

“A study of children with intermediate anorectal malformation who have undergone sacroperineal pull through with focus on bowel function, quality of life and parental stress.”

Abstract

Anorectal malformations comprise a wide spectrum of diseases that affect boys and girls and can involve malformations of the distal anus and rectum, as well as the urinary and genital tracts. Malformations range from minor, easily treated defects that carry an excellent functional prognosis, to complex defects that are difficult to treat, are often associated with other anomalies, and carry a poor functional prognosis.

166 children who had sacroperineal pull-through done for intermediate anorectal malformation from 1996 to 2005 were called for the study. The follow up period ranged 14 to 144 months with a mean follow up period of 6.5 years. The aim of the study was to study the relationship of the pulled through bowel with the anorectal sphincter complex in children who have undergone sacroperineal pull through operation for anorectal malformations (ARM), in order to determine whether the pulled through bowel is correctly sited within the sphincter complex and correlate this with the bowel function. The functional outcome after surgery especially in regard to Quality of Life (QOL), the psychosocial effects on the children due to the anomaly and the treatment were evaluated. The parental stress in bringing up a child with anorectal anomaly was also evaluated.

Ages ranged from 2.5 years to 13 years with a mean age of 7.5 years. The parents were young especially the mother with an average age of 23 years. 57% of the family belonged to a low socioeconomic group. 42.8% of the children were first born. 78% and 89% of the children were

below 50th percentile for their height and weight respectively. 28% had anal stenosis and 21% had major anal mucosal prolapse. 31% underwent secondary anal procedures. Voluntary bowel movement was seen in 83%, constipation, as detected by the abdominal x-ray, was seen in 100% and soiling in 88%. Anal manometry was done in 18 children. The average anal resting pressure and voluntary squeeze pressures were 52.5 and 144.2 cm of H₂O. Magnetic resonance imaging was done in 13 children. The pulled through bowel was central in 77% and the anorectal angle was clear in 92% of the children. 74% of the parents were emotionally affected by the birth of a baby with anorectal malformation. 52% families had difficulty in socializing and 48% children had difficulty in making friends. There was a statistically significant co-relation between soiling and satisfaction with the final result. The quality of life (QOL) scores were on average 7.8 in children with poor continence and children with good continence had a score of 10.5. There was statistically significant improvement in the (QOL) after bowel management program.

Conclusion

The main post operative complication was constipation leading to fecal impaction and soiling. Management of constipation by bowel management program improves the QOL. Sacroperineal pull-through ensures proper placement of the bowel within the sphincter complex.

A STUDY OF CHILDREN WITH
INTERMEDIATE ANORECTAL
MALFORMATION WHO HAVE UNDERGONE
SACROPERINEAL PULL-THROUGH WITH
REGARD TO BOWEL FUNCTION, QUALITY
OF LIFE AND PARENTAL STRESS

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STRESS

A DISSERTATION SUBMITTED IN PARTIAL FULFILLMENT OF
M.Ch BRANCH-V (PAEDIATRIC SURGERY) EXAMINATION OF THE
DR.M.G.R MEDICAL UNIVERSITY, CHENNAI, TAMIL NADU
TO BE HELD IN AUGUST 2008.

CERTIFICATE

This is to certify that Dr. Viju John is doing M.Ch Paediatric Surgery training course in the Department of Paediatric Surgery, Christian medical College and Hospital, Vellore from 1st August to July 31st 2008.

He has done the dissertation “A study of children with intermediate anorectal malformation who have undergone sacroperineal pull-through with regard to bowel function, quality of life and parental stress” under the guidance of Dr. Jacob Chacko, to be submitted in partial fulfillment of Branch – V (Paediatric Surgery) examination of Dr. M.G.R Medical University, Chennai, TamilNadu, to be held in August 2008.

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ACKNOWLEDGEMENTS

- Heart-felt gratitude and thanks
- To the God Almighty for his blessings
- To Dr. Jacob Chacko who was in true sense a guide. Guided me in this work with lots of patience and perseverance.
- To Dr. Sudipta Sen for all the useful insights and valid suggestions.
- To all the children in the study who have become so dear to me and their parents especially mothers who shared with me their personal travails and joys.
- To Dr. Gordon Thomas, Dr.Sampath Karl and Dr.Jyotish Kumar for their support
- To Dr. Sridhar and Dr. Anu Eapen for their valuable help.
- To Ms. Nithiya for the help in statistical analysis and Reshma for the help with anal manometry.
- To Hema, Vimala, Jasmine and George for their help
- To Molly for her support and help. Abigail for pitching in at times and the little ones for allowing the computer to take their place.

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INTRODUCTION

Anorectal malformations comprise a wide spectrum of diseases that affect boys and girls and can involve malformations of the distal anus and rectum, as well as the urinary and genital tracts.

Malformations range from minor, easily treated defects that carry an excellent functional prognosis, to complex defects that are difficult to treat, are often associated with other anomalies, and carry a poor functional prognosis.

During the last 15 years, significant advances have occurred in the management of anorectal malformations. We now recognize the importance of the concept of quality of life. The current goals in the management in these defects are (1) to anatomically reconstruct all malformations; (2) to recognize and treat any associated defects that may be life threatening; and (3) to treat the functional sequelae of the malformations, in order to provide these patients with a good quality of life. Over the last half century or more, the treatment of ARM evolved from a simple cut back procedure/translocation anoplasty to the abdominoperineal and later the sacroperineal pull through procedures. The sacroperineal pull-through was widely practiced till the introduction of the posterior sagittal anorectoplasty (PSARP) operation. The majority of children have significant problems in anorectal function beyond their childhood.

The major postoperative problem is motility disturbances leading to chronic constipation with over flow incontinence, and true sphincter insufficiency with stool incontinence.

OBJECTIVES AND AIMS OF THE STUDY

1. To study the relationship of the pulled through bowel with the anorectal sphincter complex in children who have undergone sacroperineal pull through operation for anorectal malformations (ARM), in order to determine whether the pulled through bowel is correctly sited within the sphincter complex.
2. To correlate this with the anal squeeze pressure.
3. To correlate this with the functional outcome after surgery especially in regard to Quality of Life (QOL)
4. To assess the psychosocial effects on the children due to the anomaly and the treatment
5. To evaluate parental stress in bringing up a child with anorectal anomaly

REVIEW OF LITERATURE

History of the Procedure

Paul of Aegineta in the seventh century recorded the earliest account of successful surgery for imperforate anus (1). He suggested rupturing an obstructing membrane with the finger or knife point and then dilating the tract until healing was complete. In 1576, Galen described the anal sphincters, levator muscles, and coccyx (2). In 1976 Cooke treated a child by making a small incision over a blind anal membrane and dilated the aperture. In 1787, Bell suggested using a midline perineal incision to find the bowel (3). In 1783, acting on Littre's suggestion, Dubois performed an inguinal colostomy for imperforate anus (4). Other surgeons followed suit, but almost all of the infants died; thus, colostomy remained unpopular and a procedure only of last resort. In 1834, Roux of Bringnoles attempted to preserve external sphincter function and used a midline longitudinal incision extended toward the coccyx (5). In 1835, Amussat described formal perineal proctoplasty (i.e., mobilization of the bowel through a perineal incision and suturing to the skin) (6). This technique gained rapid acceptance. Strictures were less common than observed in earlier procedures. In 1860, Bodenhamer championed the midsagittal incision first described by Roux 27 years earlier. McLeod in 1880 described an abdominoperineal (AP) procedure for cases where the rectum is not found low (7). In 1886, McCormac suggested two stage procedure- preliminary colostomy and subsequent proctoplasty. In 1930, Wangensteen and Rice first advocated imaging to delineate the abnormality (8). Dr. Ladd and Gross at Children's hospital Boston kept their dissection

close to the rectum and divided the external sphincter into two halves and resutured in front and behind the proctoplasty (9). Up to this time, the emphasis was on perineal procedures and colostomy was resorted to as a last resort and was associated with a high mortality. Following World War two, with development of better antibiotics and better anesthesia, interest in combined abdominal procedures was rekindled.

Denis Browne popularized initial colostomy and then followed by an abdominoperineal pullthrough through a hole stretched (not cut) in the pelvic floor. He also popularized the cutback anoplasty for perineal fistulas. In 1953, Stephens while working with Denis Browne described the Sacroperineal rectoplasty and emphasized the role of levator ani and downplayed the importance of internal and external anal sphincters (10). In 1959 Fritz Rehbin reintroduced the endorectal pull- through combined with AP approach. Rehbein divided the bowel at laparotomy and stripped the mucosa from the distal atretic segment and pulled the proximal bowel through the resultant muscular sleeve to the anal dimple to perform an anoplasty (11). He missed the puborectalis in performing the procedure. In 1961 Stephens proposed the importance of puborectalis as the main muscle of continence. In 1963, because of the high incidence of incontinence with the abdomino-perineal approach, Kiesewetter modified Stephens' technique by performing the abdominosacroperineal procedure (12). Unlike Stephens he believed the external sphincter was present and worth saving. Gross and later Nixon, in autopsy studies, found the external sphincter to be present. Swenson in 1967 described his AP procedure and the importance of puborectalis sling. He also completely ignored the external sphincter (13). In 1978, dissatisfied with the results of other procedures, Mollard proposed an anterior perineal approach bringing the atretic bowel in front of the puborectalis muscle (14).

There was however, a high incidence of soiling and mucosal prolapse. The results of the various procedures were difficult to assess as different subjective criteria for grading and definitions were used by various authors to assess the function. Incontinence remained a major problem.

In 1980, the introduction of the posterior sagittal anorectoplasty (PSARP) by Alberto Pena and deVries was a major event in the history of ARM (15). This approach allowed pediatric surgeons to clearly view the anatomy of anorectal malformations and to repair them under direct vision. They redefined the arrangement of the pelvic muscles and the external sphincters, as a fused sphincter muscle complex. He divided the muscles posteriorly in the midline from the anal dimple to the coccyx, and placed the tapered rectum within it. The divided muscles were then sutured posteriorly in the midline. It was apparent that many of the children had constipation and fecal soiling and hence they instituted a close follow up program to assure patients and parent compliance with postoperative anal dilatations and appropriate rectal washouts. Thus the bowel management program was established to carefully follow up all operated patients (16). Pena also brought the concept of urogenital advancement in the repair of high cloacal defects (17). In 1992, Malone described the ante-grade colonic continent enema (MACE) procedure as a means of flushing the colon instead of the retrograde enema (18). By 2000 laparoscopic assisted one stage abdomino-perineal pull-through procedures, was also introduced in the management of ARM. This was refined a step further by introducing a laparoscopic muscle stimulator to accurately identify the sphincter.

There remain several areas of controversy regarding the choice and timing of the procedure and methodology used to assess the results. Designing a protocol that will

define the precise location of the pulled through bowel and pelvic and sphincter muscle assessment using MRI and /or anal endosonography, anal manometry, colonic motility studies in addition to the evaluation of soiling, sensation and other subjective analyses is necessary.

Mechanism of Defecation

Various reflexes, such as the gastrocolic reflex and the iliocolic reflex contraction of the colon, caused by the filling of stomach and colon, respectively, as well as the voluntary contraction of the abdominal musculature, may initiate defecation by suddenly filling the rectum with colonic contents. The increasing intrarectal pressure stimulates the distension receptors in the puborectalis muscle and the parapuborectal tissues, and the desire to pass a stool is constantly felt. At the same time a reflex relaxation of the internal anal sphincter occurs. This allows even the smallest amount of stool to reach the anal canal. The hypersensitive mucosa of the anal canal in the region of the anal valves is able to discriminate between flatus and liquid or solid stool. The reflex contraction of the external anal sphincter and the puborectalis prevent the expulsion of stool from the anal canal and thus inhibit fecal soiling. This effect is increased by the compression of the lower anal canal by the cavernosum of the rectum, and by the corrugated muscles of the anus. This allows the rectum time to adapt itself to the increased intraluminal pressure. The aboral-oral pressure gradient of the rectum will propel the stools upward into a more proximal rectal segment. This however will stimulate further propulsive waves via a feedback mechanism. An intrarectal pressure of between 25-35 mm Hg will stimulate a reflex inhibition of the anorectal sphincters and the puborectalis muscle. The voluntary contractions of the abdominal muscle will also cause a reciprocal inhibition of the striated

muscles of the pelvic floor. This, in turn, will decrease the acuteness of the anorectal angle formed by the puborectalis muscle, and the defecation commences. When the rectoanal reflex operates following sudden distention of the rectum, sampling of whether the waste is solid, liquid or gas occurs at approximately the level of anal valves. If the defecation is not intended, voluntary contraction of the puborectalis will return the contents back into the rectum off the sensitive zone and the desire to defecate will diminish.

