

## Post-operative strictures in anorectal malformation: trends over 15 years

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### Abstract

**Aim** For decades, paediatric surgeons have employed the standard posterior sagittal anorectoplasty (PSARP) approach to deal with patients with anorectal malformations (ARM). In recent years, we noted an apparent increase in the incidence of anal stricture after surgical repair of ARM following the introduction of laparoscopic pull-through and techniques aiming to preserve the internal sphincter—the internal sphincter sparing approach (ISSA). We decided to analyse our data to find out if these new trends had added to the problem of post-operative strictures.

**Methods** All patients with ARM at our institution from January 2000 to December 2015 were identified. A retrospective case note review was carried out. Data collected included patient demographics, type of ARM, operative details, and post-operative outcomes.

**Results** 114 patients were identified. Ten patients were excluded. Of the remaining 104 children, 48 (46%) were female. Median age was 8.3 (range 1.2–16.8) years. Types of ARM were as follows: perineal fistula (15 patients), anterior stenotic anus (12), imperforate anus without fistula (10), vestibular fistula (32), rectourethral (bulbar) fistula (11), rectourethral (prostatic) fistula (14), rectovesical fistula (7), and cloaca (3). Twenty-seven patients with a perineal fistula or anterior stenotic anus underwent perineal procedures that were variably described by the different operating surgeons. The majority (15 patients) had an anoplasty, 5 had anal transposition, 5 had limited PSARP,

and 2 patients had ISSA. Two patients with a cloacal anomaly underwent open cloacal reconstruction. Of the remaining 75 patients, 45 had a PSARP approach, 6 had a laparoscopic-assisted pull-through, and 18 had ISSA. Four girls with vestibular fistula had anal transposition and two boys with imperforate anus without fistula had anoplasty. 15 (14%) children developed anal stricture. Stricture incidence differed according to operation type. PSARP was the most commonly performed procedure, with only 6% developing a stricture. In contrast, 30% of ISSA patients and 50% of children who had laparoscopic pull-through developed a stricture. Strictures also occurred in 11 and 12% of children having anal transposition and anoplasty, respectively.

**Conclusion** The laparoscopic-assisted pull-through involves tunnelling the sphincter muscle complex. We found that often the tunnels were not wide enough, resulting in narrowing not just at the ano-cutaneous junction but also at the deeper level. 50% developed strictures. We have modified our technique by ensuring that the tunnels are generous enough to allow the rectum to be pulled through without any resistance. ISSA unfortunately resulted in 30% of our patients developing strictures. This approach, started in 2004, was, therefore, abandoned in 2013. The standard Pena's PSARP, with or without a laparotomy, has stood the test of time. Any modification of this approach must be carefully thought through and audited meticulously. Strictures can cause significant morbidity, which may need several revisions, and the resulting redo anoplasties run the risk of sphincter damage, ironically which the newer modifications of ISSA were trying to conserve.

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## Introduction

Since its introduction by Pena in 1981, PSARP has become widely used by surgeons in the management of anorectal malformation (ARM). However, two new trends emerged in the last decade, both motivated partially with the unease over cutting the sphincter tissue widely in the midline, which is believed to disrupt the physiology of sphincter function. The new approaches are as follows:

- (a) The laparoscopic-assisted pull-through.
- (b) The internal sphincter sparing approach (ISSA).

This internal sphincter sparing approach in our centre comprised a perineal approach with preservation of the sphincter muscle complex, without dividing it in the midline (in contrast to PSARP). Instead, a tunnel was made through the complex and the rectum was brought through, with preservation of the distal tip of the fistula where the internal anal sphincter is thought to reside.

In recent years, we noted an apparent increase in the incidence of anal stricture following surgical repair of ARM. We decided to analyse our data to find out if the new trends had added to the problem of post-operative strictures.

## Methods

This was a retrospective review of all children with ARM managed at our institution from January 2000 to December 2015. Patients were identified by searching both an electronic database of neonatal unit admissions and written records of neonatal surgical admissions. A case note review was carried out. Data collected included patient demographics, type of ARM, operative details, and post-operative outcomes.

