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Single Stage Reconstructive Surgery to Treat Anorectal Malformations in Neonates; Ten Years Experience

Mumtaz Hussain Khan[1], Naila Yaqub[2]

[1]Section of Pediatric Surgery, Northern Area Armed Forces Hospital, Hafr Al Batin, Saudi Arabia.
[2]Department of Pediatrics, Northern Area Armed Forces Hospital, Hafr Al Batin, Saudi Arabia.

Author`s Contribution

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Address of Correspondent
Dr Mumtaz Hussain Khan
Northern Area Armed Forces Hospital, Hafr Al Batin, Saudi Arabia
mumtazhkhan@yahoo.com

ABSTRACT

Objective: To present single surgeon's ten years' experience of Single stage reconstructive surgery in 26 neonates born with anorectal malformations (ARMs) and review of literature.

Methodology: This is a retrospective study of 26 neonates admitted in NICU with diagnosis of ARMs between period of June 2011 to Oct 2021 and managed by single stage reconstructive surgery with an average follow up of three years. All these patients were full term. The diagnosis was confirmed on clinical examination supported by cross table lateral film in prone position after 24 hours and ultrasound evaluation of level of rectal pouch. Single stage reconstructive surgery was done under general anesthesia within 24 to 48 hours after birth. The outcome of single stage reconstruction of neonates with anorectal malformations is evaluated post operatively during an average regular follow up of 3 years.

Results: All the 26 neonates are treated by single stage reconstructive surgery between 24 to 48 hours after birth during the period from June 2011 to Oct 2021 who were admitted with anorectal malformations in NICU after excluding associated congenital anomalies. All the patients had excellent cosmetic and functional outcome without significant problem of wound infection. Only 3 female patients with recto-vestibular fistula had mild superficial wound infection and were managed conservatively.

Conclusion: Single stage reconstructive surgery to treat ARMs in neonates I effective, safe and feasible with good continence. It avoids morbidity and higher cost associated with three stage surgeries and colostomy. This fact may further be confirmed through multi-institutional experience in large number of patients.

Key Words: Anorectal malformations, Posterior Sagittal Anorectoplasty, Recto-urinary fistula, Rectoperineal Fistula, Recto-vestibular fistula, Recto-vaginal fistula.

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Introduction

ARMs are congenital anomalies varying from minor to complex defects.[1] Consequently, the conventional classification of low, intermediate and high anomalies leads to ambiguous results. ARMs present as low version in 90% of the females and 50 % of times in the males.[2] ARMs are usually associated with other congenital anomalies. [3]

The incidence of ARMs is estimated as 1 in 4000 to 5000 live births and affects boys and girls equally.[4] ARMs are diagnosed on clinical examination supported by cross table lateral film in prone position and ultrasound examination.[3,5] Ultra sound evaluation is helpful to classify ARMs and to plan treatment regarding need of initial colostomy.[5]

The factors which contribute in diagnosis include, pouch-perineal distance, location of fistula, optimal timing of the ultrasound examination and the approach used for the examination. The surgical classification of type of ARM is based on the relationship of level of distal rectal pouch and puborectalis muscle.[3,5]

The ARMs in neonates with Down syndrome are generally not associated with fistula. The incidence of ARM with rectovaginal fistula is less than 1%. Pena concluded that the higher incidence of isolated recto-vaginal fistula is due to misdiagnosed recto-vestibular fistula and persistent cloaca.[6]

ARMs usually require immediate surgery to treat obstruction unless a fistula can be relied on. The anomaly it is treated depending upon its type, either with perineal anoplasty alone or with initial colostomy followed by a definite repair by PSARP later.[7,8]

Pena has recommended three stage approach for the surgical correction of high ARM in neonates with high sigmoid colostomy, PSARP and closure of colostomy.[9]

