Discussion about methods of X-ray diagnostics of anorectal malformations in children. Review.

Introduction.

Radiological diagnosis depends on understanding the normal anatomy and physiology of the anorectal zone, as well as on their changes in anorectal malformations (ARM). It is obvious that, working in tandem, a radiologist and a pediatric surgeon should have the same views on these problems. In the historical aspect, 2 views on the pathological anatomy and physiology of ARM are announced, on which various surgical methods of treatment depend. The goal of surgical treatment is to provide the patient with the best functional results. Radiological examination helps to choose the most rational method of surgical treatment [1].

The purpose of this review is to compare scientific evidence and treatment outcomes for these two trends in pediatric colorectal surgery. Therefore, I consider separately the scientific evidence of those scientists who believe that at low ARM there is an anal canal that needs to be preserved (Group 1). The second group of evidence comes from who believe that the distal intestine is a fistula or rectal pouch, the function of which does not provide normal retention of feces and defecation. Therefore, it must be removed (Group 2).

I. Evidence of the presence of the anal canal in low types of ARM (Group 1)

Stephens proposed the concept of a pubococcygeal (P-C) line, which runs from the lower part of the pubic bone to the distal coccygeal vertebra. He showed that this line corresponds to the location of the PRM, which is located between the rectum and the anal canal and plays a significant role in stool retention. If the blind end of the intestine is located above this line, these cases are considered a high type, and if below the P-C line these cases are a low type [2]. This understanding of the pathological physiology of APM was reflected in the classification of Wingspread (1984). Since then, it was believed that if the gut is located caudally of the P-C line, it means the patient has an anal canal that needs to be preserved during surgery [3,4,5,6,7]. At that time, the low type included anal stenosis and ARM with fistulas on the perineum and vestibule.

A) Histological studies. Alamovich et al. (citation from Duhamel) investigated the innervation of the normal internal anal sphincter (IAS). This study shows that the IAS has no autonomous innervation unlike the rest of the digestive tube [8]. Lambrecht and Lierse in 3 neonatal pigs with ARM found that the proximal region of the fistulae in ARM has most features of a normal anal opening. They consider that the fistula should be designated as an ectopic anal canal [9]. The most important result was the demonstration of a normal functioning internal anal sphincter (IAS) even in high and intermediate types of

ARM [10]. Rintala et al have shown that in anorectal malformations the distal rectal pouch with the fistulous connection is anal canal ectopy [11]. The study by Uemura et al allowed them to conclude: "Epithelial and ganglionic distribution was similar in the distal rectal end of ARMs and in a normal anal canal. The anal transitional zone is the epithelial boundary between the rectum and skin in a normal anal canal. Anal transitional zone preservation could reproduce anal canal structure in ARM reconstruction" [12].

B). Manometric studies. In 1877, Gowers discovered a decrease in pressure in the anal canal after insufflation of air into the rectum [13]. This reflex was called the rectoanal inhibitory reflex. At the same moment, there is an increase in pressure in the lower anal canal because of the contraction of the puborectalis muscle (PRM) and the external anal sphincter (EAS) [14,15,16]. To graduate the rectal pressure, the use of a rectal balloon has been proposed. Since then, a rectal balloon has become a mandatory element of this technique. For many years, manometric examination in patients with visible fistulas was not performed since it was impossible to pass a rectal balloon through a narrow fistula. The rejection of the rectal balloon made it possible to perform a manometric study in ARM with visible fistulas. To create high pressure in the rectum, a sharply injected 50 cm^3 of air into the rectum was produced. Anorectal inhibitory reflex was found in all patients in whom it was possible to carry out a measuring device (endotracheal tube) into the rectum. Basal pressure in the anal canal was within the normal range [17]. Ruttenstock et al produced preoperative rectal manometry of rectoperineal or rectovestibular fistula. The manometric device was introduced from the colostomy. The rectoanal inhibitory reflex was found in all patients [18]. Thus, a manometric study in ARM with visible fistulas revealed the functional characteristics of the normal anal canal.

In 5 infants with anorectal malformations (high type 2, intermediate type 3), a preoperative manometric study at the rectal end was performed with a probe introduced from the distal colostomy. This study showed the presence of rhythmic activity in all, and positive reflexive pressure fall by rectal distension in 4 [10]. The presence of a rectoanal inhibitory reflex is a characteristic of the anal can.