## Results

Data for 114 patients admitted at our institution between the study period were identified. Nine had a diagnosis of anterior anus and did not require any surgery. Another child had cloaca with a >3 cm common channel and was referred to a supraregional centre for reconstruction. These 10 patients were excluded. Of the remaining 104 children, 48 (46%) were female. Median age at analysis was 8.3 (range 1.2–16.8) years.

Types of ARM were as follows: perineal fistula (15 patients), anterior stenotic anus (12), imperforate anus without fistula (10), vestibular fistula (32), rectourethral (bulbar) fistula (11), rectourethral (prostatic) fistula (14), rectovesical fistula (7), and cloaca (3).

## Surgical procedures

A number of types of procedures were undertaken at our institution during the study period (Table 1). Twenty-seven patients with a perineal fistula or anterior stenotic anus underwent perineal procedures that were variably described by the different operating surgeons. The majority (15 patients) had an anoplasty. Five had anal transposition, five had limited PSARP, and two patients had ISSA.

Of the 77 patients with a high or intermediate type lesion, there were 2 patients with a cloacal anomaly who underwent open cloacal reconstruction. Of the remaining 75 patients, 45 had a PSARP approach (of whom 5 required an abdominal approach combined with PSARP for high anomaly). 6 had a laparoscopic-assisted pull-through. 18 had ISSA (with 1 requiring concomitant abdominal approach). Four girls with vestibular fistula had anal transposition and two boys with imperforate anus without fistula had anoplasty.

## Post-operative anal strictures

Fifteen (14%) children had medium-to-long-term morbidity following their initial surgery which was attributed to anal stricture (Table 2). In the group with low malformations, 15% (4 patients) developed strictures, and in the intermediate-to-high group, a similar proportion was affected (11 patients, 14%).

9/15 patients required multiple dilatations under general anaesthesia. Disturbingly, all except two required anoplasty; four required multiple anoplasties. Five developed megarectum necessitating resection in four cases; it has been discussed as a future management option in the fifth case.

The proportion of patients developing stricture differed according to operation type (Table 3). PSARP was the most commonly performed procedure, with only 6% developing a stricture. In contrast, 30% of ISSA and 50% of children who had laparoscopic pull-through developed a stricture. Strictures occurred in 11 and 12% of children having anal transposition and anoplasty, respectively.

## Discussion

The newer trends in surgery for ARM have forced this debate on us:

- Is there an internal anal sphincter in ARM?
- Is it normal?
- Is it worth preserving?

Although we acknowledge the limitations of using retrospective data, our 15-year series of the surgical management of ARM within a single centre is valuable in demonstrating the impact of modifications in operative

**Table 1** Procedure performed by type of anorectal malformation

	Anoplasty	Anal transposition	PSARP	ISSA	Laparoscopic pull-through	cloacal reconstruction
Anterior ectopic anus	4	5	2	1		
Perineal fistula	11		3	1		
Rectourethral fistula (b)			7	4		
Rectourethral fistula (p)			6	5	3	
Rectovesical fistula			6		1	
Vestibular fistula		4	20	8		
Cloaca (<3 cm cc)						2
Cloaca (>3 cm cc)					1	
Imperforate anus without fistula	2		6	1	1	