Initial diverting colostomy is safe option for surgeons who do not have enough experience in managing ARMs to avoid wound dehiscence due to infection at the time of PSARP. However, a colostomy itself is a source of morbidity with many complications including bleeding, wound infection, sepsis, prolapse, stenosis, stricture, fluid and electrolyte imbalance and skin excoriations. [4,10]

Selection of single stage PSARP or three stage procedures with initial colostomy has been a subject of debate, especially in ARMs with recto-urinary fistula in male patients and ARMs with recto-vestibular and rectovaginal fistulas in female patients. Many series have reported excellent cosmetic and functional outcome without significant problem of wound infection in patients treated by single-stage PSARP.[12-17]

Primary single stage neonatal repair of ARMs with PSARP without colostomy not only has advantages of being safe and effective but also has psychological and financial benefits to the family. Single stage management, however, should be chosen by a pediatric surgeon with appropriate experience.[18,19] Parenteral nutrition to minimize wound contamination with stool and appropriate care are helpful to avoid wound infection.

The literature shows, that single stage repair of ARMs with intermediate and high types in both genders avoids the morbidity associated with colostomy and provides an opportunity for early training of the perineal musculature leading to improved long term fecal continence. One stage PSARP is a definitive repair that can be carried out in neonates without prior colostomy. The meconium is sterile during the first week after birth, thus risk of infection is minimal. Many centers have recorded success with primary PSARP in neonates.[20,21]

Low type of ARMs in both genders is usually treated by single stage perineal anoplasty.[22]

ARM with rectovesical fistula is treated by PSARP with simultaneous abdominal access for division of recto-vesical fistula and mobilization of rectum for tension free recto-neo-anal anastomosis. Laparoscopic assisted PSARP is widely being used for treatment of ARM with recto-vesical neck fistula.[23]

Single stage PSARP in neonates is now preferred over three stages with initial colostomy which is associated with significant morbidity.[10,24]

The aim of this retrospective study is to present single surgeon's ten years experience of management of spectrum of ARMs by primary reconstructive procedure (anoplasty in low type ARMs and classical PSARP in intermediate/high ARMs) in 26 neonates and review of the literature for comparison.

Methodology

This is a retrospective study of 26 neonates admitted in NICU with a diagnosis of ARMs between June 2011 to Oct 2021, managed by single stage reconstructive surgery with an average follow up of three years.

All these patients were full term. Sixteen patients were female and 10 were male. Out of 16 female patients, 5 had recto vestibular fistula, one had recto-vaginal fistula and 10 had rectoperineal fistula. Out of 10 males, 5 had recto urinary fistula (2 recto bulbar urethral fistula, 3 recto prostatic urethral fistula, 1 recto vesicle fistula), 3 had rectoperineal fistula and one had ARM without fistula associated with Down syndrome.

Patients with other associated congenital anomalies like esophageal atresia were excluded from the study. All the patients underwent echocardiography to evaluate for associated cardiac anomalies.

The diagnosis of ARM was made by careful perineal examination. A general physical examination was done to rule out other congenital anomalies. The diagnosis was further confirmed by a cross table lateral film after 24 hours and an ultrasound evaluation of the level of the rectal pouch and location of fistula. The other associated congenital anomalies were ruled out by echocardiogram and abdominal ultrasonography in all patients.

All the neonates with perineal fistula (both male and female) were operated between 24 to 48 hours after birth by primary perineal anoplasty. All the neonates with recto-vestibular, rectovaginal and rectourethral fistula were operated by single stage PSARP. One male patient with rectovesical fistula(with passage of meconium in urine and having rectal gas shadow above 5th ossified sacral vertebra on lateral cross table film in prone position and ultrasound suggestive of high distal rectal pouch with vesical neck fistula) was operated with initial identification of anal sphincter and muscle complex and dissection for neo anus and fixation of tube for rectal pull through followed by division of rectovesical fistula and rectal mobilization through concomitant abdominal access for a tension free, well vascularized recto neo anal anastomosis (Figure 1-8). One male patient with ARM without fistula in association with Down syndrome was treated by single stage PSARP.