C). X-ray examinations in ARM with visible fistulas.

In patients with visible fistulas, the distal intestine at rest is constantly in a closed state. Its length between the rectum and anal dimple in children without a serious megarectum is equal to the length of the normal anal canal (Figure 1. a). During a bowel movement, the anal canal opens to the width of the rectum. At this point, the caudal wall of the anal canal is approaching the anal dimple. The distance between the wall of the anal canal and anal dimple varies from 2 to 5 mm, depending on the age. It is equal to the thickness of the skin and subcutaneous tissue (Figure 1. b). During a barium enema, penetration of the

contrast agent into the upper part of the anal canal in front of the enema tip can be seen intermittently. At this time, the posterior wall of the anal canal at this level is pressed against the tip of the enema by the contracted PRM. It is the radiological equivalent of the rectoanal inhibitory reflex [17,19] **(Figure 1.c).**

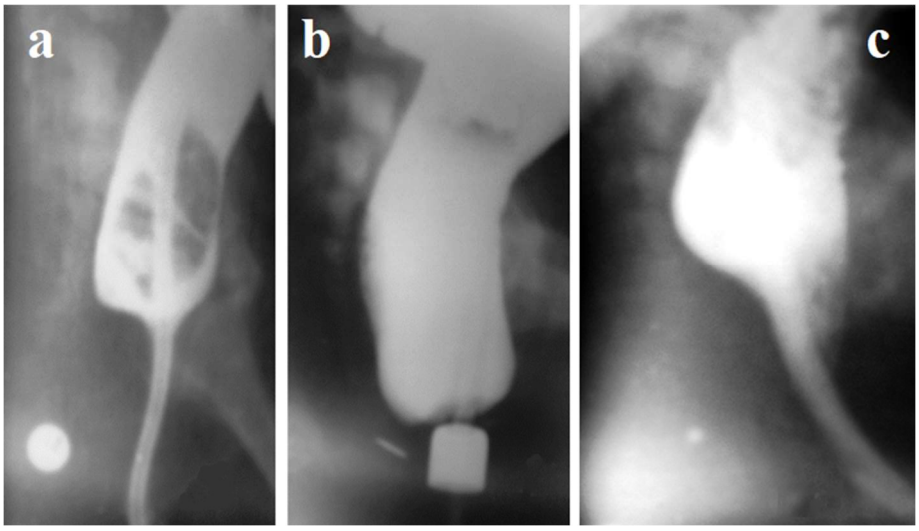

Figure 1. (a, b). The radiographs of the same girl with vestibular fistula performed at different ages. **(a).** At the age of 3 months, the rectum was filled with barium through the catheter, conducted through the fistula. A pushpin is located near the anal dimple. The distal intestine, with a length equal to the length of the normal anal canal, constantly contracts around the catheter, preventing leakage of barium. **(b)** At the age of 9 months, during a barium enema, the wide opening of the anal canal occurred. The distance from the pushpin to the distal wall of the open anal canal equals 4 mm. Barium does not penetrate outward, since the tip of the enema occluded the narrow and rigid ectopic anus. The true diameter of the marker on the enema tip is 1.6 cm. The width of the rectum is 4.3 cm (the maximum rate for children 1-3 years is 3,7 cm). Conclusion: ano-vestibular ectopy, megarectum. The diastasis between the anal canal and anal dimple is (4 mm), which corresponds to the thickness of the skin and subcutaneous tissue. **(c)** Barium was injected into the rectum through an intubation tube (№ 8), passed through the vestibular fistula. The penetration of barium into the upper part of the anal canal in front of the tube is seen, which was accompanied by a decrease in pressure in the upper part of the anal canal. The posterior wall of the anal canal is pressed against the tube by the contracted PRM. This is a typical X-ray picture of a rectoanal inhibitory reflex, i.e., relaxation of the IAS simultaneously with contraction of the PRM and EAS, except for its subcutaneous part.