**Table 2** Patients developing stricture following surgery for ARM

Pt no.	Age (current)	Type anomaly	Procedure	Morbidity
1	5.0	Vestibular fistula	PSARP	7 dilatations under GA prior to colostomy closure. Redo procedure (abdomino-perineal) age 12 months. Subsequent further dilatations under GA, then a further anoplasty age 2 years
2	5.8	Vestibular fistula	ISSA	Multiple dilatations under GA. Has megarectum, decision on further management awaited
3	6.6	Perineal fistula	Anoplasty	Multiple dilatations. Anoplasty for stricture age 12 months. Excision of megarectum
4	7.0	Rectovesical fistula	Laparoscopic pull-through	Multiple dilatations pre colostomy closure. Anoplasty age 12 months for stricture
5	7.7	Rectourethral fistula (p)	Laparoscopic pull-through	Multiple dilatations. Anoplasty for stricture age 5 years, then further anoplasty 9 months later
6	7.9	Rectourethral fistula (b)	ISSA	Initial surgery complicated by concurrent SCT. Anal stricture evident D7 post-op. Multiple dilatations pre- and post-colostomy closure. Resection recurrent SCT × 2. Megarectum resection age 3 years with ileostomy. Ileostomy now closed, has ACE procedure.
7	8.4	Rectourethral fistula (p)	Laparoscopic pull-through	Dilatation pre colostomy closure. Anoplasty for stricture age 6 months, then stoma closure. Three dilatations post-anoplasty
8	8.8	Rectourethral fistula (p)	ISSA	Two anoplasties for stricture. Subsequent excision of megarectum and ileostomy. Stoma closure and ACE. Failed management, now has ileostomy
9	9.3	Imperforate anus without fistula	ISSA	Four dilatations pre colostomy closure. Two anoplasties for stricture
10	10.9	Rectourethral fistula (p)	PSARP	Anoplasty for stricture age 2 years
11	11.4	Perineal fistula	ISSA	Anoplasty for stricture age 3 years
12	12.3	Rectourethral fistula (b)	ISSA	Anoplasty for stricture age 7 months
13	15.4	Anterior ectopic anus	Anoplasty	Anoplasty for stricture age 2 years
14	16.5	Vestibular fistula	PSARP	Anoplasty for stricture age 4 years. Excision megarectum age 12 years
15	16.6	Anterior ectopic anus	Anal transposition	Multiple dilatations. Redo anoplasty, then colostomy. Two further anoplasties. Colostomy closed age 3 years

approach and, perhaps, provides a cautionary tale in respect of introducing new techniques without careful consideration and meticulous audit. Between 2000 and 2015, while the standard PSARP procedure was used at our centre, we also noted the emergence of two other trends, namely, the

laparoscopic pull-through and internal sphincter sparing approach.

The laparoscopic pull-through involves tunnelling the distal rectum through the sphincter muscle complex, with a small perineal incision [1]. Only six such procedures were

**Table 3** Post-operative strictures by procedure type

Procedure type	No patients	Strictures	% Strictures
PSARP	50	3	<b>6</b>
ISSA	20	6	<b>30</b>
Anoplasty	17	2	<b>12</b>
Anal transposition	9	1	<b>11</b>
Laparoscopic pull-through	6	3	<b>50</b>
Cloacal reconstruction	2	0	<b>0</b>
Total	104	15	14

Bold values indicate percentage of patients undergoing procedure who developed anal stricture

performed in our centre, but it is notable that 50% developed strictures. We observed that often the tunnels were not wide enough, resulting in narrowing not just at the anocutaneous junction but also at the deeper level. In addition, often the perineal incision was a tiny cut over the anal pit, just enough to accommodate the neo-anus. Laparoscopy is advantageous as a less invasive procedure when compared with those operations that require a laparotomy (e.g., where the rectum is located superior to the peritoneal reflection [2]), so we have chosen to continue to use it in selected cases. However, we have modified our technique by ensuring tunnels are generous enough to allow the rectum to be pulled through without any resistance. In addition, we ensure that the perineal cut is at least 3 cm long to better see the underlying muscle complex, as also to allow the rectal wall be sutured to it to prevent prolapse.

Interest in using an internal sphincter sparing approach arose from reservations that the posterior sagittal approach damages the sphincters by dissecting them widely in the midline. Histological and manometric evidence of an internal sphincter surrounding the anorectal fistula started appearing in the 80s [3–5]. The anal canal is normally kept closed by the anal sphincter muscles and pressure in the anal canal is higher than in the rectum and sigmoid colon. Closure is mainly generated by the internal anal sphincter [6].

Convinced by that data, surgeons in several institutions started preserving the fistula and incorporating it in the newly created anus, in the hope that this might improve faecal continence. This meant that the very tip of the rectum in cases of rectovestibular fistula or the rectourethral fistula was brought to the neo-anus some of these looked very narrow indeed.

Yet, more clinical evidence surrounding the existence of an internal sphincter in anorectal malformations emerged in the 90s, but the evidence was viewed with some caution. Meier-Ruge and Holschneider had established that histological specimens of the distal rectum of high-type anorectal and cloacal anomalies were characterised by anomalies of the muscularis propria and/or internal

sphincter, although this was not the case in lower anomalies [7]. Further analysis of the innervation of fistula and distal rectal pouch in a prospective study comprising 40 patients showed normal innervation in only 5% of patients, whereas 66% had neuronal intestinal malformations, including aganglionosis, NID, and hypoganglionosis [8, 9].