The external sphincter is a powerful muscle, brought into action in moments of stress to supplement the sling action in arresting defecation or deflation. It too has a resting tone that mildly occludes the anus, and is forced open by flatus under high pressure. The tone of the internal and external sphincters that surround the skin-lined anal canal is probably responsible for preventing wetting of this part of the anal canal with the mucus secreted by the adjoining rectal mucosa, in the long intervals between acts of defecation

Children who become chronically constipated indicate that the sleeve-and- sling become easily tired by the impacting feces, become relaxed, and permit shortening of the anal canal to the length only of the skin-lined anus. It is found that then the short passage, although encircled by the external and the internal anal sphincters, is barely sphincteric, permitting constant leakage, which is momentarily arrested only at the time of conscious muscular contraction of the external sphincter surrounding the skin lined anus. The anal canal constructed in patients exhibiting a congenital rectourethral fistula is endowed with a high degree of sensation, content discrimination, and muscular sphincter function if the new canal is lodged within the striated muscle complex, which is then its only sphincter. Some believe that skin-lined anal canal is vital to continence (19) However

Stephens and Smith consider that proper function of the puborectalis is adequate for near complete continence (20).

Classification

ARM represents a wide spectrum of defects and conditions. A clear understanding of normal anorectal anatomy and the different types of ARM is necessary for both the planning for surgery and the procedure itself. An appreciation of the classification systems is useful in practice of the surgeon.

Ammussat in 1835 was first to attempt a classification of ARM (21). He described 5 groups. In 1934 Ladd and Gross proposed a classification system which became the standard (Table 1).

Table 1. Classification according to Ladd and Gross (9)

Type	Anomaly
1	Anal and anorectal stenosis
2	Imperforate anus
3	Imperforate anus with blind Ending pouch with fistula
4	Rectal Atresia

In 1963 a Melbourne team led by Stephens classified the lesions into two categories, either high or low. This classification recognizes the importance of puborectalis muscle and its effect in continence. Lesions above the pubococcygeal (PC) line were described as

'high' and below as 'low'. "Intermediate" was later added to this classification. The PC line represents the level of the levator ani attachment of the pelvic wall. In 1964 Santulli proposed his classification based on the work of Ladd and Gross. This also divided the lesions into low, infralevator, and high, supralevator (66). In 1970 the international classification system was proposed at a symposium on Anorectal Malformation at the pediatric surgical congress in Melbourne. Based on the earlier work of Stephens and Smith, this classification was based on the principle of normal and abnormal anatomy and divided the lesion in three groups high (supralevator), intermediate and low (infralevator). Though it was too complex and had nearly 40 subtypes, it is still used in literature.

New research and variations in surgical techniques in the late 1970s and 1980s altered previously held concepts. This led to "Wingspread classification" (Table 2) which evolved from a conference held in Wingspread Convention center, Racine, Wisconsin (USA) in 1984 (22). It was created to update the Melbourne classification.

However the Wingspread classification was not fully endorsed by the surgical community as it is based on anatomical principles. Alberto Pena proposed a classification based on the anatomical defect and how they correlate with surgical management (Table 3) (23).

Table: 2 Wingspread Conference classifications

Level of anomaly	Male	Female
High	1. Anorectal agenesis A. Rectovesical fistula B. Without fistula 2. Rectal atresia	1. Anorectal agenesis A. Rectovaginal fistula B. Without fistula 2. Rectal atresia
Intermediate	1. Rectourethral fistula 2. Anal Agenesis without fistula	1. Rectovestibular fistula 2. Rectovaginal fistula 3. Anal agenesis without fistula
Low	1. Anocutaneous fistula 2. Anal stenosis	1. Anovestibular fistula 2. Anocutaneous fistula 3. Anal stenosis
Miscellaneous	Rare malformation	Persistent cloacal anomaly Rare malformation

Table: 3 Pena's classification

Male	Females
Perineal (cutaneous) fistula	Perineal (cutaneous) fistula
Rectourethral fistula 1. Bulbar 2. Prostatic	Vestibular fistula
Rectovesical fistula	Persistent cloaca
Imperforate anus without fistula	Imperforate anus without fistula
Rectal atresia	Rectal atresia

In 2005 an international congress for the development of standards for the classification, treatment and follow up of ARM took place in Krickenbeck Castle in Westphalia, Germany. At this meeting a new, unifying, international classification system was introduced which would enable standardization of definition and treatment protocols for various anomalies. This was known as the Krickenbeck Classification (24). It is not based on the anatomical, embryological features or the imaging (Table: 4). This classification is based on the frequency occurrence and allows management outcomes to be measured.

Standards for diagnostic procedures: International classification (Krackenbeck) (Table: 4)

Major clinical groups	<ol style="list-style-type: none"> 1. Perineal (cutaneous) fistula 2. Recturethral fistula <ol style="list-style-type: none"> A. Bulbar B. Prostatic 3. Rectovesical fistula 4. Vestibular fistula 5. Cloaca 6. No fistula 7. Anal stenosis
Rare/ regional variants	<ol style="list-style-type: none"> 1. Pouch colon 2. Rectal atresia/ stenosis 3. Rectovaginal fistula 4. H type fistula 5. Others

Besides the classification an international grouping of surgical procedures (Table: 5) for follow was developed at the Krackenbeck meeting. This standardization hopes to make the different surgical procedures comparable with each other.

Table: 5 International grouping of surgical procedures for follow up

Operative procedures

1. Perineal operations
2. Anterior sagittal approach
3. Sacroperineal procedures
4. PSARP
5. Abdomino-sacroperineal pull through
6. Abdominoperineal pull through
7. Laparoscopic assisted pull through

Associated conditions

1. Sacral anomalies
2. Tethered cord

Anorectal malformations are a spectrum of congenital defects that continue to represent a significant challenge for the pediatric surgeon. These defects are frequently associated with life-long debilitating sequelae such as fecal and urinary incontinence and sexual inadequacy. It is the obligation of all surgeons who care for these children to avoid complications that will increase the risk of these sequelae and to rehabilitate the children after surgery to put them back into the mainstream.

Scoring Postoperative results

Standardized assessment of clinical outcome is essential after repair of Anorectal malformation for appropriate quality control and comparing different treatment modalities in patients treated in single or different centers. Clinical assessment is subjective and may be biased by the observer who often is the operating surgeon. Therefore scales and scores that provide information on the condition and functional status is needed. However appropriate methods and instruments of collecting data on the outcome after the repair of ARM has been a matter of debate. In 1960 Scott introduced a simple score which included exclusively clinical data (25). Since then approximately 10 scoring systems have been introduced and used with various frequency. None of the instruments used in the scoring system has undergone proper validation. The reported differences in results of different series of patients with ARM undergoing treatment remain difficult to interpret.

A scale is an instrument that is used to measure clinical phenomenon such as the degree of incontinence or the squeezing pressure of the internal anal sphincter. A score is a value on a scale in a given patient. Scores in a specific patient may be dichotomous (yes/no) or rank-ordered. Thus qualitative scores can be differentiated from numerical scores. Principally, a score may serve three functions: prediction, evaluation over time, or description at certain time point (26). A score has to be reproducible, valid and responsive. The process of ensuring reproducibility, validity, or responsiveness should not be based on observer's knowledge but on a structured process. Patients with ARM have been scored descriptively. None of the scores suggested for use has undergone

standardized validation process concerning reproducibility, validity or responsiveness, with the exception of quality-of-life scores. The endpoints like constipation, incontinence or soiling have also not been uniformly defined.

Specific Scores used in Patients with ARM

There is consensus that fecal incontinence represents the most important end point in patients with ARM. Various scores assessing the long term outcome after treatment focuses on differentiating various levels of fecal incontinence. No consensus is there for including and scoring other symptoms.

1. The Scott score (25).

In 1960 Scott established a qualitative score that differentiated between “good”, “fair” and “poor” continence. The items used are defecation habits, stool control, perianal soreness, and the function of the puborectalis muscle on digital examination. The score was not validated and a clear definition of terms such as constipation and sphincter tension was not given.

2. The Kelly Score (27). In the Kelly score the criteria are similar to Scott but the continence is scored quantitatively.

Table: 6 Kelly score (27).

Staining/ smearing	None	2
	Occasional	1
	Constant	0

Accidental defecation/ soiling	None	2
	Occasional	1
	Constant	0
Strength of puborectalis muscle	Strong	2
	weak	1
	none	0

The determination is based on the leakage phenomena, on the strength of the puborectalis sphincter, and on sensitivity. A total of 5-6 points is considered “good”, 3-4 points is “fair” and 2 points is “poor”. This is the most commonly used instrument for assessment of fecal continence today. It is compared with other more objective measures, such as manometry, electromyography, and quality-of-life data.

Holschneider and Metzger (28) introduced a quantitative clinical score, including the parameters of frequency of defecation, fecal consistency, soiling, rectal sensation, ability to hold back, discrimination and need of therapy. Later on the score was modified reducing the clinical parameters and including manometric data without changing the numerical scoring. Each seven parameters are scored as 0-2. For these scores , 14 points means normal bowel habits, 10-13 points means good (social continence), 5-9 means fair (marked limitation in social life) and 0-4 means poor bowel habits (total incontinence).

Table: 7 The Holschneider Score (1983) (28)

Frequency of defecation	Normal (1-2/day)	2
	Often (3-5/day)	1
	Very often	0
Fecal consistency	Normal	2
	Soft	1
	Liquid	0
Soiling	No	2
	Stress/ diarrhea	1
	Constant	0
Sensitivity	Normal	2
	Reduced (No discrimination)	1
	Missing	0
Anorectal resting pressure profile	> 20-24 mm of Hg	2
	14-19 mm of Hg	1
	<13 mm of Hg	0
Maximum squeeze pressure	>30 mm of Hg	2
	20-29 mm of Hg	1
	<20 mm of Hg	0
Adaptation reaction	Normal	2
	Small amplitude, shortened	1
	Not detectable	0

Holschneider et al. (29) recently stated that neither a reference to the course of anal or rectal fistula nor a rating as good, satisfactory or sufficient nor the current score system are suitable for comparative post op studies. The authors suggested renouncing the assessment of fecal incontinence taking chronic constipation into account. The subgroups described best differentiate the type of partial continence not the degrees of continence.

The Wingspread score (Table: 8)

In the Wingspread score the grades of continence are scored qualitatively (22). They fall into the four main categories of “clean”, “staining”, “intermittent fecal soiling” and “constant fecal soiling.” Subcategories include the need for occasional or constant therapy. In an additional category, related complications concerning the anorectum, urinary, genital or the other functions are noted.

