Rectal Atresia: Multiple Approaches in Neonates

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Abstract

Aim: The aim of this study is to present multiple approaches in management of rectal atresia in neonates and to discuss the outcome of each approach.

Patients and Methods: Between 2002 to 2010, a prospective study of 15 patients with rectal atresia; they were treated at Pediatric Surgical unit at Children University Hospital. They were segregated into three groups according to surgical approach that was carried out. First group (five neonates) was subjected to three-stage procedures. Second group (five neonates) was subjected to neonatal transanal end-to-end rectorectal anastomosis. Third group (five neonates) was subjected to neonatal mini PSARP.

Results: All children were presented with rectal atresia. Their age was ranged from one to five days. The male to female ratio was 7 to 8. 13 out of them were full term with normal vaginal delivery. The remaining 2 patients were preterm. Three out of 15 patients had other anomalies. Intraoperative complications were nil in three groups. Second and third groups had less hospital stay. Two patients developed wound infection in first group. Anal stenosis occurred in one patient of second group. One patient developed fecal discharge from 3rd group and treated conservatively. All patients were subjected to postoperative follow-up program for at least six months. The parents of the second and third groups were satisfied regarding one stage procedure in addition to fecal continence of their infants.

Conclusion: Many approaches could be carried out for surgical management of rectal atresia. Neonatal approaches could be applied easily and safely, with less cost-effect. Mini PSARP is considered a less traumatizing procedure to anal sphincter with easy anastomosis that was carried out under vision.

Key Words: Rectal atresia – Neonates.

Introduction

RECTAL atresia; is an extremely rare anorectal malformation, is predominant in male with an incidence of 1%-2% in all anorectal anomalies [1]. The most common types are complete fibrous membrane or severe stenosis between distal rectum and anal canal [2]. The exact pathogenesis of rectal atresia is unknown, but it is postulated to be an acquired anomaly arising from intravascular thrombosis secondary to intrauterine infection [3,4].

Rectal atresia has been classified as type IV using the Ladd-Gross classification [5], and as a separate high or intermediate variety using the International classification of ARM and under the group of rarities by Wingspread classification [6]. Depending upon the distance between the proximal rectum and distal anorectum, four grades have been described: Grade 1- rectal atresia with a short gap between each (most common), grade 2- rectal atresia with a long gap, grade 3- membranous septal type, grade 4- rectal stenosis. Grade 2 rectal atresia has not been found in clinical practice, even in area with a very high incidence of this anomaly [7]. Moreover, recently a few cases of double rectal atresia (multiple) have been reported [8].

In this anomaly, the anal canal, external sphincter, and internal sphincter are well developed, and usually, there is no fistula between rectum and the urethra or vagina. In the surgical management of this condition, an ideal operation should preserve and use those normal elements to achieve postoperative fecal continence, so many approaches had been developed to reach optimum results [2,6-10].

The aim of this study is to present multiple approaches in management of rectal atresia in neonates and to discuss the outcome of each approach.

Patients and Methods

Over a period of eight years between 2002 and 2010, fifteen children were managed at Pediatric Surgery unit at Assiut University Hospital. Children with other anorectal malformation were excluded from this study. The patients were referred to our hospital age ranged from 1 to 5 days. Data collected
included; clinical presentation, age at diagnosis, associated congenital anomalies, age at surgery, operative technique, operative complications, and anorectal function, were studied and analyzed.

On examination, most of the patients were presented with abdominal distention, sometimes bilious vomiting, and inability to pass meconium. All patients revealed normal anus. Trying to pass small Hegar’s dilator (through the anus) could only be introduced for not more than 2cm. Rectal atresia was suggested in all our cases. Three children had other associated congenital anomalies in form of right kidney agenesis in one child, Down’s syndrome in second child, and cardiac anomalies (PDA and VSD) in third child. All patients were subjected to routine investigations as CBC, Prothrombin time and concentration, and sometime contrast X-ray. Some patients were subjected to additional laboratory investigations according the state of baby. Echocardiography was made if there was any suspicious of cardiac problem. The patients were segregated into three groups according to the type of surgical intervention. First group (five neonates) was subjected to colostomy waiting for definitive surgery. Second group (five neonates) was subjected to transanal end-to-end rectorectal anastomosis. Third group (five neonates) was subjected to mini posterior sagittal anorectoplasty (mini PSARP).