Method for determining megarectum and damage of the PRM. X-ray studies show that the distal segment of the intestine in ARM with visible fistulas performs the functions of feces retention and defecation comparable to the normal function of the anal canal. However, in contrast to the norm, the narrow outlet is displaced anteriorly. The distal 2-4 mm of the anal canal is located outside the subcutaneous portion of the EAS. Second, the anal canal usually endsby a rigid ring with varying degrees of narrowing represents. While the baby has liquid feces, the narrow opening does not prevent normal rectal emptying. When hard stool appears, it does not completely pass through the ectopic anus. Stool accumulates in the rectum causing it to expand (megarectum) [20]. De la Torre-Mondragón et al found megarectum in 60% of patients with visible fistulas during a preoperative study [21]. This information turned out to be unexpected for the authors since the literature does not describe the relationship between megarectum and the narrowness of the fistulous opening. However, for accurately determine the width of the rectum; it is necessary to compare it with the age norm. Table 1 shows the age standards for the width of the rectum and the length of the anal canal [20, 22].

Table 1. The normal width of the rectum and length of the anal canal in children of different ages adjusted for projection magnification [22].

Age	Width of the rectum (cm)	Length of the anal canal (cm)
5 day – 11 months	1.3-3.0 (2.21±0.09)	1.7-2.5 (2.21±0.15)
1-3 years	3.0-3.7 (3.21±0.11)	2.3-2.8 (2.55±0.10)
4 – 7 years	3.0-3.9 (3.43±0.14)	2.5-3.6 (3.17±0.14)
8 – 10 years	3.2-4.1 (3.72±0.05)	2.6-3.7 (3.11±0.10)
11 – 15 years	3.6-4.6 (0.07±0.07)	3.1-3.9 (3.42±0.10)

In our study, 48 (87%) of 55 patients with visible fistulas aged 1 day to 12 years had chronic constipation before surgery. Each of them on X-ray examination had a rectum wider than the maximal limit for age. The length of the ectopic anal canal was measured in 21 patients under one year of age. It ranged from 1.7 cm to 3.2 cm (average 2.44 ± 0.08 cm). Only in one case, it was shorter than the age norm. In 18 patients older than one year, the length of the anal canal ranged from 1.3 cm to 3.6 cm (average 2.38 ± 0.17 cm). Only in 8 (44%) of 18 patients, the length of the anal canal was within the age norm. In 10 (56%) patients, the length of the ectopic anal canal was shorter than the minimum age norm. In four cases, an X-ray examination was performed twice before surgery **(Figure 2)** [17].

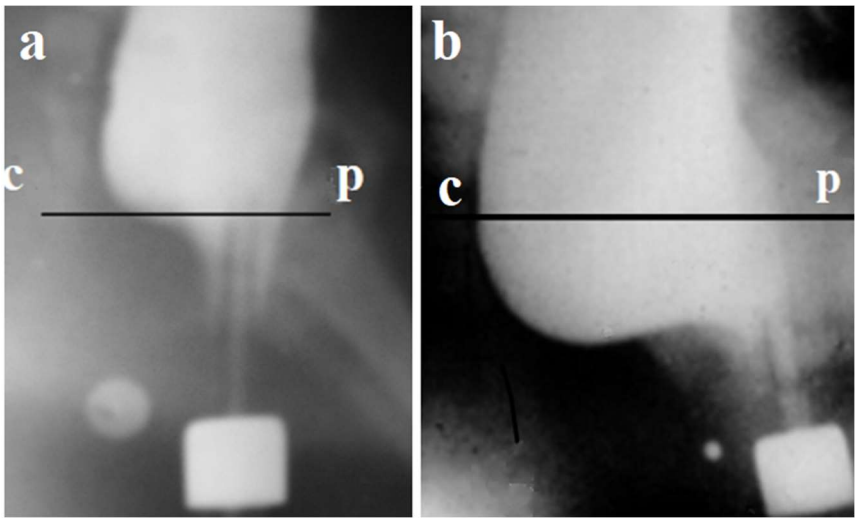

Figure 2. Lateral radiographs of the anorectum made in the same girl with a vestibular fistula at different ages. P-C is the pubococcygeal line. The true diameter of the contrast marker strung on the tip of the enema is 1.6 cm. It is located near the fistula orifice. (a). At the age of 8 month the permanent contraction of the ectopic anal canal was observed during the barium enema. Its length is 2.5 cm, which corresponds to the anal canal age norm. The width of the rectum is 3.4 cm, which is greater than the maximum normal limit (3 cm) (megarectum). Barium penetrates the anal canal behind the tip of the enema. This shows the weakness of the PRM, which does not pull the posterior wall of the anal canal forward. (b). At the age of 1.5 years, she had severe constipation and soiling. The width of the rectum is 5.5 cm, which significantly exceeds the maximum limit of the norm for this age (3.7 cm). A megarectum is combined with a significant shortening of the anal canal. Its length is 1.9 cm (the minimal limit is 2.3 cm). Conclusion. An ARM with vestibular ectopy, megarectum, and descending perineum syndrome.