Table:8 Wingspread score according to Stephens et al

1. Clean
1.1 <i>No accumulated feces</i>
1.11 No therapy
1.12 Occasional therapy
1.13 Therapy dependent
1.2 <i>Accumulated feces</i>
1.21 No therapy
1.22 Occasional therapy
1.23 Therapy dependent

2. Staining
2.1 <i>No accumulated feces</i>
1.21 No therapy
1.22 Occasional therapy
1.23 Therapy dependent
3. Intermittent fecal soiling
3.1 <i>No accumulated feces</i>
3.11 No therapy
3.12 Occasional therapy
3.13 Therapy dependent
3.2 <i>Accumulated feces</i>
3.21 No therapy
3.22 Occasional therapy
3.23 Therapy dependent
4. Constant fecal soiling
4.1 <i>No accumulated feces</i>
4.11 No therapy
4.12 Occasional therapy
4.13 Therapy dependent
4.2 <i>Accumulated feces</i>
4.21 No therapy
4.22 Occasional therapy
4.23 Therapy dependent
Related complication(Specify)
<i>1. Anorectal</i>
a. abnormal position
b. stenosis

c. prolapse
d. fistula
e. lack of contractility
f. abnormal length
2. <i>Urinary</i>
3. <i>Genital</i>
4. <i>Other</i>

Rintala Score

Rintala and Lindahl established a clinical score for the evaluation of fecal continence (30). The score is derived from standardized questionnaires, and physical examination is not required. The score consist of seven factors each given 0-3 scores, except frequency of defecation which is scored 1-2. The authors initially compared the scores of ARM children with normal controls. The score of 18 or above is taken as normal with “excellent” outcome after repair. The group with scores 9-16 was with “good” results, having occasional staining and infrequent accidents. Patients with “fair” results had intermittent daily soiling or staining and scored 7-11 points. Patients with “poor” results scored 6-9 points and had to use daily enemas because of severe constipation or had constant soiling. There were some validation steps. The scores derived from the questionnaires and the clinical outcomes noted in the hospital records were positively correlated. A pathological finding in plain spinal radiographs or MRI negatively correlated with bowel function score. Manometry did not differentiate with excellent and good clinical outcome, but showed a significantly reduced anal resting pressure in patients with fair or poor clinical outcome.

Pena score

Pena in 1995 (23) suggested a specific methodology of evaluation of long term results according to his personal experience. At the time of evaluation none of the patients had undergone any type of bowel management. Four parameters are evaluated:

1. Voluntary bowel movements, which are defined as feeling the urge to use the toilet to have a bowel movement, the capacity to verbalize it, and to hold the bowel movement.
2. Soiling is defined as involuntary leaking of small amounts of stools, which may present with or without bowel movements. Soiling grade 1 occurs occasionally (once or twice per week). Grade 2 refers to soiling that occurs every day, but does not cause social problems. Grade 3 represents constant soiling with social problems.
3. Constipation is defined as the incapacity to empty the rectum spontaneously without help everyday (Grade 1: when the patient is manageable by diet; grade 2: when he requires laxatives; grade 3 when he requires enemas).
4. Urinary incontinence is considered grade 1 when the patient has mild dribbling and wetness of the underwear day and night, and grade 2 when he is completely incontinent. Patients with voluntary bowel movements and no soiling are considered totally continent.

Other Scoring systems

Few “objective” methods of scoring are used. Electromanometry was used by Holschneider to define four grades of continence. He included selected manometric data in his clinical score for obtaining more objective results. Diseth and Emblem (31) confirmed that anal canal resting pressure and squeeze pressure correlated with fecal incontinence. Rintala et al (32) found that the only manometric parameter that correlated with the continence outcome was voluntary squeeze pressure. Fukata et al (33) compared endosonography and electromyography of the external anal sphincter with electromanometry and clinical data derived from Kelly score. Endosonographic findings for the external sphincter corresponded well with electromyographic finding, but not with manometry. Endosonography was comparable with MRI finding in only 9 out of 14 patients in one study by Jones (34). Fukuya et al (35) compared MRI with clinical assessment based on Kelly score. The proportion of “fair” or “poor” developed muscles was not significantly different between the continence groups according to Kelly.

Quality-of-life measurements

Quality of life is a multidimensional concept, which includes but is not limited to, the social, physical and psychological functioning of the individual. The relevance of quality of life assessment in children with ARM was confirmed in a study by Ditesheim and Templeton (36), who used questionnaire scoring system that included items such as school attendance, social relationships and physical capacities. Children and adolescents with fecal incontinence may suffer from emotional problems, internalizing behavior problems, and depressive symptoms. Various measures of quality of life have been used

for quantitative and qualitative scoring of children and adolescents with fecal incontinence and constipation.

Diseth and Emblem (31) used semi structured interviews and questionnaires such as Child assessment Schedule, Child behavior Check list, and self report in 33 adolescents with ARM. Psychosocial functions were impaired in 73% of the adolescents, and 58% met the criteria for psychiatric diagnosis. The authors found a significant correlation of the degree of flatus incontinence with the degree of psychosocial impairment, and of incontinence of flatus with mental health symptom scores. However in Ludman and Spitz (37) study in which the authors in addition assessed parents and teachers, the level of incontinence did not influence psychological adjustment, with the exception of incontinent young girls. There was no significant difference between continent and incontinent children concerning global self-worth measure. But there was a higher incidence of psychiatric disorder in children with incontinence. In another study it was found that there was a higher incidence of psychological or mental health problems with adolescents with ARM than with normal children. However the incidence of problem in continent or incontinent children was the same (38). Bai et al (39) used the Achenbach's Child Behavior Checklist in children with ARM and found quality of life to be significantly reduced as compared to normal control group. The authors established quality-of-life scoring criteria, including somatic assessment, social aspects, and psychological investigation.

Table: 9 Quality-of-life scoring for children with fecal incontinence.

Item	Criteria	Points
Soiling	Absent	4
	Accidental	3
	Frequent	2
Incontinence	Accidental	1
	Frequent	0
School absence	Never	2
	Accidental	1
	Frequent	0
Unhappy or anxious	Never	2
	Accidental	1
	Frequent	0
Food restriction	Never	2
	Accidental	1
	Frequent	0
Peer rejection	Never	2
	Accidental	1
	Frequent	0

Krickenbeck Score

A clear recommendation concerning specific instruments used for scoring cannot be derived from clinical experience or from data in the literature. Kelly score is the most widely used. The Holschneider score is the only one that includes an objective parameter (electromanometry). The score introduced by Rintala had a validation process and included comparison with normal children as controls. Quality-of-life measurement reveals most relevant and detailed information. At the Krickenbeck meeting in 2005 consensus was achieved regarding the use of a unified scoring system Table 10 (40). The assessment is made on children above 3 years of age who are not undergoing therapy. The analysis and assessment should be done by a person not involved in the treatment of the child

<u>Table: 10 Krickenbeck scoring syste</u>	
1. Voluntary bowel movements	Yes/no
Felling of urge	
Capacity to verbalize	
Hold the bowel movement	
2. Soiling	Yes/no
Grade1 Occasionally(once or twice per week)	
Grade Everyday, no social problem	
Grade 3 constant , social problem	
3. Constipation	Yes/no
Grade 1 Manageable by changes in diet	
Grade 2 Requires laxatives	
Grade 3 Resistant to diet and laxatives.	

Results following treatment of Anorectal malformation

Anal Complication: Postoperative anal complications including anal stenosis and mucosal prolapse have been found in 15-75% of patients (41) Anal stenosis is due to the inadequate anal dilatation during the post operative period. Anal dilatation may respond to anal dilatation but in refractory cases excision of the scar may be required. Mucosal prolapse requires operative treatment to reduce mucosal soiling and improve sensation. Few local anal problems have been reported after posterior sagittal anorectoplasty (23).

Prognostic factors determining the outcome in high and intermediate anomaly include

1. The level of the rectourogenital connection - Bowel functions of patients in intermediate anomalies (rectobulbar fistula in male, low-confluence cloacae and rectovestibular fistulae in females) is better than those with higher anomalies. The obvious cause of poor prognosis in high anomalies is marked hypoplasia of the sphincter muscles (23).
2. Sacral abnormalities – Severe sacral abnormalities like more than 2 missing sacral vertebrae or other major sacral deformities such as hemivertebrae and vertebral fusion adversely affect long term functional outcome (23). The poor outcome is usually related to the sphincter insufficiency. Sacral dysplasia also causes severe constipation by impairing rectal sensibility. Modern imaging has picked up lot of occult myelodysplasia in patients with ARM. But the impact of these lesions in long term bowel function is unclear.
3. Functional role of internal sphincter- the presence of internal sphincter in ARM is controversial. The functioning internal sphincter has been demonstrated by the

presence of the rectoanal relaxation reflex on anal manometry. In patients in whom the rectourogenital connection has been preserved at the anorectal repair, a functional internal sphincter has been demonstrated in 40-80% by demonstrating the rectoanal relaxation reflex. The presence of internal sphincter has been shown to correlate with favorable functional outcome (30).

4. Abnormal colonic motility - usually presenting as constipation is reported more with low ARM and females with vestibular fistula. However with PSARP being commonly employed constipation is one of the main functional complications of the repair (23). Constipation is more common when the internal sphincter saving repair is done. The reasons for constipation can be (a) extensive mobilization of the anorectum causing partial sensory denervation of the rectum and impairment of rectal sensation. (b) Rectosigmoid hypomotility (23) (c) dilated rectosigmoid which is present at birth or develop later in life but is rarely related to stenosis of the bowel outlet (23) (30). (d) Segmental colonic transit times in patients with ARM shows that low anomalies have rectosigmoid hypomotility and high anomalies have generalized colonic motility disturbances (42).
5. Surgical technique – in reconstruction might be an important prognostic factor. However, as there is no controlled trial, it is difficult to prove. Kieswetter and Chang (43) found abdominoperineal pull-through to be slightly better than sacroabdominoperineal operation. In a retrospective case note study Mulder et al (44) found a good continence of 40% in both groups of patients who had undergone sacroperineal pull-through and PSARP. Templeton and Ditesheim (45) suggested that the use of full thickness terminal bowel in the reconstruction gave

better results than endorectal pull through procedures. deVries (46) could not find any evidence to support the superiority of any procedure in anorectal reconstruction.

6. Timing of Surgery- the age of the patient at the time repair influences the long-term functional outcome. Neonatal abdominoperineal reconstruction of the anorectum was popularized by Rhodes et al. in 1948 and was popular in 1950s and 1960s. But many surgeons were not satisfied with the functional results and so started doing staged procedures (47). Recently early repair during the first 3-6 weeks of life is recommended, and some surgeons advocate neonatal surgery. The critical anal dilatations are easier to perform in an infant and it allows the early development for neural pathways between the anal canal and the brain, facilitating better anorectal sensation and sphincteric function. However, at present, there is no clear evidence that neonatal pull through procedures gives a better functional outcome than surgery done at 6-12 months of age (48).

Long-Term Bowel function During Childhood

Report of long-term functional outcome is variable. Most series grade the results as good, fair or poor. The good outcome does not necessarily mean the bowel function is normal but that they are socially continent. The table shows the results by various surgeons before the advent of PSARP

Table: 12 Long term functional after anorectal malformation

	N	Good	Fair	Poor
Partridge and Gough (49)	63	33%	43%	24%
Trusler and Wilkinson(50)	15	26%	20%	54%
Stephens and Smith (47)	25	56%	32%	12%
Taylor et al (51)	45	24%	20%	56%
Cywes et al (52)	38	42%	35%	23%
Smith et al (53)	18	6%	28%	66%

There are few reports of functional outcome following PSARP and here also the results have been inconsistent (Table: 13).

Table 13- Functional outcome after PSARP

	Total continence	Significant soiling	Constipation
Pena(23)	36%	41%	43%
Rintala and Lindahl (41)	35%	30%	60%
Langemeijer and Molenaar (54)	7%	56%	5%

The following results (Table:14) from follow up studies by Pena and Marc Levitt in children who had repair for ARM in terms of voluntary bowel movement, continence and constipation (55).

Table -14 Results of series by Pena

Types of malformation	VBM	Total continence*	Constipation
Rectal atresia/stenosis	100%	88%	57%
Perineal fistula	100%	100%	50%
Vestibular anus	92%	55%	61%
Imperforate anus with no fistula	89%	52%	55%
Bulbar urethral fistula	81%	31%	59%
Short cloaca	79%	28%	39%
Prostatic fistula	73%	20%	45%
Long cloaca	55%	17%	48%
Bladder neck fistula	35%	0%	15%
Series average	77%	39%	48%
VBM- Voluntary bowel movements * Voluntary bowel movements and no soiling			

Fecal Incontinence

Almost all patients who undergo repair of an anorectal malformation suffer from some degree of functional defecating disorder and abnormal fecal continence mechanism. Approximately 25% of the patients following surgery for ARM are fecally incontinent and cannot have a voluntary bowel movement. Fecal incontinence prevents a person from becoming socially accepted and results in serious psychological consequence.