All patients were placed in prone (Jack- knife) position, and the pelvis was raised. The exact location of the anus and the sphincter was determined by electrical stimulator. The skin and subcutaneous tissue was incised in midline. The anus and the rectum were placed in their correct positions and fixed there with sutures. Intravenous antibiotics were administered for 7 to 10 days postoperatively. All patients received TPN until adequate oral feeding was established.

The outcome of single stage reconstruction of neonates with anorectal malformations is evaluated to be effective and safe post operatively during an average 3 years of regular follow up.

Care was taken to prevent contamination with meconium during surgery. The patients were kept NPO for 5 post operative days to minimize the wound contamination with stool. Wound care was done with regular cleansing with normal saline followed by application of antibiotic ointment. The bladder catheter was removed after 5 days. Most of the patients were discharged home after 10th post operative day. Anal dilatations were started after 2 weeks of surgery. All patients were followed up for a mean period of 3 years. Using Pena's criteria for assessment of continence, the clinical examination for faecal continence was conducted through a parent interview.

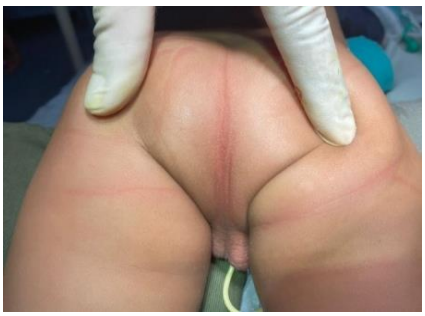

Figure 1. ARM with rectovesical fistula

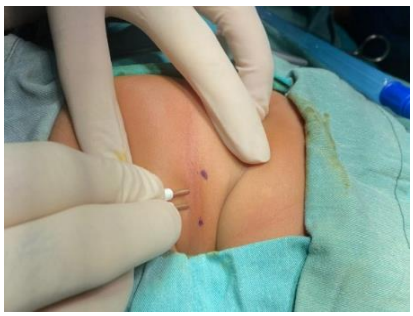

Figure 2. Identification of all sphincter components

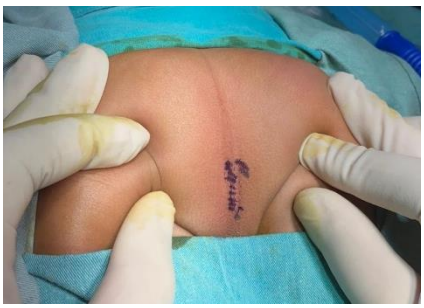

Figure 3. Midline excision marking

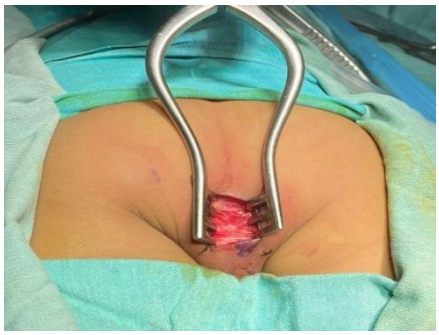

Figure 4. Incision to divide all structures in midline

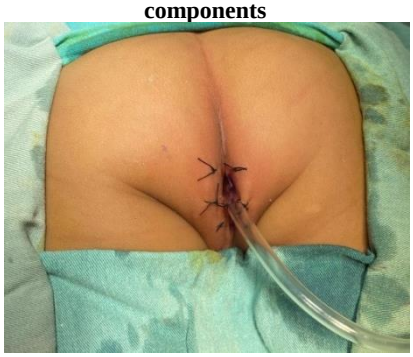

Figure 5 Tube fixed within sphincter complex to pull rectum for rectoneonanal anastamosis

