



## Perineal Fistula with High Dilated Colon: One of a Special Type of Anorectal Malformations

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### Abstract

**Background:** The perineal fistula with high dilated colon is a rare anomalous in the spectrum of anorectal malformations. The aim of this study is to explore the diagnosis and treatment of this Special type of Perineal Fistula (SPF), and to avoid the severe consequence due to misdiagnosis.

**Methods:** From March 2012 to January 2019, 7 patients who suffered from perineal fistula with high dilated colon were retrospectively reviewed. Three were operated on primarily by our department, and 4 cases were re-operated after a perineal anooplasty repair performed elsewhere. Four were operated by Laparoscopic Anorectoplasty (LARP), and 3 cases were repaired by Posterior Sagittal Anorectoplasty (PSARP). The follow-up outcomes were compared with 71 cases of Normal Perineal Fistula (NPF) in the same period.

**Results:** Seven cases have been followed up for 0.5 years to 4 years ( $M=2.57 \pm 1.26$ ) after definitive surgery. Their Bowel Function Score (BFS) was lower than normal perineal fistula (SPF=12, rang: 5 to 18; NPF=18.5, rang: 18 to 20). Four cases underwent anorectomanometry. The incidence of Rectoanal Inhibitory Reflex (RAIR) was lower in the special type group. ( $p=0.14$ ). Three cases of barium enema angiography: 2 cases of colorectal dilatation and thickening changes, 1 case showed no obvious abnormalities.

**Conclusion:** Anorectal perineal fistula should be examined by distal colostogram at pre-operation. Select appropriate surgical approach is essential for children's defecation function, and blind surgery can seriously affect long-term defecation function.

**Keywords:** Anorectal malformation; Perineal fistula; Distal colostogram; Laparoscopic anorectoplasty; Posterior sagittal anorectoplasty

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### Abbreviations

SPF: Special Type of Perineal Fistula; NPF: Normal Perineal Fistula; LAARP: Laparoscopic Anorectoplasty; PSARP: Posterior Sagittal Anorectoplasty; BFS: Fowel Function Score; RAIR: Rectoanal Inhibitory Reflex; PFO: Patent Foramen Ovale

### Background

Perineal fistula is one of the most common types of anorectal malformation in pediatric surgery. The diagnosis of perineal fistula is well recognized and most pediatric surgeons agree that it can be repaired by perineal anooplasty [1-3].

However, we found that there was part of perineal fistula associated with high dilated colon which was very rare. This special type of malformation (SPF) is characterized by perineal fistula as the main clinical manifestation but the tube is slender. Their proximal dilatation of the bowel is at a high level. Therefore, if surgeons don't perform a full preoperative assessment, this presentation may be delayed which may lead to abnormal bowel function.

As Peña's paper in 2017 emphasized, the distal colostogram is the single most important diagnostic study performed in children with imperforate anus prior to definitive repair [4]. The aim of this article is to explore the diagnosis and treatment of this special type of perineal fistula, and to improve its treatment outcomes.

### Methods

From March 2012 to January 2019, 7 patients who suffered from anorectal malformation with special type of perineal fistula were either admitted or transferred to Department of Pediatric

**Table 1:** Demographic data with SPF.

Case	Sex	Age at diagnosis	Age at operation	Weight (kg)	Associated anomalies		
					Cardiovascular	Spine	Genitourinary
1	Male	2d	6.2mo	8	---	---	---
2	Male	2d	5.7mo	7	PFO	Tethered cord	Hypospadias Isolated kidney
3	Male	2d	5.1mo	6	PFO	Tethered cord	---
4	Male	2d	4.8mo	6.2	PFO	---	Hypospadias
5	Female	10mo	12.6mo	11	---	Tethered cord	---
6	Male	2d	2.7mo	6.5	PFO	---	---
7	Male	2d	5.0mo	7.5	---	---	---

**Table 2:** Clinical features of patients with SPF.

Case	Surgical approach	Morbidity	Surgical times at anal	followed up (y)	BFS
1	LAARP	---	2	4	8
2	LAARP	---	2	4	10
3	LAARP	---	2	3	8
4	PSARP	Mucosal, prolapse	1	3	14
5	LAARP	---	1	2.5	18
6	PSARP	Partial bowel obstruction	1	1	18
7	PSARP	---	2	0.5	14