Factors determining fecal incontinence

Fecal continence depends on (1) Voluntary sphincter muscles, (2) anal sensation, and (3) Colonic motility.

Voluntary Sphincter Muscles

The muscle structures include the levators, the muscle complex, and external sphincter. Patients with ARM have abnormal voluntary striated muscle with different degree of hypo-development. Voluntary muscle can be used only with the information derived from an intact anal sensory mechanism which many with children with ARM lack.

Anal Canal Sensation

Exquisite sensation in normal individuals resides in the anal canal. Except for patients with rectal atresia most patients with ARM are born without an anal canal; therefore sensation does not exist or is rudimentary. Patients are able to perceive distention of the rectum, but for this to be felt, the rectum should be placed within the muscle structure. This sensation is proprioception caused by the stretching of the voluntary muscles. As the liquid stool or soft fecal material does not distend the rectum no sensation may be felt by the patient. So for patients to achieve some degree of bowel sensation and bowel control, the patient must have the capacity to form solid stool.

Bowel Motility

In a normal individual, the recto sigmoid remains quiet for variable periods of time depending on the specific defecation habit. The peristalsis prior to defecation is felt by the patient and the individual relaxes the voluntary smooth muscle which allows the rectal contents to migrate to the sensitive anal canal. The consistency and quality of the stool is also discerned. The voluntary muscles push the rectal contents up into the rectosigmoid and to hold them, if desired, until the appropriate time for evacuation.

During defecation the voluntary muscle structures relax. The main factor that provokes the emptying of the rectosigmoid is a massive involuntary peristaltic contraction. Most patients with ARM suffer from disturbances of this sophisticated bowel motility mechanism.

Patients who have undergone a PSARP or any other type of sacroperineal-pull through in which the most distal part is preserved show evidence of 'megarectum' which acts as a over efficient bowel reservoir. The clinical manifestation is constipation. When the constipation is not aggressively treated this results in further dilatation of the ectatic rectosigmoid and worsening constipation. The enormously dilated rectosigmoid with normal ganglion cells behaves like a myopathic type of hypomotile colon (56). In these patients who are incontinent, a daily enema successfully cleans the colon (57) However patients who have their terminal bowel resected do not have the rectal reservoir and behave like a perineal colostomy. In these individuals, a daily enema with a constipating diet and medications to slow down the colonic motility is indicated.

Bowel Management Program (BMP)

BMP consists of teaching the patient or their parents how to clean the colon once daily as to stay completely clean in the underwear for 24hr. The correct type and quality of enema can be determined by trial and error method. The x-ray of the abdomen is taken to monitor the amount and location of any stool left in the colon. Modification of their diet and drugs is also made.

It is important to differentiate real fecal incontinence from overflow pseudo-incontinence. Pseudo-continenence occurs when they have severe constipation and overflow soiling. Once the disimpaction is treated and the patient receives enough laxatives so as to avoid constipation, the patient becomes continent. Generally 75% of the patients will have good voluntary bowel movement (VBM) after the age of 3 years. About half of these patients will still soil and is usually related to constipation. When the constipation is treated, the soiling stops. Thus approximately 40% will have normal VBM and no soiling, they behave as normal children. However some of these children will have episodes of fecal incontinence especially during episodes of diarrhea. Finally there will be around 25% of children who will be fecally incontinent. The surgeon should be able to predict in advance which children will have good functional prognosis (Table: 15, 16) and which will not. This will help the surgeons in giving a realistic picture of the child's chances of bowel control. This will avoid creating false expectations and frustrations later on, among parents.

Table : 15 Common indicators of Good and Poor Prognosis	
Good prognostic signs	Bad prognostic signs
Good bowel movement pattern (1-2 bowel movements per day and no soiling)	Constant soiling and passing stools
Evidence of sensation when passing stool (pushing making faces)	No sensation (no pushing)
Urinary control	Urinary incontinence

If the child is having a defect with good prognosis one can expect VBM by the age of 3 years. These children still need supervision to avoid fecal impaction, constipation and soiling. The child with poor prognosis, the BMP is needed to keep the child clean and it should be implemented when the child is 3 or 4 years. A child with a rectoprostatic fistula has a 50% chance being continent. Therefore effort should be made to achieve toilet training by the age of 3 years.

Redo pull through: if the child with good prognosis has been operated and is incontinent and has a misplaced rectum, a redo pull through can be performed to relocate the rectum within limits of the sphincter mechanism.

Table: 16 Predictors of Functional Prognosis	
Indicators of good prognosis for bowel control	Indicators of poor control for bowel control
Normal sacrum	Abnormal sacrum
Prominent midline groove	Flat perineum
Some types of anorectal malformation <ol style="list-style-type: none"> 1. Rectal atresia 2. Imperforate anus without fistula 3. Cloacas with common channel <3 cm 4. Rectourethral bulbar fistula 	Some types of anorectal malformation <ol style="list-style-type: none"> 1. Rectobladderneck fistula 2. Cloacas with acommon channel >3cm 3. Complex malformation

Children operated for fecal incontinence can be divided into well- defined groups that require individualized treatment plans: (1) those with constipation (2) children with loose stools and diarrhea

1. Children with Constipation (Colonic hypomotility): In these children the motility of the colon is significantly reduced. The child can be dry by cleaning the colon once a day with a suppository, an enema or colonic irrigation. The fact that they suffer from constipation (hypomotility) guarantees that they remain clean in between enemas. No special diet or medication is necessary.
2. Children with Loose Stools and Diarrhea: The majority of these children had resection of their rectosigmoid (reservoir). They have overactive colons with rapid transit of stools. The enema cleans the colon but stool keeps passing fairly quickly from the caecum to the descending colon and anus. To prevent this, a constipating diet and/or medications to slow down the colon are necessary and foods that further loosen bowel movements are avoided.

A contrast enema (with hydrosoluble contrast) will help in determining the type of colonic motility, hypo motility – constipated, or hyper motility.

Types of enema: There are different types of solutions that can be used for enemas: ready made enemas or solutions prepared at home based on salt and water (0.9% saline can be made by adding 3-4 teaspoons to 1 liter of water). Saline enemas are effective and less expensive. Phosphate enemas may cause abdominal cramps. Phosphate enemas can cause phosphate intoxication and children with impaired renal function should use with caution. Glycerin can be added to the saline enema to make it more effective. The frequency and the amount of enema also can be increased initially to get the desired result. The ‘right’ enema is the one that can empty the child’s colon and allow him to stay clean for the

following 24 hrs. This can be achieved only by trial and error and learning from previous attempts.

The child with the overactive bowel, along with the enema, needs constipating diet and drugs like loperamide are recommended. Most parents would eventually know which meals provoke diarrhea and which constipate their child.

Every summer holidays, the children with some potential for bowel control (refer Table: 16) can try to find out how well they can control their bowel movements without the help of enemas. This is tried during vacations to avoid accidents at school.

Continent catheterisable stoma: Children when they are in the preschool and school-age enjoy a good quality of life with the BMP. However, when they reach puberty they feel that parents are intruding on their privacy by giving them enemas. Though it is feasible, it is difficult to administer the enema themselves. An operation called continent appendicostomy or Malone's procedure (Malone's ante-grade colonic enema- MACE) has been designed for this specific group for children (58). The Malone procedure is just another way to administer enema, and therefore the child should be perfectly clean with the BMP before implementing the Malone procedure. The operation consists of connecting the appendix to the umbilicus, and creating a valve mechanism that allows catheterization of the appendix for the enema fluid, but avoids leakage of stool through it (59). The other advantages are easy access to the colon, one-way effective irrigation, and smaller volumes for irrigation, and physiological comfort.

Continent Cecostomy Techniques:

1. Disconnection and reimplantation of the appendix
2. Orthotopic appendicostomy (+/- divided appendix)
3. Tubularised cecal/ colonic flap
4. Transverse tubularized ileal tube (Monti)
5. Laparoscopic MACE (appendicostomy only)
6. Cecostomy button
7. Percutaneous cecostomy catheter
8. LACE (Left colonic antegrade colonic enema)

The main role of long term patent channels is to improve quality of life for patients.

There is an increase in self-esteem, happiness, and social acceptance.

Dietary Prevention of Constipation:

Dietary fiber: Roughage or fiber plays an important role in the passage of chime. Roughage increases the digestive juices and it swells up in the intestine due to its absorption of water. It serves as a culture medium for the bacteria in the colon. The breakdown of the fibers by bacteria creates gases and acids, which in turn stimulates the peristalsis of the intestinal wall. The consistency of the stools becomes softer, and the distension of the intestinal wall and increased propulsive motility shortens the transit time and reduces the water resorption. For the roughage to have the optimal effect, it is important to drink enough liquids. Roughage is indigestible vegetable material which can be found in leaves, fruits and roots. Nonpurified vegetable fibers are the fibers found in

cereals, fruits and vegetables. Purified vegetable fibers are fibrous and polymer substances such as lignin, cellulose and pectin. Foodstuffs containing lots of fibers are fruits and vegetable especially berries, dried fruits, green peas, pulses, whole cereals, bran, nuts and almonds.

Dietary stimulants: in addition to the mechanical stimulation, a chemical stimulation may also improve peristalsis. Lactic acid is an example and is found in yogurt, butter milk, pickles and vegetable juices. Tartaric acid found in grapes, apple, citric acid in citrus fruits and acetic acid in wine vinegar are believed to improve peristalsis. Lactose or concentrated sugar solution made of lactose also stimulate intestinal motility.

Operations to improve continence after previous surgery: Several techniques have been described to restore continence after previous repair of anorectal malformation. Secondary operations are done mainly in 2 groups of patients. The first being in patients with a benign type of defect like 'rectobulbar fistula' who is incontinent postoperatively due to totally misplaced rectum where the options are (1) Redo PSARP (2) Stephen's Secondary Pull through. The second group is represented by patients who had previous operation but who suffer from fecal incontinence. In principle the attempts to correct this has been based on

1. Secondary repair of levator ani – Stephens' secondary repair of damaged or hypoplastic muscle complex. (Stephens FD, Smith AD(1988)-(60)
2. Reinforcement and substitution for the levator ani – (a) Kottmeier's levatorplasty (61). (b) Puri and Nixon's levatorplasty (62).

3. Free autogenous muscle transplant for strengthening of the levator ani (Palmaris longus transplant) (63).
4. Substitution of striated anal sphincter (a) Gracilis muscle transplant (64) (b) Dynamic graciloplasty (c) Gluteus maximus transplant (65)
5. Construction of sphincter from the bowel wall (a) Free smooth muscle transplantation (b) Flap smooth muscle transplantation (66).
6. Electric devices to stimulate the sphincters (67)
7. Secondary procedures for anal prolapse or strictures (a) Nixon Anoplasty (68) (b) Mollard – Laberge operation (69)

Resection of inert recto sigmoid for the treatment of chronic constipation

Many children with chronic constipation after repair of ARM suffer from different degrees of dilatation of rectum and sigmoid, a condition defined as megarectosigmoid, due to a hypomotility disorder that interferes with complete emptying of the recto sigmoid (56). This is mainly due to inappropriate treatment for constipation, leading on to fecal impaction and overflow incontinence. The impaction is removed with enemas and/or colonic irrigation to clean the megarectosigmoid. The constipation is subsequently treated with large doses of laxatives. The dosage of the laxative is increased till the child is able to completely empty the colon everyday. If the medical management is difficult and the dosage required to treat constipation is very high, sigmoid resection may be beneficial. After sigmoid resection the amount of the laxative can be minimized or eliminated. Pena et al (70) followed up 53 cases after sigmoid resection. Following the resection 10% did not require laxative and the rest required significantly less amount of

laxative. It is found that the patients who benefit are those with localized form of megarectosigmoid.