A strong peristaltic wave of the rectum tries to expel large fecal mass that cannot pass through the narrow fistulous opening. Such repeated bougienage pelvic floor impairs muscle tone until it disappears completely. During defecography, a descent of the "rectum" relative to the pubococcygeal line is determined [23, 24]. The use of a contrast marker near the anal fossa allows us to determine the descent perineum during a simple barium enema by shortening the distance from the "rectum" to the anal fossa [19, 20]. Thus, the shortening of the ectopic anal canal in ARM with a visible fistula indicates severe damage (distension) of the PRM and levator plates, because of which the upper part of the anal canal participates in the accumulation of feces, like the rectum. The PRM weakness is the cause of encopresis before surgery. Dissection of distal stenosis to ensure unimpeded passage of feces should be performed as early as possible to avoid these complications.

D) X-ray examinations in ARM without visible fistulas in males.

In a newborn's first hours of life, the rectal pressure is below a threshold level. Therefore, the anal canal is in a closed state, and meconium with gas is in the

rectum. Only after 30 hours of birth does the rectum collect enough gas and meconium to create pressure that opens the anal canal. For example, Hosokawa et al on the sonograms found, that the pouch-perineum distance on the next day was significantly shorter than on the birthday (P = .001) [13]. This is because irritation of the perineum by the probe provokes an anal reflex. In some newborns, on the second day, rectal pressure reaches a threshold value, which leads to the opening of the anal canal with a decrease in the pouch-perineum distance. In cases where it is below the threshold value, this distance remains the same as on the first day. Therefore, the average statistical indicator is significantly shorter on the second day. In **Figure 3** can be seen the importance of the rectal pressure for anal canal opening.

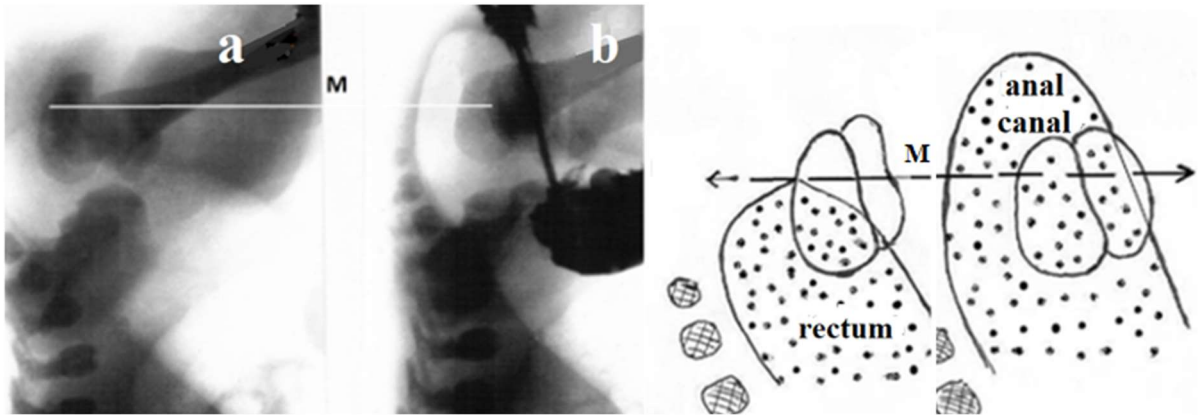

Figure 3. Radiographs of a newborn with ARM without a visible fistula. (a) Invertogram took 12 hours after birth. The distal contour of the rectum is located on a horizontal line (M) between the middle and distal third ischium, which has a typical pear shape. According to Cremin et al data, this line corresponds to the pubococcygeal line [25] (see scheme). (b) 30 hours after birth the erroneous introduction of contrast medium into the perineal tissue (instead of to the rectum) was produced. The anal canal opened, and gas is visible close to the perineal skin. The rectal width is noticeably larger than in Figure 1a. Line "M" was inscribed because there are no other bony landmarks on radiographs. Thus, 12 hours after birth, the X-ray picture corresponded to the intermediate type of ARM (this is the normal position of the rectum over the contracted anal canal), and after 30 hours during the tension of the abdomen, as a reaction to pain, the anal canal opened, which indicates a low type.