Surgery, Xinhua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine. There are 6 males and 1 female; the diagnosis time ranged from 2 days to 10 months after birth ( $M=8.57 \pm 107.70$ ). The age of the operation ranged from 2.7 months to 12.6 months ( $M=6.01 \pm 2.88$ ). Surgical weight ranged from 6.2 kg to 11 kg ( $M=7.46 \pm 1.59$ ). More than 50% of SPF patients had a demonstrable cardiac defect (Patent Foramen Ovale, PFO); 3 cases had spine syndrome and 2 cases had some degree of urological anomaly (Table 1).

Among these cases, four were diagnosed of low rectal perineal fistula, who had accepted perineal anoplasty without performed distal colostogram at a local hospital after birth. During surgery, it was found that it was difficult to find the blind end of the rectum and the operation was terminated. After the perineal was sutured, the colostomy was performed. Therefore, they only underwent colostomy, and were then transferred to our institute after three months. We did preoperative distal colostogram for them which revealed that the tube was slender, and the dilated proximal colon was located in front of the iliac crest.

One patient was diagnosed with rectal perineal fistula in the local hospital, and was treated with daily anal sphincter treatment. When she was 10 months old, she developed anal bleeding after anal sphincter, and then she was transferred to our hospital. The preoperative distal colostogram and MRI were diagnosed as NPF, so we have first performed transverse colostomy for her.

The 2 others, who came to our hospital after birth, were diagnosed as SPF by the preoperative distal colostogram as in the above situation. First of all, the colostomy was performed in our hospital.

All the 7 children underwent an anal angioplasty in our hospital. Four were operated by Laparoscopic Anorectoplasty (LAARP), and 3 cases were repaired by modified Posterior Sagittal Anorectoplasty (PSARP). In 7 cases, the proximal dilatation of the bowel was found to be at a high position in the operation. During the surgery, we found

that 4 cases of children who were re-operated anal had serious scars in the original perineal surgery site and surrounding tissues. Electrical stimulation was difficult to locate the external sphincter center point, and we finally completed the re-construction surgery combine with anatomical positioning (Table 2).

Clinical evaluation was carried out using the seven intestinal function scores (BFS) presented by Rintala [5]. Refer to the previous scale study, the maximum total score is 20, the reference value of normal children is 19, of which 18 is the tenth combined [6,7]. Therefore, in this study, the value of 18 (over 90% of the control group) was used as the lower limit of the normal value. In addition, the manometric evaluation was performed simultaneously with the high-resolution anorectal manometry. The Parameters measured in this study are the sphincter resting pressure, squeeze pressure and the presence of Rectal Inhibitory Reflex (RAIR). The reference value of normal sphincter resting pressure is 30 mmHg to 60 mmHg, and the extrusion pressure is 50 mmHg to 120 mmHg [8]. The follow-up outcomes were compared with 71 cases of Normal Perineal Fistula (NPF) in the same period.

A standard statistical software package (Windows version 23.0; SPSS Inc., Ar-monk [NY], US) was used to analyze the data. Continuous variables are expressed as medians (ranges) and compared using Mann-Whitney U test. Use the chi-square test to compare categorical variables. P value less than or equal to 0.05 is considered statistically significant.

**Results**

Finally anorectoplasty was performed successfully for all 7 cases. All 7 cases have been followed up for 0.5 to 4.0 years ( $M=2.57 \pm 1.26$ ) after definitive surgery. Six patients underwent regular outpatient follow-up and one patient was followed up by telephone. Among all cases, one had partial bowel obstruction after operations and was cured by supportive treatment. One case was complicated with