Anal manometry

The goal of treatment for anorectal malformation is to achieve anal continence, and various attempts have been made to obtain objective assessment of it. The objective assessment gives accurate information about postoperative sphincter function and sphincter distribution providing further strategies for postoperative treatments. One of the objective assessments is by anal manometry. Anal manometry has been used to measure anorectal resting and squeezing pressure profile, the rectal adaptive reaction and the internal sphincter relaxation. According to Holschneider (71) the normal findings seen postoperatively after pull through were (1) Anorectal resting pressure profile 25+/-5mm of Hg. (2) Squeeze pressure profile of more than 35 mm of Hg (3) A Normal rectal adaptation reaction (4) Normal internal sphincter relaxation.

Manometric and clinical results have been often found contradictory. Some investigators have found a positive correlation between clinical continence and the anal resting pressure profile (30), others have observed a correlation between continence and voluntary squeeze force (72); still others have reported no correlation between clinical continence and pressure profile or squeeze force (73). Decreased rectal sensitivity at rectal distension has been reported to correlate with poor functional outcome (73). Patients can perceive distension of pulled through rectum but this requires a rectum that has been properly located within the muscles structures. The sensation seems to be a consequence of voluntary muscle stretching (proprioception). However the patient should

have solid stools to achieve some degree of sensation and bowel control. A skin lined anal canal is vital for continence. Kieswetter and Nixon showed an in growth of sensory nerves following pull through operation (12). The anal canal constructed in patients with rectourethral fistula is endowed with a high degree of sensation, content discrimination, and muscular sphincter function if the new canal is lodged within the striated muscle complex.

Improvement in bowel function with puberty

Fecal incontinence in patients having undergone repair for ARM improves at adolescence, as constipation disappears (74). This improvement with time is probably related to reinforced sphincter function and an increasing use of gluteal and pelvic floor muscles, and is a manifestation of the adaptation and adjustment made by the patient himself to achieve a socially acceptable status.

MRI

Imaging studies are used to assess long term anorectal function in patients with ARM. Imaging modalities like intra-anal ultrasound, computed tomography (CT) and MRI provide excellent information about postoperative anatomy of ARM. MRI has an advantage over CT because it gives superior tissue characterization, multiplanar imaging, and lack of ionizing radiation. MRI can detect factors related to poor outcome after surgery for ARM like (1) Hypoplastic sphincter complex, (2) Misplacement of bowel in relation to the sphincters, and (3) Obtuse anorectal angle (75). However the clinical results and MRI findings may not correlate.

One of the major goals of the surgical correction of ARM is to establish a good functional outcome. Adequate placement of the neorectum in both puborectalis muscle and external anal sphincter is essential to accomplish the goal (22, 23). The sphincter muscle can be clearly demarcated with MRI in multiple planes. The levator ani muscle and puborectalis muscle and their interrelationship with external anal sphincter are best evaluated on the coronal T1 weighted images (76).

Post operatively MRI can play a role in the evaluation of patients with persistent fecal incontinence (75). An inappropriate placement of the neorectum and or interposition of mesenteric fat in the sphincter muscle complex are causes of postoperative incontinence. MRI is able to demonstrate these operative complications affecting the functional outcome. Anterior misplacement of the neorectum in the external sphincter and the lateral misplacement of the neorectum in the puborectalis are the most common errors observed. MRI may demonstrate unsuspected maldevelopment of the sphincter muscle complex as a cause of persistent incontinence. However Arnbjonsson et al did not find a statistically significant correlation between the level of continence and the sphincter muscle thickness (77). In addition Fukuya et al stated that MRI evaluation based solely on muscle development can be misleading. Therefore they included a measurement of the anorectal angle in their postoperative evaluation and found a statistically significant difference in anorectal angle between patients with good and poor clinical outcome. Anorectal angulation represents sufficient contraction of the sphincter muscles in the post operative period. deSouza et al (78) used a qualitative index of sphincter appearance. A sphincter that appeared normal was scored 0, minimal asymmetry of the muscle deemed a mild deficiency and scored 1, a 25-50% reduction in the length / thickness of any sphincter

component compared to with the other side was deemed a moderate deficiency and scored 2 and such reduction that was greater than 50% was deemed severely deficient and scored 3. Tang S et al (79) measured anorectal muscle complex in normal children using phase- arrayed MRI and found that absolute measurement values muscles cannot be compared among children of different ages. So he used the ratio between the absolute width of the muscle and the transverse anteroposterior diameter of the pelvis, as the relative width of the muscle to avoid the influence of age. In order to avoid error interference with asymmetrical development and smaller measurement value of the muscles, relative width, the ratio between total muscle width measured at 3-and 9-o'clock position and the half distance between the ischial tuberosities was taken. The sum of right and left width of puborectalis at the PC level was 7.9 mm (range 5.0-9.8mm) and the width of the EAS, the sum of the right and left parts, was measured as 7.6mm (range 4.9-8.6mm). The relative width of PR and EAS was 0.5mm and 0.44mm respectively.

MATERIALS AND METHOD

This study was conducted in 42 children with intermediate anorectal malformation, operated by sacroperineal pull through in the Department of Paediatric Surgery, Christian Medical College, Vellore from 1996 to 2005. The children were assessed by a person not involved with the surgery.

Exclusion criteria:

1. Children with low and high anorectal malformation
2. Children with intermediate anorectal anomaly operated by surgery other than sacroperineal pull through
3. Children with anorectal malformations who were operated elsewhere
4. Children with associated Hirschsprung's disease.

Methodology:

All children operated in the Department of Paediatric Surgery for intermediate anorectal malformation from 1996 to 2005 were called for the study.

166 children who had sacroperineal pull through done from 1996 to 2005 were called for the study. 42 children, who responded, were selected for the study. The follow up period ranged 14 to 144 months with a mean follow up period of 6.5 years.

Family demographic details including socioeconomic status, age and education of the parents were noted.

Associated anomalies were also noted.

All the 42 children underwent clinical evaluation to assess their somatic growth and functional status with respect to fecal continence. The children were graded according to

Krickenbeck classification for postoperative results. X-ray of the abdomen was done to assess the fecal load.

Out of these 13 children with intermediate type of anomaly had MRI evaluation of the pelvis to determine the relationship of the pulled-through bowel with the sphincter muscle complex. MRI examination was done on either 1.5T or 3 T magnet. All subjects were positioned supine with the pelvis centered on the coil and a body phase array was used. Sagittal, coronal and transverse T2 – weighted images of the pelvic region was obtained in all subjects. Slice thickness was kept at 3 mm with interslice gap of 0.3. Anorectal angle and thickness of the sphincter muscle was also studied.

Anal manometry was done in children in 18 children. Balloon tip pressure transducer probes were used. Pressure was recorded at 1cm, 2cm and 3cm of anal verge and the maximum value was selected. Anal resting pressure profile and anal squeeze pressure profile at voluntary squeeze was recorded. Afterwards a balloon tipped catheter was passed into the rectum and inflated and the volume at which the first sensation, urge to defecate and the maximal tolerable volume was noted. Anal sensation was assessed by electrical stimulation using small electrode placed 1 cm into the anal canal and passing a small current at 15 output.

Anal pressure profile, rectal and anal sensation were also noted.

The psychosocial evaluation of both the child and the parent was done. Parents were assessed for parental stress based on their reaction to the deformity, relationship with spouse and relatives, attitude towards the child financial implications of the treatment, future worries and whether they were satisfied with the final postoperative results. The

child assessment was made based on the schooling, interaction with friends and whether aggressive behavior was evident or not. Quality of Life was assessed for the children.

Intervention

Aggressive bowel management program was started for all those children who had fecal incontinence. The children were reassessed after the bowel management program and revised scoring was done with Krickenbeck scoring system and Quality of Life (Bai) scores (61).

Statistical Analysis

Statistical significance was assessed using chi-square test. Statistically significant difference were observed if $P < 0.05$.

RESULTS AND ANALYSIS

A total of 42 cases were followed up who had undergone sacroperineal pull through from 1996-2005. The ages ranged from 2.5 years to 13 years with a mean of 7.5 years. Of the total, 40 were males and 2 were females.

DURATION OF FOLLOW UP

Follow up after colostomy closure ranged from 14 months to 144 months with a mean follow up of 78 months (6 1/2 yrs).

AGE OF PARENTS

The average of the father was 29.8 years and that of the mother 23.6 years.

EDUCATION

The level of education of the parents was available for 41 fathers and 41 mothers.

Table: 1 Level of parents' education

Class	Father	Mother
None	4	4
< 5 th	5	6
6 th -9 th	10	9
10 th -12 th	17	16
Graduates	5	6

INCOME OF THE PARENTS OF THE CHILDREN (Table: 2)

The income of the families ranged from less than Rs.1000 per month to about Rs.10000 per month. In 24 families the income was less than Rs.1500 per month, 9 had income above Rs.1500 but below Rs.3000, and 8 had an income between Rs.3000-Rs 5000. Only one family, who were in business, had a monthly income of Rs.10000.

Table: 2 -Monthly income of the families

Monthly Income(RS)	Number of families
<1500	24 (57.14%)
1500-3000	9 (21.42%)
3000-5000	8(19%)
10000	1(2.3%)

POSITION OF CHILD IN FAMILY

18 (42.8%) of these children were the first born, 14(33.33%) were 2nd and 10(23.8%) were 3rd.

ASSOCIATED ANOMALIES

Associated anomalies were seen in 15 of the 43 patients. 4 patients had hypospadias of which 3 were penoscrotal and 1 was distal penile hypospadias. Three patients had major vesicoureteric reflux and 3 had undescended testis. Renal anomalies like dysplastic kidney, crossed fused renal ectopia and horse shoe kidney were seen in one patient each.

The child with crossed fused ectopia had associated vaginal atresia. Other anomalies seen were exomphalos, esophageal atresia with tracheo-oesophageal fistula and lobster claw deformity with rudimentary thumb. Twelve children had vertebral anomalies which are described next.

VERTEBRAL ANOMALIES

9 children had sacral anomalies and according to Cama (80) classification of sacral anomalies, 8 children type 3 sacral anomaly and 1 child had type 2. Two children also had lumbar hemi vertebrae and one had multiple vertebral anomalies.

HEIGHT AND WEIGHT (were available for 36 children)

The height and weight were plotted against the growth curve graph recommended by the Growth monitoring Guidelines Consensus Meeting of Indian Academy of Pediatrics (81).

The reference growth chart was applicable to the Indian population.

20/36 (55%) of the children were below 25th percentile for height and 27/36(75%) were below 25th percentile for weight (Table: 3).

Table: 3 Growth pattern in children with anorectal malformations

Height(percentile)	No:	Weight (percentile)	No:
< 3	8	<3	11
3-25	12	3-25	16
25-50	8	25-50	5
50-75	6	50-75	3
50-97	2	50-97	1

PERINEUM

Position of neoanus

The anus post operatively was in the normal position in all patients.

Patency

19 of the 42 (45.2%) children had a supple and patent neo anus. One was patulous and the remaining 22 (52.3%) had varying degrees of stricture. All the strictures were at the skin level. 12 were major strictures out of which 4 underwent anoplasty; the rest of the cases had mild strictures. One case which underwent anoplasty also had shelving.

Mucosal prolapse

15 of 42 (35.71%) cases had mucosal prolapse. Six were minor and 9 had major prolapse which required mucosal trimming.

Secondary anal procedures

13 (31%) patients underwent secondary anal procedures. Four anoplasties for strictures and 9 mucosal trimmings were done.

Urinary symptoms

Four children had features of neurogenic bladder on follow up. Three of them presented with dribbling and one of them, with retention of urine. Two of them are on CIC and drugs while the other two are on drugs (amitriptyline and oxybutynin) alone.

BOWEL FUNCTION

Krickenbeck scoring system

1) Voluntary bowel movements were present in 35/42 (83.3%) patients.