As in normal, the reflex opening of the anal canal takes several seconds. Then, the rectum, adapting to the increased volume of contents, relaxes, which leads to a drop in rectal pressure. This causes a reflex contraction of the anal canal and the displacement of gas from the anal canal into the rectum. In the increasing of rectal contents volume, this situation is repeated several times [26]. Therefore, even 30 hours after birth does no guarantee that at the time of the radiograph the opening of the anal canal will be recorded. The abdominal compression increases rectal pressure and causes the anal canal to open at the time of fluoroscopy (Figure 4).

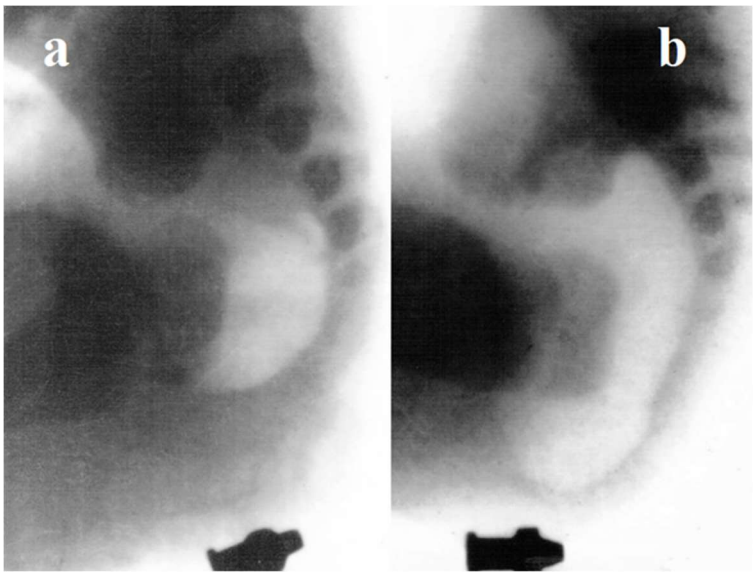

Figure 4. Radiographs of a newborn with ARM without fistula were performed horizontally. A radiopaque marker was glued to the anal dimple. (a). At rest. (b).During abdominal compression, the gas approached the marker. The distance between the marker and the intestine is the thickness of the skin and subcutaneous tissue.

With low rectal pressure, gas is in the rectum, because the anal canal is in constant contraction, i.e., performs normal fecal retention. When the rectal pressure reaches the threshold value, there is a reflex wide opening of the anal canal to allow the passage of feces out. This is a normal defecation function. Thus, neonates without fistula, previously thought to be high type ARM, have a normally functioning anal canal. Therefore, they belong to the low types of ARM.

Augmented-pressure distal colostogram

Kraus et al in the article, on augmented-pressure distal colostogram in boys, state: "… it is extremely important in this regard to understand that the lowest part of the rectum is usually collapsed from the muscle tone of the funnel-like striated muscle mechanism that surrounds the rectum in 90% of cases…" [27]. Meanwhile, it is known from anatomy that there are no muscles around the rectum that are compressing it. The terminal section of the intestine, which is surrounded by muscles, the tone of which overlaps the intestinal lumen and does not allow contrast agent to pass through, cannot be the rectum. The characteristics of this section correspond to the idea of the anal canal, the contraction of which performs the function of the fecal retention. The inference from this article suggests that at least 90% of boys without a visible fistula, including those with recto-bulbar and recto-prostatic fistulas, have a functioning anal canal. The augmented-pressure distal colostogram, with X-ray examination

or with the use of CT or MRI, convincingly shows the presence of an anal canal in urethral fistulas (Figure 5).

