Theoretical rationale for new surgical tactics for anorectal malformations without visible fistulas

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Abstract

Recently, the diagnosis and surgical treatment of anorectal malformations (ARM) is based on the idea of the absence of the anal canal in these patients. The caudal part of the intestine, called the fistula or rectal pouch, is removed during all pull-through procedure. Poor results from these surgeries (fecal incontinence 16.7% to 76.7%; chronic constipation 22.2% to 86.7%; urinary incontinence 1.7% to 30.5%; ejaculatory dysfunction 15.6% to 41.2%; and erectile dysfunction 5.6% to 11.8%) are explained by the congenital absence of the anal canal. In the previous articles, we proved that these patients have a normally functioning anal canal. This was confirmed by the European Consortium. This paper provides a theoretical justification for new surgical interventions that preserve all the elements of the anal canal to exclude severe complications during ARM correction. In newborns, we offer a perineal perforation (PPP) procedure by inserting a needle into an open anal canal. The opening of the anal canal is performed under X-ray control due to compression of the abdomen. As a result of simple manipulations, a tracheostomy tube is inserted into the rectum, which is removed after 7 days. Diastasis between the intestine and the skin heals without the formation of fibrous tissue since there are no sutures between the intestine and the skin. In the presence of a colostomy, we recommend the method proposed by Pakarinen and Rintala. The anal canal is visualized within the intestinal lumen using retrograde flexible endoscopy through a previously performed sigmoid mucosal fistula. Perforation of the skin and subcutaneous tissue is performed with a special endoscope needle. After the same manipulations as in PPP, a tracheostomy tube is inserted into the rectum. Fecal retention and defecation did not differ from the norm after the use of PPP in several patients. This work substantiates the opportunity for clinical trials of the proposed methods.

Keywords: anorectal malformations without visible fistula; anal canal; X-ray diagnosis; surgical treatment; perineal perforation procedure; colostomy.

I. Introduction. Stephens (1953) proposed the concept of the pubococcygeus (P-C) line, extending from the inferior part of the pubis to the distal part of the coccygeal vertebras. He showed that this line corresponds to the location of the puborectalis muscle (PRM), which separates the rectum and anal canal. In accordance with this concept, anorectal malformations (ARMs) were divided into high if the distal part of the intestine was located cranially from this line, intermediate - at the level of the P-C line, and low if they were located caudal to this line. Stephens showed the need to preserve PRM, which plays an important role in fecal retention [1]. Since then, it was believed that if the colon is located below the P-C line, this means that the patient has an anal canal, which must be preserved during surgery. In practice, ARMs were divided into high and low types, i.e. without anal canal and with anal canal, respectively. However, the diagnosis of ARM was not accurate because it was based on two misconceptions. (1) Radiological studies (invertogram or lateral radiograph with the pelvis elevated) assumed that gas in the rectum moves upward. In fact, the contents in the digestive tract move only by peristaltic waves. (2) The fact that the anal canal at rest, i.e., at low rectal pressure, is in a closed state and opens only at a certain (threshold) pressure in the rectum was not considered [2]. In such cases, ARMs were mistakenly thought to be high when in fact they were low.

From 1982 until recently, it was believed that a portion of the bowel, which existed caudal to the P-C line, is pathological changed and it is called a fistula or rectal pouch. Although there has been no scientific evidence to support this claim, this so-called fistula is removed during posterior sagittal anorectoplasty, anterior sagittal anorectoplasty and laparoscopy-assisted pull-through surgery [3,4,5]. Long-term treatment outcomes were shown in a systematic review by Rigeros Springford et al. Long-term active problems were as follows: fecal incontinence, 16.7% to 76.7%; chronic constipation – from 22.2% to 86.7%; urinary incontinence - from 1.7% to 30.5%; ejaculatory dysfunction – from 15.6% to 41.2%; and erectile dysfunction - from 5.6% to 11.8%. [6]. Even though the results of pull-through surgery were significantly worse than operations that preserve the anal canal (cutback procedure) [7], many authors continue to argue that better results cannot be achieved, since the patient with ARM does not have an anal canal. Based on a review of the literature and our own research, we have proven that most patients with ARM without a visible fistula have a normally functioning anal canal [8]. Members of the European Consortium agreed that “According to current knowledge, the “fistula” of ARM is an ectopic anal canal and should be preserved as far as possible to increase the likelihood of fecal retention” [9]. Based on the modern understanding of the pathophysiology of ARM, based on an analysis of the literature and our own research, we

propose a theoretical justification for a new surgical tactic for ARMs without the visible fistulas.