**Table 3:** Comparison of clinical and manometric results of patients between SPF vs. NPF.

	SPF (n=7)	NPF (n=71)	p value
Functional assessment			
- Constipation	14.30%	7.00%	0.44
- Soiling ( $\geq 1$ time per week)	42.80%	7.00%	<0.05
- Median BFS	12.4 (8-18)	18 (16-28)	0.82
- % of patients with normal BFS	42.90%	86%	0.17
Manometric assessment			
-Resting pressure at sphincter (mmHg)	23 (21-26)	34 (13-65)	0.1
- % of patients with normal sphincteric resting pressure	0%	72%	<0.05
-Sphincteric squeeze pressure (mmHg)	50 (29-68)	56.3 (13-115)	0.59
- % of patients with + RAIR	0%	50.70%	<0.05

mucosal prolapse and did the surgical resection. During the Follow-up, all 7 cases did the seven-itemed Bowel Function Score (BFS) and high-resolution anorectomanometry. Three cases did the barium enema angiography.

There was no significant difference in the bowel function score between the different surgical approaches. Three cases who only had one operation have no obvious symptoms such as incontinence and constipation up to now. But 4 cases that were experienced more than 2 repair operations had lower score in bowel function. One of them have constipation and the remaining 2 have fecal incontinence. And their BFS were only 8, 10, 8, and 14, which far below the normality (Table 2).

For functional evaluation, the rate of constipation (as defined by the Roman III standard) is 14.3% (n=1) and 7.0% (n=5) ( $p = 0.44$ ) in the SPF and NPF groups respectively but this was not statistically significant. The rate of soiling (more than once per week) of SPF (n=3, 42.8%) was higher than that of NPF (n=5, 7.0%) ( $p < 0.05$ ). The median of BFS in the SPF group was significantly lower (SPF=12.4, range: 8 to 18; NPF=18, range: 16 to 20,  $p=0.82$ ) and there were fewer patients with normal BFS in the SPF group (SPF vs. NPF=42.9% vs. 86%,  $p=0.17$ ) (Table 3).

The median sphincter resting pressure in the SPF group was 23 mmHg (range: 21 mmHg to 26 mmHg) in the SPF group and in the NPF group it was 34 mmHg (range: 13 mmHg to 65 mmHg) ( $p=0.10$ ). Compared with the age-matched control group, higher patients in the NPF group had normal sphincter resting pressure (SPF vs. NPF=0% and 72%,  $p < 0.05$ ). There was no significant difference between the two groups in the measurement of sphincter compression pressure (SPF= 50 mmHg, range: 29 mmHg to 68 mmHg; NPF=56.3 mmHg, range: 13 mmHg to 115 mmHg,  $p=0.59$ ). However, RAIR was present in 0% (n=0) and 50.7% (n=36) of patients in the SPF and NPF groups ( $p < 0.05$ ) (Table 3).

## Discussion

The definition of the perineal fistula is undisputed. And the perineal fistula is one of the most common anorectal malformations in pediatric surgery [9]. But the Special type of Perineal Fistula (SPF) with high dilated colon is very rare and the number of cases in the literature is limited. So far, no such case has been reported. In our cases, the dilated proximal colon was located in front of the iliac crest which undoubtedly leads to the complexity of the operation that can be defined as SPF.

In this study, incidence of the special type of perineal fistula with high dilated colon was 8.97% (7/78). This incidence may reflect that surgeons often ignore to raise the importance of SPF. About 42.8% of all patients with SPF had spine syndrome of tethered cord which is far higher than NPF.

The diagnosis of perineal fistula is well recognized, and almost neonates can be diagnosed after birth. A true perineal fistula is a narrow perineal foramen with no visible anal canal, located in the center of the anal sphincter, and not completely surrounded by the sphincter mechanism [9-11]. The boy's fistula is often in the normal anal area and the root of the scrotum. The girl's fistula is mostly between the normal anal area and the posterior labia. The clinical presentation of SPF is similar to perineal fistula, but the position of dilated colon is unlikely to this normal perineal fistula. Based on the above findings, some surgeon usually ignored preoperative examination to assess the correct understanding of the anatomical structure of these children's anorectal malformations, including the related fistula communication between the rectum and genitourinary tract. In our neonatal unit, all newborns with anorectal malformation of perineal fistula were screened by distal colostogram to confirm the position of the distal rectum. And we found that there was partial special type of perineal fistula which was associated with high dilated colon. This special type of perineal fistula, if unrecognized preoperatively, may complicate surgical operations to repair anorectal deformities. Therefore, it is imperative that a thorough evaluation (e.g. distal colostogram) be performed to identify SPF so that they can be correctly method for treatment. There is a consensus in the literature on the best imaging method for accurate anatomical diagnosis at birth. Some authors believe that perineal ultrasound is the best option, while others prefer MRI [12,13]. We believe that no matter which evaluations is used to assess accurate understanding of the anatomy of the anorectal malformation, preoperative evaluation is essential for these children who were diagnosed perineal fistula to identify the possibility of a high dilated colon. We, therefore, suggest distal colostogram in the newborn period as well as pelvic MRI to thoroughly evaluate for the anatomy of the anorectal malformation.