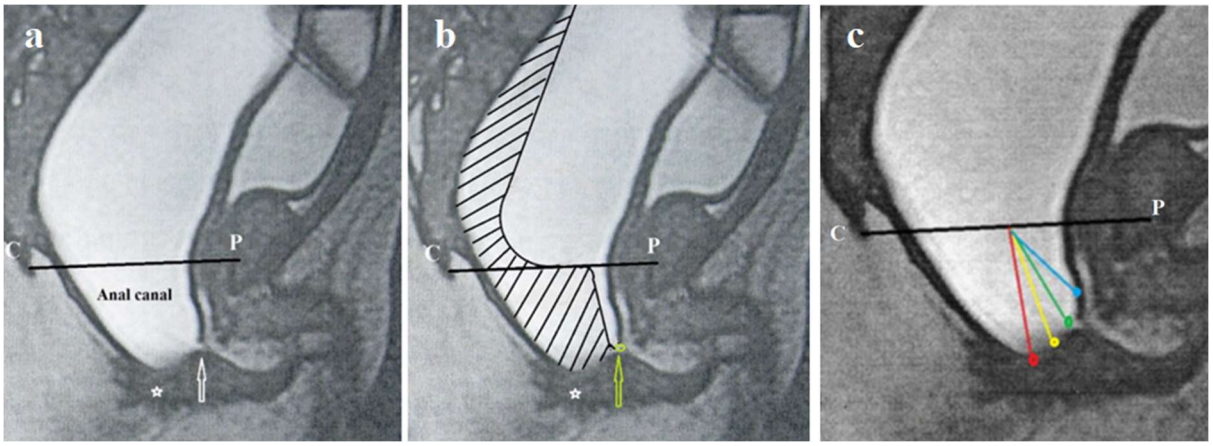

Figure 5. (**a**). MRI imaging during augmented-pressure distal colostogram in a male with recto-bulbar fistula (arrow). Distal to the pubococcygeal line (p-c), a wide-open anal canal is visible. Its blind end is located ≈ 2-4 mm from the proposed site of the anal fossa (asterisk). (**b**). MRI reconstruction scheme with low rectal pressure. The anal canal is closed. Conclusion: Ectopia of the anal canal into the bulbar part of the urethra. (c). Scheme of varying degrees of ectopic anus. The closed IAS is fixed in the place where it penetrated some cavity. Red - normal, yellow - perineal fistula, green - bulbar, blue - prostatic.

In the embryonic period, the endodermal part of the anal canal develops normally by penetration of the IAS inside the sphincters (PRM and EAS). The absence of an opening in the anal fossa indicates a violation of the formation of its ectodermal part. Failing to meet the ectodermal primordium on its way, IAS continues its penetration. Having reached the subcutaneous tissue, it continues its penetration, but it is represented by a narrow, rigid canal. If it penetrates into the center of the subcutaneous part of the EAS, congenital stenosis occurs. More often he continues his way with an anterior displacement. This is how fistulas are formed on the perineum, the vestibule and vagina in females, or perineal and urethral fistulas in males. IAS is fixed to the place of penetration. Therefore, with full disclosure of the anal canal, its length is the same for all types of ARM. The higher the ectopia of the anus, the shorter the closed anal canal. And the whole channel is shifted forward. This means that the EAS, exclusing of its subcutaneous part, also shifts anteriorly. Therefore, it makes no sense to look for a muscle complex on MRI in children with ARM in the same place where it is observed in healthy children [28].

Thus, radiological studies confirm results of histological and manometric studies, that urethral fistulas in boys, which were previously considered high types, have a functioning anal canal, i.e., are low types of ARM.

E) ARM in females with vaginal fistulas.

Vaginal fistulas in females are the symmetrical counterpart of urethral fistulas in males. They may be less high, like bulbar and higher like prostatic fistulas

(Figure 6). Except for different heights there are two varieties of vaginal fistulas, . To understand their formation in the embryological period, one should pay attention to the fact that with the progression of the IAS outside the tissues of the perineum, they form long and narrow fistulous tracts. So, for example, in males, such fistulas go in the subcutaneous tissue and open under the scrotum. In such cases, the entire course of the fistula filled with black meconium can be seen through the skin along the sagittal suture (Figure 6.a). In cases where the fistula has penetrated the vaginal cavity, the vagina has a normal lumen throughout. In cases where the fistula has penetrated the vaginal wall, but the cavity inside the vagina has not yet appeared, the IAS continues to create a narrow channel until it penetrates out. Therefore, there are narrow lower parts of the vagina at different heights. Sometimes they are so narrow that obstruction develops and hydrocolpos. As seen in the Stephens diagram, these fistulas are located below the pubococcygeal line. In addition, on the x-ray, a closed anal canal is visible **(Figure 6).**