Diagnostic tests and surgical methods depend on the age of the patient. Therefore, we are considering two options: (1) in newborns, and (2) in the long-term period in the presence of a colostomy.

II. Diagnosis and surgical treatment of ARM without functioning fistulas in newborns.

A) X-ray examination

(a). Rationale of the method. In the first hours of life, both in a healthy child and in a newborn with ARM, rectal pressure is below the threshold level. Therefore, the anal canal is in a closed state, and meconium with gas is in the rectum. It is not until 30 hours after birth that enough gas and meconium have accumulated in the rectum to create the pressure at which the anal canal opens. However, in the initial stage the opening of the anal canal lasts several seconds, after which the rectum adapts to the rectal volume, the pressure in it decreases below the threshold level and the anal canal contracts, squeezing the gas that has entered it into the rectum [2]. To see the opening of the anal canal and prevent its closure, we create additional pressure in the rectum [8,10].

(b). X-ray method. The child is on the X-ray table in a horizontal position, on his side. During fluoroscopy, the doctor squeezes the child's abdomen between his hands from the back and abdomen. Evidence of normal anal canal function is the penetration of gas from the rectum to the anal canal and its approach to the radiopaque marker glued to the anal dimple **(Figure 1).**

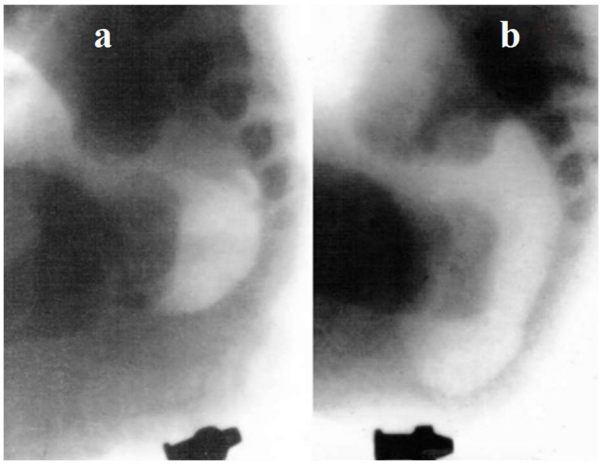

Figure 1. Radiographs of a newborn with ARM without a visible fistula were taken in a horizontal position. An X-ray contrast marker is glued to the anal dimple. **(a).** At rest, the

distance between the rectum and the marker is less than the normal length of the anal canal because the photograph was taken at the beginning of the opening of the anal canal. **(b).** When the abdominal cavity is compressed, the gas approaches the marker. The distance between the marker and the intestine is equal to the thickness of the skin and subcutaneous tissue.

Compression of the abdomen between the doctor's two palms from the back and abdomen provokes a more stable opening of the anal canal. In this case, the following situations are theoretically possible:

1) If, when the abdomen was compressed, the gas approached the perineum, but immediately disappeared, and the true width of the rectum decreased, then there is a previously undiagnosed fistula.

It should be noted that over a long period we did not have a single similar case in boys with urethral fistula. This indicates that in most newborns, anourethral fistulas do not function under physiological conditions. Secondly, in almost all cases when, based on an X-ray examination with abdominal compression, a low type of ARM was diagnosed, during the pull-through operation, surgeons found the rectum at about 2 cm from the anal dimple, and considered these cases as a high type of ARM [10]. This phenomenon is explained by the fact that the pressure in the rectum decreases under anesthesia, which leads to a contraction of the anal canal, which is almost impossible for the operator to enter.