Surgical approaches:

The first group; the patients were subjected to sigmoid colostomy; the surgery was done under general endotracheal intubation anesthesia, performing a simple loop sigmoid colostomy. The patients were sent back home on the second or third day after stabilization of the general condition. Their parents were informed to return back to hospital every month to evaluate the patient and to prepare to definite surgery. Two types of surgical interventions were carried out for those patients (depending upon the surgeon preference). PSARP was performed for four patients of this group at age ranged from 6 to 8 months. One child out of the four patients, developed wound infection and was treated conservatively. The remaining 5th child was subjected to transanal end-to-end rectorectal anastomosis at age of 9 months. Both techniques were performed under general endotracheal intubation anesthesia. The hospital stay ranged from 7 to 9 days in four patients and 4 days for the 5th patient. They were scheduled for follow-up program to perform anal dilation every week for first month, every month for three months. All patients of this group were scheduled for colostomy closure. The third stage colostomy closure was carried out after a period ranged from 6 to 8 months, and the patient stayed at hospital for another 9 to 11 days. One patient developed infection and wound dehiscence at site of colostomy closure and was treated by secondary sutures at 10th day and returned back to home on 11th day.

The second group; the patients were subjected to neonatal definitive surgery in form transanal rectorectal anastomosis. Using the technique that was described by Upadhyaya 1990, in infants and Ibrahim 2006, in neonates, under general endotracheal intubation anesthesia, the patient was positioned in lithotomy position, after anal dilation a transverse incision was done at the dome of the distal rectum, upward genital distal dissection to reach the proximal pouch, which may be plugged with thin membrane due to accumulation of meconium. A transverse incision had been made and the meconium was aspirated to clarify the field. The wall of the proximal rectum was dissected and intusscepted to the distal segment to perform end-to-end rectorectal anastomosis. Hospital stay ranged from 4 to 5 days. Intra-operative or postoperative complications were nil. Postoperative anal dilation was performed after one week, twice per week for one month, once every week for another second month. One patient developed marked rectal stenosis at the age of six month (this patient performed his follow-up program badly), dilation under anesthesia was made and his parents were instructed to repeat his dilation program perfectly.

The third group; the patients were subjected to neonatal definitive surgery in the form of mini posterior sagittal rectorectal anastomosis. This approach was carried out under general endotracheal intubation anesthesia; the patient was positioned in knee- chest position after catheterization. Hegar’s dilator was introduced via anus to facilitate identification of the distal rectum. An incision was performed in exact posterior midline about 2 to 3 cm starting from the tip of the coccyx to 1.5cm above mucocutanous junction to keep the external sphincter intact. The dissection was carried out to reach the both ends of the proximal and distal rectal pouches, then anastomosis was performed by 5/0 absorbable interrupted sutures. A tubal drain was placed in pararectal space and removed on the second day, the muscles re-rapped around the rectum as in Figs. (1-6). Postoperative anal dilation was performed after one week, twice per week for one month, once every week for another second month. One patient developed fecal discharge from the wound on 3rd day, this patient performed daily anal dilation for two weeks and the discharge stopped.
Results

Fifteen children presented with rectal atresia in the study period between 2002 to 2010. Other varieties of anorectal malformation were excluded from this study. Eight out of 15 were females. 13 out of 15 patients were full-term babies and the remaining two patients were preterm 31 ± 2 weeks. The patients were referred at different ages ranged from 1 to 5 days. All patients presented with abdominal distension, no passage of meconium, and vomiting, in addition to normal appearance of anus. Three neonates had other associated congenital anomalies in form of right kidney agenesis, Down’s syndrome, and cardiac anomalies. These associated congenital anomalies did not prevent surgical correction. Three different surgical approaches were carried out in our study for correction. The choice of surgical procedure for correction was haphazardly. The results were collected and analyzed. The age at surgical intervention was determined after establishing the medical state of the child and it ranged from one to two days after admission to the neonatal intensive care unit (NICU).