2) Soiling

1. No soiling was seen in 5 (11.9%) patients
2. Grade -1 soiling seen in 12 (28.57%) patients
3. Grade-2 soiling in 7 (16.66%) patients
4. Grade-3 soiling in 18 (42.85%) patients

3) Constipation – as perceived by parents

1. No constipation seen in 27 patients
2. Grade-1 constipation in 6 patients
3. Grade-2 constipation in 6 patients
4. Grade-3 in 3 patients

Constipation

Constipation as perceived by the parents was incorrect as evident by the x-ray findings of fecal load.

Fecal load as seen on X-rays

X-rays were evaluated in 38 pts.

Abdominal X-rays of the patients were taken to assess the fecal load in patients on whom bowel management program had not been earlier started, and a score of 1 to 4 were given according to the extent of fecal matter present in the colon. Score of 1 was given if the fecal load was limited to sigmoid and ascending colon, score of 2 if the fecal load extended up to the whole of transverse colon and score of 3 if the fecal load extended up to the caecum. Score of 4 was given if a mega rectum or mega sigmoid was evident.

(Table: 4). **Radiological extent of fecal load**

Fecal load extent	Grade	No. in the study
Sigmoid and ascending colon	1	7 (18.4%)
Whole of transverse colon	2	5 (13.15%)
Upto caecum	3	26 (68.4%)
Mega sigmoid/ mega rectum	4	0

According to the above scores, score of 1 was seen in 7, score of 2 in 5 and a score of 3 in 26 patients (Table:4).

Table: 4

Constipation, of any degree, was reported by parents of 15 children, only. However, fecal loading (Grade 1-3), was seen in all the 38 children in whom x-rays were taken, indicating that they were actually constipated, and had significant fecal retention. 31 of these 38 children (81.57%) had major fecal retention (Grade 2 and 3). So, constipation was a significant problem.

Soiling

Out of 42 children, 37 (88%) had some form of soiling. Major soiling of grade 2 and 3 were seen in 25 (59.5%).

Table: 5 Soiling with relation to level of X-ray fecal load

Grade of Soiling	Grade of fecal load in x-ray	No:
3	3	13
	2	3
	1	1
2	3	4
	2	1
	1	2
1	3	7
	2	1
	1	3
0	3	2
	2	0
	1	1

Soiling compared with fecal loading

Out of the 17 children who had Grade 3 soiling, 16 had features of major fecal retention (Grade 3 or Grade 2) on the x-ray.

Out of the 7 children with Grade 2 soiling, 5 had features of major fecal retention on the x-ray.

Out of the 11 children with Grade 1 soiling, 8 had evidence of major fecal retention on the x-ray.

Therefore out of the 35 children who have soiling in some form or other, 29 had x-ray evidence of major fecal retention (Grade 3 or Grade 2), i.e. 82.8%. Out of the 3 who have no soiling, only 2 had x-ray evidence of major fecal retention (66.6 %).

Anal manometry

The average anal resting pressure was 52.5 cm of H₂O and the average voluntary squeeze pressure was 144.2 cm of H₂O.

Squeeze pressures were done in 14 children. All, except 1 child who had a rectovaginal fistula, had squeeze pressures above 30 mm Hg (40 cm of H₂O) (score of 2 in Holschneider's scoring system) (Table: 6). One child in whom a squeeze pressure could not be elicited at all, has constant soiling and the MRI showed sacral agenesis and the sphincter was extremely thin superiorly and scarring was seen at 3 and 9'o clock positions.

Of the 13 children with good squeeze pressures, 11 have soiling in some form. However, we believe that these children have the potential to improve, when they cross puberty.

Table: 6- The pressure profile was correlated with the degree of incontinence

Grade of soiling	RP (cm of H ₂ O)	VSP (cm of H ₂ O)
Grade 3 and 4	47.8	94.8
Grade 1 and NS	57.11	173.8
RP- Resting pressure, VSP- voluntary squeeze pressure NS-No soiling		

The rectal sensation was determined by inflating the rectum with a balloon. The initial sensation at distension was present at an average of 36 ml, there was an urge to defecate at 65ml and the maximum tolerable volume was on average 80ml. This was correlated with soiling.

Table: 7- Soiling in relation rectal sensation

Grade of Soiling	1 st sensation	Urge to defecate	Maximum tolerable limit
Grade 2 and 3	25.8ml	53.16ml	64ml
Grade 1 and NS	45.28ml	75.7ml	94ml
NS- No soiling			

Anal sensation was assessed by electrical stimulation of the new anal canal. Anal sensation was tested in 15 children. 11 (73.3%) children could appreciate anal sensation with an average of 10.18 mA and 4 patients could not appreciate anal sensation with electric stimulation. 7/11(63.63%) children who had anal sensation had grade 1 or no soiling whereas 1/4 (25%) children who had no sensation on electric stimulation had grade 1 soiling

Table: 8 Soiling tabulated along with fecal load in x-ray, anal pressure profile and anal sensation

Sl.no.	Soiling	Constipation	Fecal load (x-ray)	VSP (cm of H2O)	Resting Pr (cm of H2O)	Anal sens (mA)
1	3	0	3			
2	0	0	1		40	NA
3	2	0	3			
4	1	2	1	90	20	6
5	1	0	1			
6	3	2	3			
7	2	0	3			
8	3	0	3	180	40	10
9	3	0	3			
10	3	0	3		30	NA
11	1	0	3	130	80	6
12	3	0	3			
13	0	3	3			
14	1	0	3	110	70	Nil
15	0	2				

16	3	0	2	0			
17	3	0	1			0	Nil
18	0	2	3	309			
19	1	1	1	227		45	6
20	2	0	1			63	10
21	1	0	3				
22	3	2	3	125			
23	2	0	2			50	Nil
24	1	1	2	100		30	NA
25	2	0	3			30	12
26	3	0	2				
27	1	3	3				
28	2	1	3	130			
29	1	0	3			70	20
30	3	0	2				
31	3	0		80			
32	1	2				80	12
33	3	0	3	136			
34	3	0	3			81	nil
35	3	0	3				
36	3	0	3				
37	3	1	3				
38	2	1	1				
39	1	3	3	275			
40	1	0	3			91	10
41	3	0	3	170			
42	0	1		150		50	6
						75	14

NA: Not assessed, Nil: No sensation

MRI

Magnetic resonance imaging was done in 13 children. The position of the rectum was central in 10/13 (76.9%). In 2 of the children it was slightly on to the right and in one child the pulled through bowel had slipped out through a defect in the right side. 6/10 (66%) of children with properly placed rectum had no soiling or grade 1 soiling and 4/10 (40%) children had major soiling. One child with major soiling had severe sacral agenesis in the MRI.

In the MRI of 12/13 children the anorectal angle was clearly seen in the sagittal plane. The average anorectal angle was 122.1. In 4 children the angle was above 121 and major soiling (grade 3) was seen in 3/4 (75%) of them. In 9 children the angle was below 121 and major soiling was seen in 3/9 (33.33%). The puborectalis muscle (PRS) was thin in all patients. In 2/3 patients PRS was deficient on the right side. In these children the pulled through rectum was eccentrically placed to the right in one patient and completely slipped out through the defect to the right. Both of them had grade 3 soiling. The external anal sphincter was measured at the I-plane and M-plane (midway between I line and PC line). The maximum thickness was taken on either side of the rectum. If the thickness on one side was greater than twice that on the other side, the rectum was judged to be

mispositioned in relation to striated muscle complex (82). Accordingly 3/12 (25%) children were mispositioned in relation to the striated muscle complex. The axials were suboptimal in one of the patient. 2/3 (75%) children who had the rectum mispositioned had major soiling. The sphincter thickness was scored in our study according to deSouza (78). In our study 12 children the sphincter thickness was scored. 7/12 (58.3%) had score of 1, 2/12 (16.66%) had score of 2 and 3/12 (25%) scored 3. In 7 children with score of one, 2/7 (28.57%) had major soiling and 5/7 (71.5%) had minor/ no soiling. In 2 children with a score of 2, both (100%) had major soiling and the children with a score of 3, 2/3 (66.66%) children had major soiling.

Psychosocial Evaluation

PARENTAL STRESS

Initial reaction

The birth of a child with an anorectal malformation had various emotional impacts on the parents, especially the mother. (14/42) 33.33% mothers described fear as the initial reaction on hearing about the birth of their child with ARM. Sadness was the initial response of another fourteen parents. Three parents contemplated abandoning the child. There was wonder in the village in the case of few children, as this condition was unheard of. Few parents were confused and perplexed and two mothers felt this was from God and accepted the deformity.

Where they properly informed?

Thirty seven (37/42) 88% parents said that they were properly informed regarding the condition, treatment and complications.

Resentfulness / taking it out on the child

18/42 (42.85%) mothers said that they were at some point of time angry with the child especially in the initial periods before the commencement of the bowel management program(BMP), when they had soiling and they used to take it on the child either verbally or physically. This reaction was more when others especially their neighbors complained about the smell because of soiling and sent them off from playing with their children.

Socializing

22/42 (52.38%) parents said that in one way or other they restricted themselves from socializing with others mainly because of soiling and the attendant embarrassment. One family makes the child clean and avoids disclosure of the child's condition in order to socialize with others. A few parents are socializing more, as their children have grown up with some amount of continence attained by the BMP or otherwise.

Blaming mother

Six mothers said that they were blamed for the child. One father in anger threw the child from the bed as the child had soiled the bed sheets. Four mothers complained that the in-laws blamed the mother for the birth of the child.

Care giver

Mother was the main care giver in 41/42 (97%) cases. (1 child was adopted by the aunt). 18 (42.85%) of them had some help from their parents and 13 of the children had one of their grandparents especially maternal, helping the mother in taking care of the child. The role of the mother was in the majority, one of never ending chores of washing and cleaning soiled linen, with hardly any time for herself, and her work was often taken for

granted and unappreciated. All mothers broke down and cried during the interview while they elaborated on the role as care giver

Marital life

Marital life was affected in one way or the other in 16 (38%) families, because the mother was involved in taking care of the child most of the time. One family delayed having a second child for the fear of having the same abnormality in the next child.

Behavior of other children towards child

18/42 (42.85%) children had problems with other siblings and friends. They were completely avoided or made fun of. 11(26%) children remain secluded and did not mingle much with others.

Strained relations

Mothers of 10 (23.8%) children said that they had strained relationships with the relatives, especially the in laws. Since most families lived with the paternal grandparents, an 'abnormal child' and especially a fecally incontinent child was not fully accepted. Many mothers had to move with the child to her own parent's house. Acceptance came much later as the child grew up.

Future worries

Almost all mothers were worried about the child's future. The main area of concern was the child's future studies and marriage in 23/42 (54.76%). As many children were dependent on their mothers for the toilet needs, many mothers were concerned as to how their child would manage alone in the future. Interestingly six of the parents did not feel that the malformation was a reason for more concern about their child's future.

Satisfaction with final result

20/42 (47.61%) parents were fully satisfied with the outcome and 22 parents were either not satisfied at all or satisfied partly (11 each). In the children whose parents were satisfied 7 (35%) had major soiling and 13 had none or minimal soiling. In the children whose parents were not satisfied or not fully satisfied, major soiling were seen in 19 (86.36%) patients compared to 3 with minimal or no soiling. There was a statistical significant association between soiling and the parental satisfaction (statistical significance assessed by the chi square test; $\chi^2=11.72$, $p< 0.001$)

Table: 9 Satisfaction with results		
Satisfied with outcome	Soiling Gr	Number
Yes	3	4
	2	3
	1	8
	0	5
No	3	7
	2	2
	1	2
	0	0
Not Fully	3	8
	2	2
	1	0
	0	1
Grade 0 = No soiling		

Expenses

Only 3 (7.1%) patients could afford payment on their own and the rest 38/42 (92.8%) needed financial help. Four patients had relatives helping them. Twelve parents (28.57%) could pay for their treatment only after selling off their property, house, cattle or some valuables. Parents of seventeen children (40.47%) went into debt with the treatment of their child and six of them had not fully paid back the debt at the time of this study. This was in spite of concession given to 8 of them. Four children were given free treatment.