2) If, during compression, the gas approached the perineum and remained there until the compression of the abdomen continued, and the width of the rectum did not change, then there is an anal canal without a functioning fistula (**see Figure 1**).

3) If, despite abdominal compression, the gas does not reach the perineum and the true rectal width is < 2 cm, then there is too little content in the rectum to cause defecation, and the test should be repeated after 4 hours.

4) If the gas does not approach the perineum, but the rectum is wide, then the child has a high type of ARM. We do not have reliable data to rule out the possibility of a high-type ARM. However, in our practice, we have not diagnosed a single such case using abdominal compression.

B). Surgical treatment in newborns without a functioning fistula.

For neonates with a low type of ARM, we recommend a perineal perforation procedure (PPP). This method preserves all elements of the anal canal and does not damage their blood supply and innervation.

(a). Perineal perforation procedure in newborns without visible and non-functioning fistulas.

This operation is recommended: (1) If, despite careful attempts, a fistula in the perineum is not detected within 30 hours of life; (2) If no traces of meconium are found in the urine; (3) If, during X-ray examination, during abdominal compression, gas penetrated the perineum, approached the marker in the anal fossa, and did not disappear from the anal canal during compression. (4) This also applies to cases where the ectopic anus is attached to the urethra, since the above conditions indicate that this fistula does not function under physiological conditions. In other cases, i.e., if a functioning fistula is suspected, a colostomy should be performed. The PPP diagram is presented in **Figure 2.**

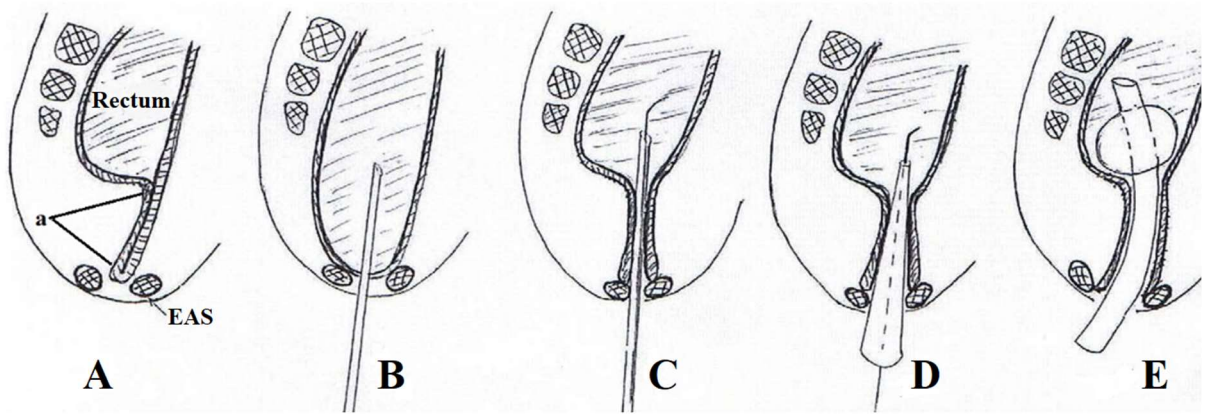

Figure 2. The PPP scheme. Side view. **(A)** The anal canal (a) is closed. The "EAS» is the subcutaneous part of the external anal sphincter. **(B).** The anal canal is open during compression of the abdominal wall. Under X-ray control, a needle is inserted into the anal canal. **(C).** A guidewire for vascular catheterization is inserted into the rectum through a needle. **(D).** A conical dilator is inserted by a guidewire to widen the newly created channel in the skin and subcutaneous tissue. **(E).** After removing the dilator, a tracheostomy tube is inserted through the guidewire. The balloon of the tube is inflated in the rectum to fix the tube in the anal canal.