The patients in this study were segregated into three groups according to the type of surgical approach. No selective characters for each approach. Regarding hospital staying; first group, the patient was admitted three times to the hospital with a total hospital stay ranged from 22 to 25 days. Among that time the patients were subjected to three-time anesthesia and operations. This approach was considered as a time consuming particularly for parents and had a high cost-effective ratio. Moreover, the child was liable to get cross-hospital infection that may add more hospital stay. In second group, the patient stayed at hospital for 4 to 5 days. Anal stenosis in one case was due to his parent’s incompliance and they neglected the postoperative dilation program. These patients were subjected to single surgical procedure with hospital stay ranged from 4-5 days, that was much less than that of the first group with less cost-effective ratio. This approach may be suitable for poor community. In 3rd group, the hospital stay ranged from 7 to 8 days that was more or less similar to that of the 2nd group. One patient developed fecal discharge that improved conservatively.

Regarding postoperative complications, one child developed wound infection in second stage operation and another baby developed wound dehiscence on third stage in first group. Anastomotic leakage occurred in one child of third group and improved conservatively. Anal stenosis was developed in one child from the second group that corrected by vigorous dilation under anesthesia and repeating of postoperative dilation program.

Follow-up program for all patients for six months showed no significant difference regarding
anal function. Furthermore, their parents have been satisfied regarding fecal continence and simplicity of the procedures in second and third groups. In first group?? the families suffered from multiple hospital admissions.

**Discussion**

Rectal atresia is a rare variety of anorectal malformation, which may be passed unnoticed for a time due to normal appearance of the shape and the position of the anus. In our institution the incidence of rectal atresia (4%) is less common than that of the anorectal malformation and this agreed with that of most literatures [2]. The pathogenesis of rectal atresia is unknown and there are many postulations for explanation. The most accepted one; is an acquired anomaly arising from intravascular thrombosis secondary to intruterine infection [34].

Rectal atresia is characterized by the presence of the rectal pouch which usually terminates at, or within, the pelvic diaphragm (pubococygeal-PC-line), and a well formed anus that is in its normal appearance [9].

Dorairajan (1988) classified rectal atresia into four grades; grade I, short gap less than 2cm, was the most common type of his series [7]. In our study, rectal atresia with a complete fibrous membrane (grade III) was encountered in 13 patients (87%), and short gap between the two segments in the remaining two (13%) cases.

Fortunately, most of investigators had an acceptance that the rectal atresia usually has well developed pelvic structures particularly; the anal canal, external sphincter, and internal sphincter. Also the incidence of fistula between rectum and the urethra or vagina is extremely rare, and this was confirmed in our study. The significance of near normal anatomical pelvic structure is to preserve this anatomy to achieve postoperative fecal continence, and this encouraged me to minimize the posterior sagittal incision in 3rd group for more preservation of the anal sphincters.

The incidence of associated anomalies is much lower in cases of rectal atresia when compared to other intestinal atresias [10]. But our study had three (20%) cases with other associated congenital anomalies, this is a relatively high ratio that might be due to a few number of our cases.

In our study, the diagnosis of rectal atresia depended upon the clinical presentation of abdominal distension, vomiting, and failure to pass meconium, in addition to simple rectal examination using rectal dilator. An inverogram with a Hegar dilator in anal canal was the main radiography used to evaluate the distance of separation in our study. Some authors (centers) had used perineal ultrasonography and magnetic resonant image to verify the diagnosis and the exact relation between anal canal and sphincters. Contrast study had been only done for first group by performing distal loopgram to confirm the diagnosis and to exclude presence of fistula.

Rectal atresia is considered as a type of colonic atresia, so it has the same debate over whether primary anastomosis or a staged operation is required [11,12]. Several authors have recommended resection and primary anastomosis as a reasonable treatment option regardless of location of CA if the newborn’s condition permits [13].

Rectal atresia is a matter of surgical interest for pediatric surgeons, and via Medline, Pubmed, Scince Direct researchs we found a very long list of various operative procedures have been reported and described since the 1950 used for the correction of rectal atresia as shown in Table (1).