Another important decision before surgery is to determine whether to transfer to a center with rich experience in pediatric surgery. Although there is no consensus on this point, there is strong evidence from adult studies that the surgeon's physique is independently associated with better outcomes [14-16]. For some local hospitals, surgeons may lack awareness of the anorectal malformation. Therefore, we suggest that if there is no pediatric

surgeon in local hospital, patients should be transfer to a centre with extensive pediatric surgery experience.

In most patients born with SPF, management includes a diverting colostomy. Colostomy is valuable because it can avoid the complications of mega-colon and pseudo-incontinence [17]. In our series, all patients were managed in diverting colostomy before the repair.

Although the most straightforward of the anorectal malformation groups to repair [18,19], the perineal fistula is still a consistent point of confusion. It is uniformly known that the best effect of radical surgery is to select appropriate surgical approach which is essential for children's defecation function, and blind surgery can seriously affect long-term defecation function. In this study, we introduced the experience of correcting this abnormality using the posterior sagittal approach and laparoscopic approach which can accurately expose the position of the distal rectum. Two approaches can avoid possible complications of fistula, urethral injury or intrasphincter anal dislocation.

Our routine is to calibrate the anus 3 weeks after surgery, and continue to expand for several months every day. The postoperative outcome regarding continence after ARM treatment is reported to be 100% of the patients had bowel control in recto-perineal fistulas and the postoperative pain is also minimal [3]. In our review 86% of the NPF patients followed up were continent after correction at the end of the follow-up period. However, 57.1% of these SPF patients had abnormal bowel function, which include constipation and fecal incontinence. An inadequate surgery, coupled with a natural propensity for constipation, can lead to severe faecal impactions that can cause substantial rectal dilation. This dilation can hamper rectal mobility and ultimately affect faecal control. Especially when surgeon did not make adequate pre-operative evaluation to accurate understanding of the anatomy of the anorectal malformation including an associated fistulous communication between the rectum and the urogenital tract, it would affect the optimal surgical management. Based on that, patients may suffer multiple operations, and may cause defecation dysfunction. In our study, children who were experienced more than 2 repair operations at anal had lower score in bowel function than those suffered only one operation. Besides, the measurements of bowel function score in SPF patients were lower than NPF patients though there is no significant difference. Therefore, we recommend that repeated operations and high dilated colon may be one of the risk factors for abnormal function of defecation in these patients. Although most of these patients can be managed with drugs, careful follow-up is vital (as with all patients who have anorectal malformations) so that patients who need early management of their bowel function can be identified [20].

## Conclusion

In conclusion, this study reported the special type of perineal fistula. Compared with NPF patients, the measurements of bowel function score and sphincter resting pressure in anorectal manometry were lower in SPF patients. In order to avoid misdiagnosis, anorectal perineal fistula should be examined by distal colostogram before repair. Select appropriate surgical approach is essential for children's defecation function, and blind surgery can seriously affect long-term defecation function.

## Declarations

Ethics approval and consent to participate the study was

approved by the ethical committee of the Institutional Review Board of Shanghai Jiao Tong University. Informed written consent was obtained from the parents of each participant. Study procedure was carried out in accordance with the approved guidelines.

Consent for publication written informed consent for publication was obtained from all participants. All parents of the study participants gave written consent for their clinical details along with identifying images to be published in this study.

Availability of data and materials the data used to support the findings of this study are included within the article.

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