On the X-ray table, under general anesthesia, a cross-shaped incision is performed in the skin above the subcutaneous part of the external anal sphincter. After stretching fibers of the sphincter, the child is transferred to the lateral position. During abdominal compression, and the anal canal opening, the needle is inserted from the skin incision into the rectum through the

open anal canal (Figure 2, A-B). Only this step is performed under fluoroscopic guidance. The sound of gas escaping is indicated that the needle is inside the gut. Then a conductor with a soft floating end is inserted through the needle into the rectum (Figure 2, C). The needle is removed and a dense conical bougie with a maximum diameter of about 0.8 cm is inserted into the intestine along the guidewire (Figure 2, D). After this, a tracheostomy tube with a diameter of about 0.8 cm is inserted into the rectum and the guide is removed (Figure 2, E). The 5 cm^3 of air is blown into the balloon of this tube, which makes it possible to fix the tube for 7 days. The internal anal sphincter is not mobilized or sutured to the skin. After 7 days the tube is removed. After 2 weeks, the wound heals without scars and the mother begins to dilate the anus by inserting the little finger.

In one case, no gas was heard escaping. A contrast agent was injected to make sure that the needle was in the rectum **(Figure 3).**

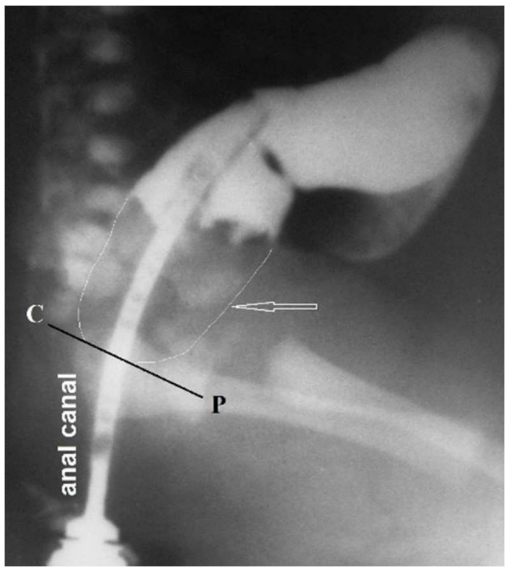

Figure 3. Newborn with ARM without visible fistula. After insertion of the needle, no gas was heard escaping. Therefore, doubt arose that the needle was in the intestinal lumen. A tube was passed through the guidewire, through which a contrast agent was injected. It turned out that there was a large amount of meconium in the rectum (arrow), which blocked the lumen of the needle.

The PPP was performed in 4 newborns with ARM without visible fistulas. In one of four children, ARM was combined with esophageal atresia. He died after correction of the esophagus from aspiration pneumonia. In three patients the postoperative period was uneventful. They were called for examination at the ages of 1.5, 1.9 and 2.2 years, and in each

of them the functions of fecal continence and defecation did not differ from the norm. We recommend this method, despite the small number of children operated on, because the described method is scientifically proven. Secondly, because the results of treatment are strikingly different from all described methods of the pull-through surgery.

(b). Why can't the internal anal sphincter (IAS) be sutured to the skin of the perineum?

After PPP, a small diastasis remains between the wall of the IAS and the skin. The generally accepted method of correction is suturing the intestinal wall to the skin. Analysis of the literature and the results of the observations described above prove the fallacy of this tactic.

First, to suture these tissues without tension, it is necessary to isolate the IAS from the surrounding tissue by more than 2 cm, essentially the entire depth of the newborn's anal canal. As a result, the blood supply and innervation of the IAS are disrupted, which leads to degeneration of the sphincter muscle fibers and loses neural connections with other participants in the anorectal reflexes. As shown by Rintala et al., "preservation of the internal sphincter in patients with high or intermediate anorectal anomalies provides good fecal continence but is associated with a high incidence of symptomatic constipation" [7]. However, one should distinguish between the "use" of the IAS, in which its function is disrupted during "separation" from surrounding tissues, and the "preservation" of the IAS, when it is not released from the surrounding tissues. In the latter case, its vascularization and the anorectal reflexes are completely preserved.

Secondly, after the IAS is disconnected from the vagina in girls and the urethra in boys, fibrous tissue, and scars in the suture area form around the IAS.

Thirdly, as X-ray examination shows, the distal wall of the anal canal always reaches the subcutaneous tissue in low types of ARMs, and the distance from it to the skin in the anal fossa in newborns is no more than 2 mm [10].