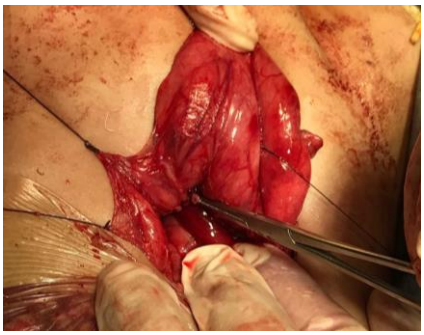

Figure 6 Division of rectovesicle fistula through simultaneous abdominal access

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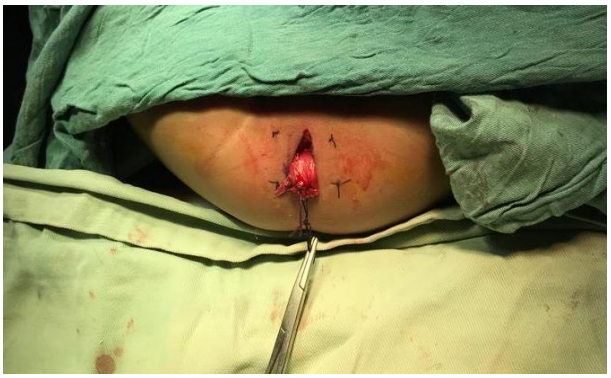

Figure 7 mobilized rectum pulled through Previously dissected passage within all Sphincter components guided by Prefixed tube.

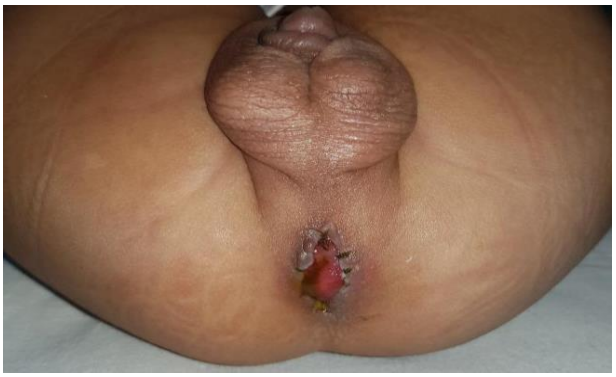

Figure 8 Rectal neoanal anastomosis on follow up after 3 weeks.

Results

All patients were operated by PSARP between 24 to 48 hours after birth. The fistula with urinary system (recto-bulbar urethra, recto-prostatic urethra and recto-bladder neck) was found in 6 male patients. The fistula with genital tract (5 recto vestibular and 1 recto vaginal) was found in 6 female patients. The dissection was easy in all patients. Only one patient with recto vesical fistula needed concomitant abdominal approach to divide recto vesical fistula and mobilization of rectum for recto neo anal anastomosis. Rectal tapering was not needed in any of our patients. Thirteen patients with rectoperineal fistula (3 male and 10 female) were treated by perineal anoplasty (Table I). There was no intraoperative complication. All patients were started on oral feed after 5 days. All patients needed gentle regular strict wound cleansing. No patient had urinary symptoms after removal of bladder catheter after 5 days. Three female patients with recto-vestibular fistula had superficial wound infection of anoplasty site and were treated conservatively. No patient had disruption of anoplasty or rectal retraction. Most of the patients were discharged home after 10 days. All patients underwent anal dilatations after 2 weeks of surgery and there was no complication of anorectal stenosis. No patient had chronic constipation in this series. No patient had urinary or fecal incontinence on regular follow up for a mean period of three years (Table II). In our series we found that primary PSARP is effective and safe approach in management of neonates with ARMs.