CHILD ASSESSMENT

Schooling and school absence

Thirty children (71.42%) are going to school in classes ranging from kindergarten to 7th. Four children of school-going age are not going to school. Parents of 2 children were not willing to send them to school for fear of soiling. One child refused to go. The fourth one had Down's syndrome. He was not allowed admission in a nearby school because he was a child with Down's, and the mother could not send him to a school for children with 'special needs', as it was far from home and had only residential facility.

11/30 (36.6%) children are going to a class level lower than that for their age. Parents prefer their children to go a school nearby as they can come home for use of the toilet and to clean themselves when they soiled.

7/30 had frequent absence from school.

Withdrawal and aggressive behavior

Six children often withdrew from other children and played by themselves due to fear of teasing.

19/42 (45.23%) children showed aggressive behavior.

Quality of life (QOL) score

QOL score was assessed in 32 school going children from ages 5 and above. The average QOL score was 9.1.

QOL score	Grade	Number
9-13	Good	21
5-8	Fair	9
0-4	Poor	2
Total		32

QOL ON FOLLOWUP

The QOL score was assessed in 11 children on follow up after institution of bowel management program. The average QOL scores increased from an average of 8.1 to 12 that is from 'fair' to 'good'.

QOL	Grade	Number	QOL(after BPM)	Grade	Number

9-13	Good	6	9-13	Good	11
5-8	Fair	3	5-8	Fair	0
0-4	Poor	2	0-4	Poor	0
Total		11			11
BMP =Bowel management program					

On follow after bowel management program the QOL score of 6 patients improved from 'poor' or 'fair' group to good score. The children who were in the 'good' group improved on their QOL scores. There is a correlation with bowel management program and increase in the QOL score which was statistically significant (t-value is 5.17, $p < 0.001$)

DISCUSSION

This study was done in children who had sacroperineal pull through operations for intermediate anorectal malformation according to the International classification (22) in Christian Medical College, Vellore from 1996 to 2005. A total of 42 children were included in the study. Children were followed up from 14 months to 12 years from the time of colostomy closure.

The age of the patients ranged from 2.5 years to 13 years with an average of 7.5 years. The majority of children were male 40/42. This might represent a true male preponderance in intermediate anomaly group (83) or may be because many girls are not brought for follow up in this society.

The parents at the birth of the children with anorectal malformation were very young (mean age-23), and they, especially the mother, most of the time, single handedly took the responsibility of looking after the child.

Of the parents who had some form of education, 46% (19/41) had only an elementary education, and only 5/41(12%) of the fathers and 6/41(14.6%) of the mothers were graduates. Educated parents were more regular in their follow up for their children.

Majority (57%) of children belonged to low socioeconomic group with a monthly income below Rs.1500. It was difficult to meet the expenses of the treatment of the anorectal malformation which was done in 3 stages with 3 separate admissions to the hospital.

Majority of the children (18/42) were the first born. Hassink et al (84) reports 'the idealized image of a perfect baby the parents may have, is instantly shattered with the birth of the first child. All of a sudden, they have to cope with the unexpected situation of having a child with an ARM and the consequences of the disorder, such as hospitalization, treatment and possibly other congenital defects in the child, and also in the subsequent children'. They are often not prepared for these challenges. Therefore, there is a great need for such parents to be adequately counseled and supported throughout their treatment period.

Associated anomalies mostly involved the urogenital system 13/42 (31%) and the commonest was vesicoureteric reflux.

The somatic growth in the children was affected as 28/36 (77.7%) and 32/36 (88.8%) of children were below 50 percentile for height and weight respectively. This might also reflect the poor socio-economic status of the family or even the negligence of the child. Studies on the somatic growth pattern in patients on follow up of pull through operation are lacking. In 13 patients (59.2%) the height and weight were less than the 50th percentile of the expected values for their age in a follow up study of pouch colon (85).

Perineum

The position of the neoanus was in normal position in all the patients. The anal opening had major anal stenosis in 12/42 patients (28%) and all the strictures were at the anal verge. Major mucosal prolapse was seen in 9/42 (21.42%). In literature the incidence of

anal stenosis and mucosal prolapse ranges from 15-78% (86). The local anal problems are less after posterior sagittal anorectoplasty (23). In our series there was 13/42 (31%) of secondary anal procedures (Anoplasty-4 and anal mucosal trimming-9). Our tendency had been to leave the bowel protruding out of the anal verge, out of fear of retraction which would explain the high incidence of mucosal prolapse. Pena emphasizes suturing the bowel to the anal verge under mild tension and accurate skin to mucosa approximation. Anal stenosis in our series was high because there was no strict anal dilatation program. Pena considers lack of anal dilatation as an important factor for anal stenosis and advises a strict dilatation program starting 2 weeks after repair. The parents are advised to dilate daily twice and continue dilatation beyond closure of colostomy according to the following protocol: once a day for 1 month, every 3rd day for a month, twice a week for a month, once a week for 1 month and once a month for 3 months (87)

Urinary incontinence

3/42 (7.1%) patients had urinary dribbling. The urinary symptoms may be because of the neurological damage following repair, which may present as retention and may recover. It may be as a result of spinal abnormalities causing neurogenic bladder. Mosiello and colleagues observed neurogenic bladder in 8 out of 39 patients with low lesions, 18 out of 45 patients with high lesions and all 5 cloacal patients (88). In Pena study the urinary incontinence in the entire series was 9.3% (23).

Bowel Function

It is difficult to compare results of different series in the past due to lack of uniformity in the classification of the defects.

Voluntary bowel movement was seen in 35/42(83.33%). Children who had VBM experienced sensation of rectal fullness and indicated the desire to defecate The VBM of the similar subgroup of intermediate anomaly which included recto bulbar fistula, imperforate anus without fistula and vaginal fistula (29) were 10/15 (66%) in a study by Bliss (89) and 54/68 (79.4%) by Pena (23). However total continence i.e. presence of VBM with no soiling was seen only in 5/42 (11.9%) in our series probably because we did not include any patients even with slightest soiling. Total continence in a similar subgroup was 4/15 (26.66%) in the series by Bliss (89) and 25/68 (36.76%) by Pena (23). The occurrence of voluntary bowel movement does not permit an assessment with regard to continence as only a small percentage of patients with high frequency of voluntary bowel movements show total continence.

Table: 1 - Patients with Voluntary Bowel movements and Total continence

Series	VBM		Total continence*	
	Patients	%	Patients	%
Seattle CHMC(89)	10 of 15	66.66%	4 of 15	26.66%
Pena Study (23)	54 of 68%	79.4%	25 of 68	36.76%
CMC Vellore	35/42	83.3%	5 of 42	11.9%

*Voluntary bowel movement and no soiling

Constipation was seen in 59% of bulbar fistula, 55% in imperforate anus without fistula and 45% of prostatic fistula on follow up in a series of 1192 patients by Pena (55)

which was comparable to his early results (23) of 387 patients followed up which showed constipation in 55.5% of bulbar fistula, 50% of imperforate anus with no fistula, 41.4% of prostatic fistula and 25% of vaginal fistula. Another series by Chau-Jing (90) had constipation in 21% of bulbar and 17% of prostatic fistula. Constipation is the most common sequelae after ARM repair (55). In our series the incidence of true constipation was 100%. The parents perceived constipation erroneously so we considered fecal loading seen in the x-ray as indirect evidence of constipation. The fecal load of grade 1 was considered as constipation grade 1 and fecal load above grade 2 was considered as constipation above 2.

Accordingly grade 1 constipation (fecal load of grade 1) was seen in 7/38 (18.42%) and grade >1 constipation (fecal load >1 grade) was seen in 31/38 (81.57%).

Soiling was seen 37/42 (88%) in our series and out of it grade 1 soiling was seen in 12 (28.5%) and soiling above grade 1 was seen in 25 children (59.5%). In Pena's study (87), imperforate anus without fistula had grade 1 soiling in 1/18 and soiling above grade 1 in 6/18 patients with a total 38.9%. Bulbar fistula had grade 1 soiling in 14/48 patients and above grade 1 soiling in 17/48 with a total of 64.6%. Prostatic fistula had grade 1 soiling in 11/58 patients and above grade 1 soiling in 32/58 patients with a total of 74.1%. Vaginal fistula had grade 1 soiling in 2/4 patients and above grade 1 soiling in 2/4 patients with a total of 100%.

Soiling is related to constipation and fecal overloading. So by treating fecal overloading by laxatives, diet or enema soiling can be reduced. The high incidence of soiling in our series is high because of the fact that many patients did not come for regular follow-up and had fecal overload and overflow incontinence of stools. With regular follow-up, with

clearing of the fecal load and bowel management the incidence of soiling is likely to come down and a trend towards this is already evolving in our patients.

One case of rectovaginal fistula had constant soiling and urinary incontinence also. The MRI pelvis showed sacral agenesis. Pena had 4 cases of vaginal fistula and soiling was seen in all. Pena (23)

Anal manometry

Resting anal pressure : According to Schweinger (91) internal sphincter contributes to 85% of the anorectal pressure profile resting pressure. The presence of internal sphincter in the pulled through bowel of ARM however is disputed. Rintala observed good continence in children with positive internal anal sphincter relaxation and a high anorectal pressure profile in patients with internal sphincter preserving repairs(92) Chen-Lung reported that if internal anal sphincter relaxation was positive and a high anorectal profile was established, the development of constipation was six times higher(93).. Sangkhathat by studying 24 children who had ARM came to the conclusion that rectoanal inhibitory reflex plays a crucial role in emptying function after anoplasty and that these functions should be preserved during reconstruction (93). The deepest point of the rectal pouch does not always correspond with the rectal origin of the fistula or the confluence to the urethra. Only in patients without fistula is the lowest point of the rectal pouch thickened by a cluster of smooth muscle cells that could be easily implanted in the perineum inside the external sphincter fibers. In many patients with fistulae, however, the smooth muscle fibers are on a higher level than the deepest point of the pouch.

The voluntary squeeze pressure profile depends on the quality of the sphincter complex, some may be dysplastic, and it indicates the presence of bowel within the sphincter complex.

The anal resting (RP) and voluntary squeeze pressure (VSP) were assessed in our series: The average resting and voluntary squeeze pressure was 52.5 and 144.2 cm of H₂O respectively. In children with severe soiling the RP and VSP were 47.8 mm of H₂O and 94.8 mm whereas in children with mild or no soiling it was 57.11 and 173.8 mm H₂O of respectively. In a study by Diseth and Emblem (31) the anal canal resting and squeeze pressure was significantly lower than that of controls. The anal canal pressure in patients with high anomaly was significantly lower than in those with low anomalies. The anal pressure profile were RP of 65 and 50 cm of H₂O in low and high atresias respectively compared with 98 in controls; and a VSP of 18 and 30 in low and high atresia respectively compared to 200 cm of H₂O in normal controls.

Rectal sensation was assessed in our children using balloon tipped catheter and volume of 36ml, 65 ml and 80 ml caused the 1st sensation, urge to defecate and maximal tolerable volume respectively. In children with severe soiling the volumes were 25.8ml, 53.6 ml and 64 ml respectively. In children with mild incontinence or continent children, the volumes were 45.28ml, 75.7 ml and 94 ml.

A skin lined anal canal is vital for continence. Kieswetter and Nixon showed an in growth of sensory nerves following pull through operation (12). Anoderm in many cases is well developed or reconstructed by Nixon's anoplasty or retraction at the neoanus at the end of the procedure after having sutured the distal bowel under tension to the perineal skin.

In our series anal canal skin sensation by electric stimulation, was present in 11/15 (73.3%) and 7/11 (63.63%) children were continent or had grade 1 soiling.

MRI

1. Anorectal angulation: in a study by Hisato, anorectal angulation was clearly seen in the intermediate anomaly operated by PSARP and sacroperineal pull through (95)

2. Misposition of the rectum- In the same study done by Hisato (95) there was misposition of the rectum in 21% in PSARP and 25% in the controls who had conventional surgery. In our series 13 children underwent MRI of the pelvis. The position was central in 10/13(76.9%) and in the remaining 3 it was misplaced. However it was within the sphincter complex in 2 and one child had the rectum slip posterior and to the right through a defect of the sphincter on the left side.