Fourthly, in the embryonic period, the anal canal is formed due to the penetration of the IAS into the perineal tissue. The wall of the IAS retains the ability to recanalize in the postnatal period. Our observations confirmed that after PPP, the 2 mm diastasis between the wall of the anal canal and the perineal skin is filled with progressive neoplasms of the IAS within a week, usually without an inflammatory reaction and without the formation of fibrous tissue and stenosis.

III. Diagnosis and treatment of ARM without visible fistula in the presence of colostomy

A) Rationale. If a newborn without a visible fistula shows signs of a functioning fistula in the urethra or vagina, it is customary to perform a colostomy. It unloads the intestines, prevents the development of megacolon, and protects the genitourinary system from infection. It is advisable to make a radical correction when the child grows up and gets stronger. The examination begins with establishing the presence or absence of the anal canal, since this information radically changes treatment tactics. Performing a distal colostogram with elevated pressure, firstly, does not make sense, since the presence or absence of a functioning fistula does not change the treatment tactics. Secondly, this study carries a high risk of intestinal rupture. Therefore, most researchers try not to create high pressure, but in this case, they do not achieve the desired effect. Thirdly, by creating very high pressure, doctors stretch the non-functioning fistula, turning it into a functioning one. Therefore, this study does not have a functional basis.

In pediatric surgery, there is an axiom that it is better to perform surgery not in the neonatal period, but at an older age. However, pediatric surgeons performing pull-through surgery prefer to perform the final correction in the neonatal period, since the functional results do not depend on the age of the operated patients. The reason for these results is that complete destruction of the anal canal equalizes the functional results in patients operated on at different ages.

B) The diagnostic method proposed by Pakarinen and Rintala is completely safe and can also be used for surgical treatment. The anal canal is visualized within the intestinal lumen using retrograde flexible endoscopy through a previously performed sigmoid mucosal fistula. The distal end of the anal canal is clearly defined by the convergence of the anal columns. Bright transmission of endoscope light from the anal canal to the anal fossa within the external anal sphincter indicates a low type of ARM (**Figure 4**) [11].