Table I: Neonates with spectrum of ARMs

ARMs	Gender (No. of patients)	
	Male	Female
ARM with perineal fistula	3	10
ARM with recto-bulbar-urethral fistula	3	-
ARM with recto-prostatic urethral fistula	2	-
ARM with recto-vesicle fistula	1	-
ARM with recto-vestibular fistula	-	5
ARM with recto-vaginal fistula	-	1
ARM without fistula	1	-

Discussion

Primary corrective single stage surgery is a valid option in management of ARMs in neonates.[12] This approach has been practiced at many centers with variable success. Such correction has successfully been done in female patients with recto-vestibular fistula with good outcomes. Pena and Devries advocated classical 3 stage approach.[4] However, there is significant morbidity associated with a colostomy and there is higher cost of three surgeries. Also, the chance of benefit from early restoration of intestinal continuity is lost.[25]

Table II: Long-term outcome with regards to continence and constipation

Single-stage Procedure	Gender (No. of patients)		Continence	Constipation
	Male	Female	good	-
Single-stage anoplasty for ARM with perineal fistula	3	10	good	-
Single-stage PSARP for ARM with recto-vestibular fistula	-	5	good	-
Single-stage PSARP for ARM with urethral fistula and recto-vaginal fistula.	5	1	good	-
Single stage PSARP for recto-vesical fistula with concomitant abdominal access (for division of recto-vesicle fistula and mobilization of rectum for tension-free recto-neo-anal anastomosis)	1	-	Good	-
Single stage PSARP for ARM without fistula	1	-	Good	-

Recently, the literature shows that there is emphasis aiming at single stage repair of ARMs in both genders to avoid morbidity associated with colostomy.[13-17]

Moor first reported anterior approach for sagittal anorectoplasty performed without colostomy in neonates with recto-urinary fistula with excellent results.[8] Albanese et al reported successful outcome in 5 male neonates treated by primary PSARP.[18] Liu and Hill also have reported good results in 7 male patients with recto-urinary tract fistula treated by primary PSARP.

Mishra et al in their comparative study of primary PSARP and staged procedures reported good results in their series of 14 neonates who were treated by primary PSARP at birth. Mirshemirani reported 17 male neonates with rectourethral fistula who were treated effectively and safely by primary PSARP.[17] One of the concerns of primary correction is damage of local surrounding structures. Expert pediatric surgeons agree to the fact that most mishaps occur in patients with recto bladder neck fistulas. Such accidents include urethral damage, division of vas, pull through of dilated ectopic ureters and a neurogenic bladder. Thus, skin excoriations Pena recommends to repair these malformations in neonates by primary procedure only with low type of anomalies.[4,22] Albenese et al have used cystoscopy in their 5 neonates but they could identify urinary fistula in only 3 cases.[26]

Most surgeons proceed with primary PSARP to deal with urinary fistulas only intraoperatively.[14-17] We used the same approach in our series. We used lateral cross table film in prone position and ultrasound to evaluate the level of the rectal pouch and site of fistula. We did not have to do tapering of rectal pouch in any of our patients.

Wound infection is an important concern during primary PSARP. However, wound infection does not appear to be a concern in reported experience of primary PSARP.[27] We also, did not find wound infection as a problem in our series. Neonatal bowel takes approximately one week to be colonized with gram negative and anaerobic bacteria and thus meconium is sterile during this time.

Continence after correction of ARM depends upon multiple factors including development of perineal musculature, spine, and placement of rectum within sphincter during surgery, uneventful postoperative recovery and proper conditioning of defecation reflex.[28-30]

A meticulously done primary PSARP gives chance to all existing factors of their best utilization and most surgeons have reported good results. [31-35] In our series, all patients have normal/voluntary bowel movements after an average of 3 years of follow up. Primary single stage neonatal repair of ARMs with PSARP without initial colostomy not only has definite advantages of correction being safe and effective but also has psychological and financial benefits to the family. However, the procedure should not be taken lightly and it should be performed only by an experienced pediatric surgeon.

Conclusion

Single-stage reconstructive surgery is effective, safe and feasible with good continence. This fact may further be confirmed through multi- institutional experience in large number of patients.

We recommend single stage reconstructive surgery to treat ARMs in neonates for better clinical outcome and to avoid morbidity and higher cost associated with three stage surgeries and colostomy**.**

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