The anorectal angle was preserved in 12/13 children. The average angle was 122.1. In children with angle above 121, 3/4 (75%) had major soiling and in children where the angle was less than 121 only 3/9 (33%) children had major soiling. Children with anorectal angle above 121 had 2.25 times risk of having soiling compared to children who had an anorectal angle below 121. (R.R=2.25 95% CI (0.76, 6.65) It was not statistically significant. The puborectalis was thin in all patients. The sphincter muscle measurement was done in 12 children. The sphincter thickness measured in our series showed that 3/12 children had mispositioned rectum in the sphincter complex according to criteria proposed by Tsuji 82). 2/3 children had major soiling. In Tsuji's series 3 patients (21%) had mispositioned rectum in the sphincter complex. According to deSouza,

the thickness of the sphincter muscle was scored. 7/12 children had a score of 1, 2 children had score of 2 and 3 had a score of 3.

Parental Stress

About 74% (31/42) of parents indicated they were emotionally affected by birth of baby with anorectal malformation. The most frequently described emotional effects were sorrow and apprehension. There was wonder in the village. A similar finding of 75% of parents indicating fear and apprehension was noted by Hassink (84).

Most of the parents (88%) parents said they were well informed by the concerned surgeon regarding all the aspects of anorectal malformation. This is similar to finding by Hassink (84). In another study done by self help grouping Germany SoMA, parents received no information (4%) or insufficient information (42%) about the malformation prior to surgery (96) The parents (18/42) 42.85% indicated that they were angry with the child and even some indicated that they physically took it on the child.

More than half of the parents (22/42) 52.38% kept some restriction in socializing mainly because of the fear of fecal incontinence. One parent would conceal the handicap and made efforts to socialize. This was also mentioned in the study by Hassink (97) Children had difficulty in making friends and difficulties in peer relationship in our series 18/42 (42.85%). Hassink et al (97) found that 40% of children with anorectal malformation had difficulty making friends until age 13. One study showed that fecal incontinence did not affect peer relationship. However other studies have shown difficulties in peer relationships among children with imperforate anus because of the fear of incontinence (39). When a child is incontinent for feces, there is always a risk of comments from

friends. The smell of feces is difficult to hide (98). In 18/42 38% of the parents, birth and care of the child with anorectal malformation affected their marital life adversely. In another study by Diseth (98) 59% of parents reported their child's malformation negatively impacted their marital relationship and family life.

Mothers assumed the main responsibility of taking care of the child 41/42 (97%). Only half of them had the father (18/42) or the grandparents (13/42) involved in the care. This fact is also reported by Nisell (99). For the mother it was never ending chores of washing and cleaning soiled linen with hardly any time for her self, which was often taken for granted and unappreciated. Invariably all the mothers cried during the interview. A similar experience was noted by Nisell et al (99).

The parents worried about the future of the child in 23/42 (54.70%). The main worries were future studies and marriage. Ekkehart W.D (96) reported 66% had serious concerns regarding the future of the child. Many parents worried about how the children will manage when they are alone. Nisell et al (99) described the same concern of the parents in their study.

Satisfied with the final result:

The parents hoped and believed that surgery would restore a normal anus with normal function, and they are disappointed when this failed to occur (99). In our study 20 parents were satisfied and 22 were either not fully satisfied or not satisfied at all. Incontinence was the main reason for parents' dissatisfaction. In the parents who were not satisfied, 19/22 (83.63%) of the children had major (grade 2 and 3) incontinence. There was a statistically significant correlation between soiling and satisfaction. Ekkehart et al (96)

noted high contentment when there was no soiling (80%) however the parents in grade 2 soiling also showed a high rate of contentment.

Financial burden

Ekkehart et al (96) reported that financial burden on the family with ARM when compared with other chronic conditions were similar or worse in 54% of parents. However in our group the financial burden was very significant. 40.47% of parents went into debt with the expense of the treatment. 28% had to sell off their land, house, cattle or other valuables. The staged repair of ARM with the hospitalization for each has significant financial burden on the family

Schooling

Studies have reported normal intellectual ability in patients with anorectal malformation (100). Hassink (97) however reported that the overall education level was lower than that of the normal population. In our study out of 34 children of school going age 4 (12%) were not going to school. Parents did not want to send 2 of them to school for the fear of soiling. One child refused to go. 36.6% were studying in a lower class than that of their peers. Frequent school absence was seen in 23% of the children. School absence was reported in 50% by Hassink (97) and 18.3% by Bai et al (39).

Aggressive behavior

Bai et al (39) reported that 18% of children had behavioral problems like internalizing, depression, withdrawal, schizoid, and unsocial behavior according to Achenbach's Child behavior check list. The behavior problems were seen more in children with poor

continence. Ludman and Spitz et al (37) did not find incontinent adolescents more emotionally disturbed than those of good bowel control.

In our study 19/42 (45.23%) showed aggressive behavior. 12/19 (63.15%) children who showed aggressive behavior had major (grade 2 and 3) soiling.

Quality of life:

Fecal incontinence influences the quality of life adversely. Probably no aberration of the bodily function is as socially unacceptable as the inability to control one's stool. Quality of life was assessed according to the Quality-of-Life (QOL) Scoring criteria for children with fecal incontinence developed by Bai et al (39). This was assessed in children 5 years and above. 21/32 children had a 'good' score (scores 9-13), 9 had 'fair' (scores 5-8) and 2 (scores <4) had 'poor' scores. The average QOL was 9.1. The scores for children with poor continence was lower (7.8) compared to children with good continence (10.53). In a study by Bai (39) the QOL scores children with surgically corrected anorectal malformation was 9.4 and children with poor fecal incontinence had significantly lower score than those with good fecal continence. In children who were followed up after the bowel management program the QOL scores improved from an average of 8.1 to 12. The improvement in the score after BMP was statically significant. All the patients in the 'fair' and 'poor' group moved to the good group and children in the 'good' group improved on their QOL scores. This change was because the soiling had stopped after the bowel management program.

CONCLUSION

1. The main postoperative complication is chronic constipation , fecal overloading with overflow incontinence
2. Soiling was the main cause for parent's dissatisfaction of the treatment
3. The main thrust of the treatment is bowel management program aimed at eliminating fecal overloading.
4. There is significant improvement in the quality of life score after bowel management program
5. Parents go through lot of parental stress and financial burden if a child is born with anorectal malformation. A compassionate long term follow up and counseling is necessary
6. MRI findings are difficult to interpret but were useful in determining if the bowel was within the sphincter complex. And also whether there were sphincter deficiencies.
7. Anal manometry records a good anal pressure profile after sacroperineal pull-through.
8. The distal blind rectal pouch should be always preserved and not resected. The internal sphincter surrounds the fistula, so bowel around the fistula is preserved
9. The pouch should be mobilized as little as possible to preserve the innervation to the rectum.

10. The pulled through bowel should be sutured to the skin under mild tension to prevent prolapse and maintain a skin lined anus. Studies have shown ingrowth of sensory nerves from the perianal skin.
11. Children with ARM should be followed up into the childhood to deal with not only bowel problem, but also psychosocial issues. Their families require a lot of psychosocial support in order to raise their children.

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PROFORMA

A. PATIENT DETAILS AT FIRST ADMISSION

1. NAME
2. AGE
3. HOSPITAL NUMBER
4. ADDRESS AND CONTACT NUMBER
5. TYPE OF ANOMALY
 - a. INVERTOGRAM
 - b. DISTAL COLONOGRAM
 - c. OPERATIVE FINDING
6. COLOSTOMY
 - a. LEVEL
7. PROCEDURE
 - a. AGE
 - b. TYPE OF PROCEDURE
8. COMPLICATION
 - a. RETRACTION
 - b. STENOSIS
 - c. PROLAPSE
 - d. URINARY
9. SECONDARY ANAL PROCEDURE
10. AGE OF COLOSTOMY CLOSURE
11. ASSOCIATED ANOMALY
CARDIAC
VERTEBRAL
GIT
LIMB
RENAL
MISCELLANEOUS
12. SOCIOECONOMIC STATUS
 - a. AGE OF PARENTS AT THE TIME OF BIRTH OF THE CHILD
 - b. LEVEL OF EDUCATION
 - c. NUMBER OF CHILDREN
 - d. POSITION OF CHILDREN
 - e. OCCUPATION

B. DETAILS ON FOLLOW UP

1. DURATION FROM THE LAST SURGERY (COLOSTOMY CLOSURE)
2. HEIGHT in percentile

3. WEIGHT in percentile

1. SCORING (KRICKENBEG)

1. Voluntary bowel movements	Yes/no
Feeling of urge, capacity to verbalize, hold the bowel movement	
2. Soiling	Yes/no
Grade 1	Occasionally (once or twice per week)
Grade 2	Every day, no social problem
Grade 3	Constant, social problem
3. Constipation	Yes/no
Grade 1	Manageable by changes in diet
Grade 2	Requires laxative
Grade 3	Resistant to laxatives and diet

b. XRAY (TO FIND FECAL LOAD)

c. PER RECTAL EXAMINATION FOR LOADED COLON

d. TOILET TRAINED OR NOT

e. EPISODE OF ENTEROCOLITIS (DIARRHOEA, FEVER, ABDOMINAL DISTENSION, FALLING SICK)

6. PHYSICAL FINDINGS:

A. EXAMINATION OF THE PERINEUM

a. POSITION

b. PATENCY

c. PROLAPSE

d. FISTULA

- e. SHELVING
- B. MUSCLE STIMULATION
- C. SQUEEZE PRESSURE
- D. MRI

7. URINARY PROBLEMS

- A. DRIBBLING
- B. RETENTION
- C. RECURRENT UTI
- D. STRUCTURAL ABNORMALITY

8. PSYCOSOCIAL EVALUATION

A. PARENTAL STRESS

1. INITIAL REACTION
2. WERE THEY PROPERLY INFORMED
3. ANGRY WITH THE CHILD / RESENTFUL.
4. TAKING IT ON THE CHILD.
5. DO NOT SOCIALISE / INVITE OTHERS TO THEIR HOUSE.
6. SPOUSE PUT THE BLAMES ON THE OTHER.
7. WHO TAKE CARES OF THE CHILD / DO OTHER MEMBERS OF THE
FAMILY ESP. GRANDPARENTS INVOLVE IN THE CARE
8. DOES IT AFFECTS THE MARITAL LIFE

9. OTHER CHILDREN'S BEHAVIOUR

10. SECLUDED THE CHILD

11. STRAINED REALTION WITH THE INLAWS

12. WORRIED ABOUT THE FUTURE ESP. THE MARRIAGE.

13. SATISIEDWITH THE FINAL RESULTS

14. FINANCIAL IMPLICATION- DEBT/ LOAN/ TREATMENT CONCESION

B.CHILD ASSESSMENT

1. WHICH CLASS –SAME /BELOW FOR THE AGE

2. FREQUENT ABSENCE FROM THE SCHOOL

3. WITHDRAWN/ PLAYS WITH OTHER CHILDREN

4. AGGRESSIVE BEHAVIOUR

B. QUALITY OF LIFE (ABOVE 8 YEARS)

SOILING	ABSENT	4
	ACCIDENTAL	3
	FREQUENT	2
INCONTINENCE	ACCIDENTAL	1
	FREQUENT	0
SCHOOL ABSENCE	NEVER	2
	ACCIDENTAL	1

	FREQUENT	0
UNHAPPY OR ANXIOUS	NEVER	2
	ACCIDENTAL	1
	FERQUENT	0
FOOD RESTRICTION	NO	2
	SOMEWHAT	1
	MUCH	0
PEER REJECTION	NEVER	2
	ACCIDELNT	1
	FREQUEN	0

9. FOLLOW UP BOWEL MANAGEMENT

1. ENEMA- TYPE / FREQUENCY

2. DURATION OF FOLOW UP

3. RESULTS- FUNCTIONAL SCORE COMPARISON