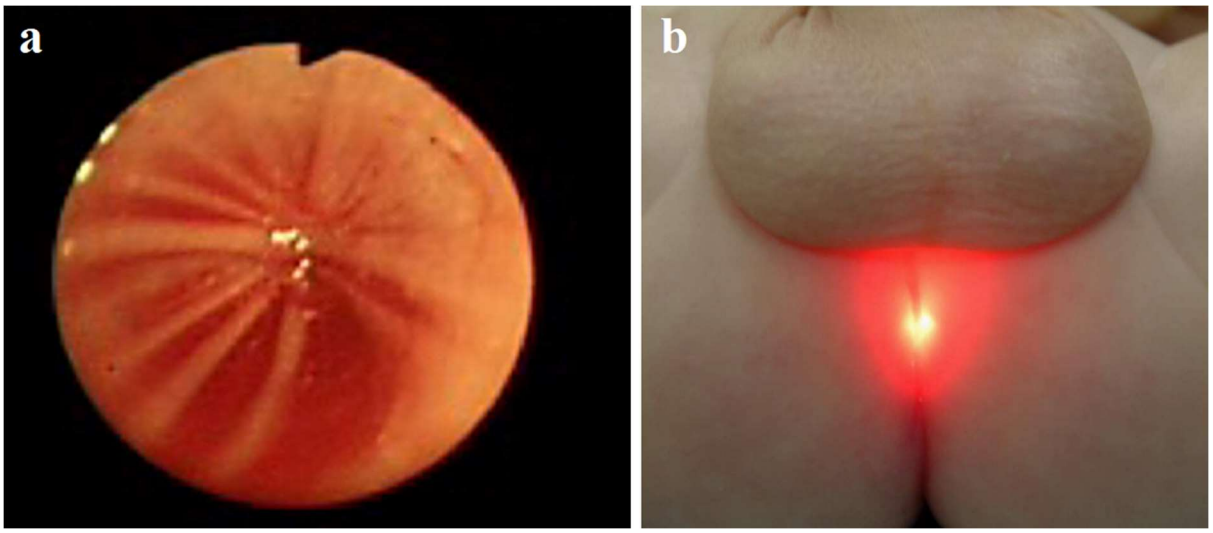

Figure 4. Examination of the terminal bowel using retrograde flexible endoscopy through a sigmoid fistula from the article by Pakarinen and Rintala [11]. **(a)** Endoscopic image of a blind anal canal in a patient with ARM without fistula. Note the convergence of the anal columns. **(b)** Translumination of endoscope light in a patient with ARM without a fistula. The light is visible so brightly because it passes from the lower part of the anal canal only through the blindly ending distended wall of the IAS, subcutaneous tissue, and skin. Conclusion: there is low type of ARM.

C) Treatment of ARM without visible fistula in the presence of colostomy.

Pakarinen and Rintala described "transanal endoscopic proctoplasty, which requires significantly less extensive anorectal dissection without the risk of anatomical reconstruction" [12]. However, since any dissection leads to circulatory failure and tissue denervation, we suggest using this method without tissue dissection.

(a). Rationale for a new method of surgical treatment of ARM in the presence of colostomy.

Rationale for a new method of surgical treatment of ARM in the presence of colostomy.

The location of the subcutaneous portion of the external anal sphincter (EAS) is determined using electrical stimulation. The skin is cut and the ring of the subcutaneous part of the NAS is stretched. The anal canal is visualized in the intestinal lumen using an endoscope inserted retrogradely through the previously created sigmoid fistula. The distal end of the anal canal is clearly defined by the convergence of the anal columns. A bright endoscope light visible from the outside indicates a low type of the ARM (**see Figure 4**). In these cases, the endoscope needle is passed through the middle of the convergence of the anal columns and through the

EAS ring. The guidewire clings to the needle and is pulled into the rectum. The anus is dilated using a conical dilator along a guidewire, after which a tracheostomy tube with a diameter of 1 cm is inserted into the rectum. A balloon inflated in the rectum fixes the tube in the anal canal for 7–10 days. During this time, the diastasis between the wall of the IAS and the skin, several millimeters long, is closed due to hyperplasia of the wall of the anal canal. Healing occurs without inflammation and without the formation of scar tissue. If during observation an ectopic opening is found between the anal canal and the urethra, it can be sutured through the anal canal (**Figure 5).**

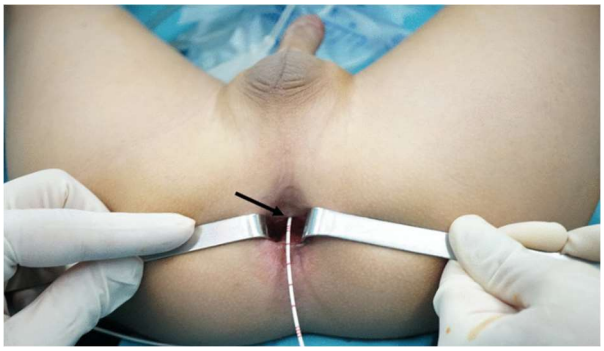

Figure 5 and caption from the article by Huang et al. "Examination under anaesthesia: the appearance of the anus and sphincter function were good. The fistula (arrow) was located 1 cm above the anus verge without obvious scar" [12].

Figure 5 shows an opening between the urethra and the anal canal, which can easily be sutured or sealed in some other way. Treatment ends with closure of the colostomy. This tactic cannot damage the genitourinary system and preserves all elements of the anal canal, so such children can be completely healthy.

Conclusion. This review of the literature and our own research is a theoretical basis for new methods of diagnosis and surgical treatment of patients with ARM without a visible fistula. They are a scientifically proven alternative to pull-through surgery, regardless of the approach (posterior, anterior or with endoscopy), that irreversibly destroys the anal canal. Of course, It is necessary to evaluate their effectiveness in clinical practice. However, the choice of every pediatric surgeon is very simple: destroy or preserve the anal canal.

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