followed up for a period ranged from 2 - 62 months. During follow up the patients were evaluated as regards to the presence of postoperative anal stenosis, any urinary problems, and for the degree of continence.

The anorectal functional results were designated according to the modified Wingspread scoring as excellent, good fair, and poor. Continence was defined as the ability to stay clean without staining or soiling both day and night without pads or diapers. Soiling was defined as an inadvertent loss of small amount of feces staining the underwear. Incontinence was defined as regular loss of solid feces. Constipation was defined as less than 3 spontaneous bowel movement per week, rectal fecal impaction or abdominal fecal mass.
Fig 1. Transperineal ultrasound shows high ARM, the rectal pouch was located 26 mm from the skin of perineum.

Fig 2. MRI Sagittal plane shows the termination of rectal pouch just above pelvic floor with recto-bladder neck fistula.

Fig 3 A. Clear visualization of both superficial muscle and muscle complex in a male neonates with high ARM.

Fig 4. Intra-operative meticulous evacuation of gas and meconium.

Fig 5 A. High ARM with rectovesical fistula. Combined posterior sagittal and abdominal approach was needed.

Fig 5 B. Operative photo shows the termination of rectum to the posterior wall of the urinary bladder.
RESULTS

The definitive repair was accomplished entirely through the posterior sagittal approach in all patients except one with rectovesical fistula who needed laparotomy to separate the fistula and for mobilization of the rectum and sigmoid colon. The repair was done within the first 72 hours in all males and in 8 of the 22 females. In the remaining 14 females decompression of the bowel was carried out through the rectovestibular fistula for 3-4 weeks then surgery was carried out.

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The mean operative time was 140 ± 20 minutes for male patients, and 110 ± 15 minutes for the females. The posterior wall of the vagina was opened in 6 cases during the separation of the rectum from the vagina; these minor openings were repaired without any postoperative consequences. Injury of the seminal vesicle occurred in another male patient. No other significant technical difficulties or intraoperative complications were met during the rectal mobilization. No blood transfusion was needed.

Postoperative wound infection occurred in 9 patients (23.7%) one of them required colostomy and 2 required secondary sutures. The other 6 patients were treated conservatively with local wound dressing and parental antibiotic therapy. One patient had anesthetic complications and died 5 days after surgery.

All patients (except one) passed stool within 24 hours after surgery and tolerated full enteral feeding within 3-5 days. The hospital stay ranged between 4 - 15 days (median 5 days). Thirty-four of the 37 survived patients have a strong urinary stream; three had variable degrees of neurogenic bladder.

Regular postoperative dilatation was started 10 days postoperatively. The regimen for dilatation was as described by Pena. Five patients developed anal stenosis secondary to parental non-compliance with postoperative dilatation regimen, these responded to frequent anorectal dilatation.

Twenty-four of the survived 37 patients have spontaneous evacuation of the stool. Variable degrees of constipation occurred in 12 patients (32.4%), 9 of them were females. Six patients required laxatives occasionally, 4 required laxative and occasional enemas and 3 required regimen of enema program.

Three male patients out of 8 and 2 female patients out of 10 older than 3 years have incontinence. Incontinent patients were treated conservatively. Two patients improved with time and the other 3 patients had some improvement on enema program and biofeedback therapy.

DISCUSSION

The major objectives in the management of ARM are the relief of intestinal obstruction if present, restoration of anorectal continuity with optimal sphincter function, early postnatal establishment of the brain-defecation reflex, reduction of the physical and psychological stress to the patient and his family. All these should be done with insurance of cost effective care of the patients.
Since 1980s PSARP operation has become the slandered treatment for high and intermediate ARM.\textsuperscript{10} Though the postoperative outcome of ARM has been improved significantly after PSARP, these 3 stage-operations are of immense disadvantage to the patients and the parents. The cost is significant and the incidence of complications is considerable.

There are a variety of complications related to colostomy performed in infancy reported in the literature. Common complications include colostomy prolapse or retraction, skin dehiscence and excoriation, intestinal obstruction and stomal ulceration and bleeding.\textsuperscript{11-14} Patwardhan et al reported 32-57\% complications of colostomy in their series, 32\% were due to mechanical complications related to colostomy formation.\textsuperscript{11} Urinary tract infection was observed after colostomy in 29\% of infants.\textsuperscript{11} Nour et al reported an incidence of 28-74\%,\textsuperscript{12} colostomy-related complications. Likewise, Liu et al reported 39.6\% complications in their patients in whom there were more than one complication as stoma prolapse, skin dehiscence and urinary tract infection.\textsuperscript{8} The reported frequency of complications after closure of colostomy are also significant.\textsuperscript{12} A significant incidence of wound infection, incisional hernia, and adhesive intestinal obstruction have been reported after colostomy takedown.\textsuperscript{11}