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Reference</th>
<th>Year</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perforation of membrane and serial dilation.</td>
<td>Brayton et al.</td>
<td>1958</td>
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<tr>
<td>Abdominal pull-through with nonsuture anastomosis.</td>
<td>Nixon</td>
<td>1961</td>
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<tr>
<td>Swenson’s pull-through in the neonatal period.</td>
<td>Synder</td>
<td>1961</td>
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<tr>
<td>Abdomino-perineal pull-through after stripping the mucosa of the distal pouch.</td>
<td>Trusler</td>
<td>1962</td>
</tr>
<tr>
<td>Swenson’s pull-through at age 6-12 months.</td>
<td>Kiesewetter et al.</td>
<td>1964</td>
</tr>
<tr>
<td>Stephen’s sacro-perineal pull-through using nonsuture anastomosis.</td>
<td>Santulli</td>
<td>1965</td>
</tr>
<tr>
<td>Intraluminal anastomosis using special intraluminal anastomosing device.</td>
<td>Santulli et al. [19]</td>
<td>1965</td>
</tr>
<tr>
<td>Duhamel pull-through.</td>
<td>Louw</td>
<td>1965</td>
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<tr>
<td>Swenson’s anastomosis via Stephen’s sacral approach.</td>
<td>Upadhyaya</td>
<td>1978</td>
</tr>
<tr>
<td>String placement through the atresia, followed by progressive dilations using Tucker’s dilators.</td>
<td>Dias et al. [22]</td>
<td>1982</td>
</tr>
<tr>
<td>Sacroperino-perineal or abdomino-perineal pull-through using zig-zag anastomosis.</td>
<td>Japan Study group</td>
<td>1982</td>
</tr>
<tr>
<td>Transanal end-to-end reectorrectal anastomosis (TERA).</td>
<td>Gauderer et al. [23]</td>
<td>1984</td>
</tr>
<tr>
<td></td>
<td>Pena</td>
<td>1993</td>
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</table>
This extensive list of ingenious procedures is testimony to the great difficulty faced in treating this anomaly, moreover, absence of standardized recommendations in literatures encouraged us to perform this study. The aim of our study is to evaluate and modify surgical procedures for correction of the rectal atresia.

In our study, three procedures had been carried out; neonatal one-stage surgery with two different approaches for second and third groups, and multiple-stage procedure for first group. The neonatal procedures were mini PSARP as a modification of Pena’s procedure; in which the midline incision did not reach the anocutanous junction but stopped away (about 1.5cm) from anal verge. The aim of this modification is for more preservation of normal anatomy and to minimize the surgical trauma to the anal sphincters. The procedure was easy and feasible with favorable results.

The second neonatal procedure was transanal end-to-end rectorectal anastomosis using the original procedure described by Upadhyaya in infants and Ibrahim [25] in neonates. The procedure was easy, simple and feasible.

In colostomy group (first group), the definitive surgery was delayed for more than 6 to 8 months, we performed a classical Pena procedure in four cases, and transanal end-to-end rectorectal anastomosis in one case. This group had a very long hospital stay comparing to neonatal procedures and this made the child more vulnerable to hospital cross infection in addition it was a hard thing to families to come to hospital to perform three operations for their children.

Our study had low postoperative complications rate in neonatal one-stage procedure; one of these complications was anal stenosis, due to parent’s incompliance and negligent of postoperative follow-up program.

The advantages of neonatal procedures are decompressing the abdominal obstruction with establishing anorectal continuity, with short hospital stay, without colostomy and its susceptible complications. If it occurred it may need more hospital stay and additional surgery for fixation. There were no significance differences between two neonatal procedures, but we thought it would be simple and perhaps safe to attempt primary one-stage procedure in neonate. We found initial colostomy to relieve the obstruction should be used in neglected cases with severe abdominal distention and bad general condition.

One to six months postoperatively all patients were in good condition and had one to three intestinal emptying daily. There had been no diarrhea or sign of intestinal obstruction. Post operative follow-up dilation program is very important.

\textbf{Conclusion:}

Many approaches could be carried out for surgical management of rectal atresia. Our study concluded; that neonatal surgical correction of rectal atresia is easy, simple, feasible procedures, and with less-cost effective. The anatomy can be identified from surrounding structures and anastomosed easily.

Mini PSARP is less traumatizing procedure to anal sphincter, moreover the anastomosis was done under vision. Three-stage approach can be carried out to neglected cases with bad general condition.

\textbf{References}