Rintala, however, followed 40 patients who had undergone an internal sphincter-saving posterior sagittal anorectoplasty and, histologically, ganglion cells were found in the proximal anal canal in all cases [10].

Later, publications showed that the subject was still surrounded by controversy. In one study, preoperative rectal manometry of rectoperineal or rectovestibular fistula showed the presence of functional anal structures within the fistula in all 12 patients [11]. Prokurat et al. also demonstrated good post-operative functional results after anterior sagittal anorectoplasty [12], and to date, Rintala's group in Helsinki continues the practice of preserving the fistula, with a reasonably low complication rate: out of a 159-patient cohort, 3% developed an anal stricture in the early post-operative period, and 4% developed severe constipation requiring further surgery in the late post-operative period [13, 14].

Meanwhile, Khan reported a 10% anal stenosis rate and a 28% chronic constipation rate after PSARP with fistula preservation [15]. Gangopadhyay's data supported the earlier findings of Holschneider and confirmed that the terminal end of the distal rectal pouch and the proximal fistula region possess distorted anal features with aganglionosis; these data brought into question the utility of incorporation of this region in anorectal reconstruction [16]. Another study concluded that actually the internal sphincter-saving procedure might not be essential for the development of the rectoanal relaxation reflex and that compensation or adaptation most likely contributes to the presence of the rectoanal relaxation reflex [17]. An experimental study performed on dogs confirmed that the IAS contributed to the anal resting tone but concluded that resection of the IAS did not completely interfere with faecal continence [18].

Our clinical data demonstrated quite a significant rate of anal stenosis in sphincter sparing operations—as high as 30%. This approach, started in 2004, was, therefore, abandoned in 2013. It seems probable that the combination of creating a tunnel through the sphincter complex in combination with preservation of the fistula resulted in a narrow anal opening from the outset, which became further compromised with the inevitable development of scar tissue post-operatively. Retrospectively, we may speculate that a vigorous anal dilatation regime and a very close post-operative follow-up might have helped to reduce the rate of stenosis. Interestingly, although anal dilatations are performed worldwide routinely after surgery, there are no established, standardised national or international protocols [19].

Strictures were not entirely confined to these two groups. We also saw it in 6% of those who had PSARP and just over 10% off those having procedures for ‘low’ anomalies. Although it was not possible to confirm from our data, a third trend was noted by us, which warrants discussion—the tendency to create a neo-anus which is not analogous in size to a ‘normal’ infant. Certainly, the calibre of the anal opening at the end of the ISSA operations was often reduced.

This begs the obvious question—what should the calibre of the neo-anus be, at different ages? The senior author (DM) advocates the following:

- (a) A suitable calibre for the neo-anus of a term neonate with a typical weight of 3.5 kg would equate to a size 11 Hegar dilator and it should be possible to pass the dilator without resistance. This applies to neonates with a ‘low’ anomaly who are having a primary PSARP or anoplasty at birth.

In smaller infants (e.g., premature) with a weight of less than 3 kg, a size 10 Hegar is recommended.

- (b) For an infant having PSARP at the age of 3–4 months, a size 12 Hegar is preferable. Post-operative dilations should take this up to size 13 or 14 before the colostomy is closed.

The morbidity resulting from anal stricture is not insignificant. Multiple dilatations under general anaesthesia (reflecting the severity of the narrowing) were required in nine patients, with little success as the majority of children with stricture (87%) subsequently had further anoplasty and 26% had more than one anoplasty. One of the most significant sequelae of tight stricture—megarectum—developed in five children necessitating resection in four cases; it has been discussed as a future management option in the fifth case.

## Conclusion

The standard Pena’s PSARP, with or without a laparotomy, has stood the test of time. Any modification of this approach must be carefully thought through and audited meticulously. Strictures can cause significant morbidity, which may need several revisions, and the resulting redoanoplasties run the risk of sphincter damage, ironically which the newer modifications were trying to conserve.

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