The advantages of bypassing the colostomy stage are many. First, the above mentioned colostomy complications are eliminated completely which is specifically important in developing countries where the idea of colostomy is socially unacceptable, beside the shortage of stoma nurses and deficient parents knowledge about colostomy care.\textsuperscript{15} Having a colostomy offers the advantage of performing distal loop colostogram to distinguish between various types of malformation, to localize the location of the rectal pouch and to show the different sites of fistulae. But in the current series we found that MRI was very useful in preoperative evaluation of those patients with minimal X-ray exposure.

Liu et al raised a question about the safety and feasibility of one stage PSARP. They had no operative mortality in their series of 65 neonates.\textsuperscript{8} Coon's also had no operative mortality in their 32 patients.\textsuperscript{7} In the current series we had one postoperative mortality due to anesthetic problems. We believe that strict adherence to the exclusion criteria is mandatory to ensure the safety of this approach in our institutions.

As regard the feasibility, this one-stage approach was technically feasible in all cases in the current series either through an entirely posterior sagittal approach or through combined posterior sagittal and abdominal approach. Liu et al reported 2 cases of conversion to Laparotomy one with rectovesical fistula with too high rectal pouch and the other with rectoprostatic fistula with torn rectum. They recommended extension of the upper incision to 2-3 cm higher than the level of the coccyx and the lower incision to 0.5-1 cm higher than the level of the anal dimple and removal of the coccyx to liberate and facilitate pull through of the rectum and to decrease the incidence of conversion to Laparotomy.\textsuperscript{8} On the other hand, Pena warned against attempting to find the very high rectum and dissect it through the posterior sagittal approach for fear of injury of the seminal vesicle, vas deference, prostate and bladder innervations.\textsuperscript{16} In this series we had injury to the seminal vesicle in one patient with very high rectum. This might have been avoided if combined transabdominal and posterior sagittal approach was used in that particular case.

In our study postoperative wound infection occurred in 9 out of 38 patients (23.7\%) one of them required colostomy and the other 2 required secondary sutures. It seems that colostomy can help diminish the frequency of wound infection.\textsuperscript{8} So if we choose to do one-stage PSARP every effort should be done to diminish the incidence of infection by good colonic irrigation and meticulous tissue handling. Beside the operation should be done by well-experienced neonatal surgeon. Also the infants should have good postoperative nursing facilities. One of the methods to decrease the incidence of wound infection is to nurse the patient in prone position after surgery to minimize soiling of the wound by the stool. This approach was adopted in the last 6 patients with very encouraging results where no wound infection occurred in this group of patients.

Recent theories suggest that the neuronal framework for normal bladder and bowel function exists at birth. But there is learning or training period in which long-lasting, activity-driven neuronal changes take place during neuronal circuitry development.\textsuperscript{6} Moor highlighted the importance of restoring gastrointestinal continuity in the neonates to establish brain- defecation reflexes early.\textsuperscript{6} Likewise, Albanese et al reported that early restoration of gastrointestinal continuity would train the perineal
musculature and improve long term fecal continence. So delayed repair of anorectal anomaly will result in loss of critical time needed to form neuronal network and synapses resulting in normal or near normal function. Therefore, at least theoretically, the earlier the definitive operations are done, the higher the chances of achieving continence. Liu et al didn’t note any significant difference in the postoperative bowel habit in their comparative study between one-stage and three-stage PSARP. In the current study 3 patients (7.9%) had constipation that responded only to enema program and 5 patients who are older than 3 years are incontinent. Two of them showed gradual improvement. We agree with Templeton and Dietcheim that there is a gradual improvement of continence with age. Although we noted some improvement of continence in the other 3 patients after enema program, regulation of the diet and biofeedback therapy, but as mentioned by Gangopadhagy et al, we do not know how much the degree of improvement and how long it will take to achieve normal bowel habit. We believe that we may need a large number of patients and longer period of follow up to prove that.

CONCLUSIONS

Primary repair of intermediate and high ARM in male and female neonates is technically feasible and safe. It avoids the risk of complications related to colostomy, the risks of multiple anesthesias and reduces the economic and psychological burden to the family.

Acknowledgement:

The author thank Hisham Elsafory, MD, Professor of Pediatric Surgery, Ain Shams University, for permission to include 6 patients from his institution in this study, and Amel Hashish, MD, Lecturer of Pediatric Surgery, Tanta University for her assistance in preparing and reviewing this manuscript.

REFERENCES