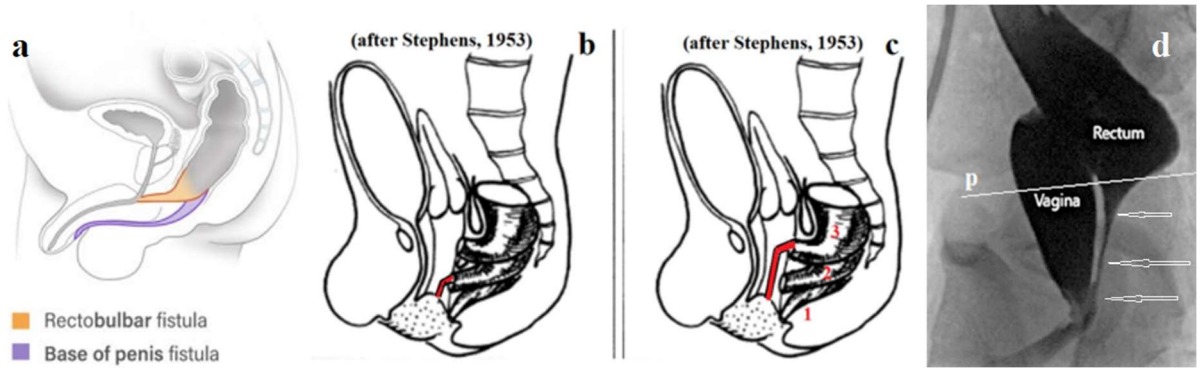

Figure 6. Anatomy and physiology of ARM with fistulas in the vagina. (a). Ano-penial long and narrow fistula in males (b, c) Stephen's scheme of fistulas in females (my additions in red): 1 - vestibular, 2 - low vaginal, 3 - high vaginal fistula. (b) a red narrow and short canal that appeared in the embryonic period, when a cavity had not yet developed in the vagina. (c) a long and narrow channel created under the same conditions. (d) ARM with vaginal fistula. A contracted anal canal (arrows) is determined.

The above evidence indicates that most patients with ARM have a functioning anal canal, and its preservation is essential to achieve acceptable fecal retention and defecation.

F) Results of ARM treatment with anal canal-preserving methods.

First, we should be separate two different methods: (a) preserving the anal canal, and (b) using the BAC.

a) The anal canal in ectopic anus differs from the normal anal canal in that its terminal part is outside the subcutaneous portion of the EAS. However, it should be remembered that the subcutaneous part of the EAS is the smallest of all its parts and "because of muscular fatigue, maximal voluntary contraction of the EAS can be sustained for only 30-60 seconds" [29] in respond to increasing intra-abdominal (rectal) pressure, such as during a cough, lifting from a place, etc. Study of Raizada et al "shows that with voluntary contraction, pressures increase significantly in cranial part of anal canal that is surrounded by PRM" [30]. "Most investigators agree that the PRM is a key component of anal continuity mechanism" [30]. Until now, only two methods ensured the safety of all elements of the anal canal. This is a cutback procedure and incision or stretching of a narrow and rigid fistula длиной 2-4 mm long.
b) Perineal anal transplantation or anoplasty involves high isolation of the IAS along with the rectum from the surrounding tissues and moving it inside the subcutaneous portion of the EAS with suturing it to the skin of the perineum [32,33]. This leads to denervation and devascularization of the IAS and part of the pelvic floor. The IAS, which normally provides about 50% of the rectal pressure, loses its properties. Sclerotic tissue forms around it, which leads to chronic constipation and fecal incontinence.

Secondly, if the defect corrected late, when a megarectum has already appeared because of a narrow fistula, chronic constipation will not be a complication of the operation, but the result of its late application.

Results surgery after cutback procedure.

Stephens and Smith believed that a 'cut-back" anoplasty with the creation of a "shotgun perineum" usually provides the patients with adequate bowel control and genital function. Smith et al, analyzing the experience of 90 patients with ARM, noted "increasing satisfaction with the "cut-back" anoplasty as a definitive procedure or as a temporary stage in low recto-vaginal or recto-vestibular fistulas [34]. Nixon believed that "the simple cut-back described Denis Browne is all that is needed to make the imperfect anus large enough to work where it lies" [3]. The results of the cutback procedure in low types of ARM in males and females in accordance with the Wingspread classification are shown in **Table 2.**

Authors	Good (%)	Fair (%)	Poor (%)
Ackroyd et al. [35]	98	0	2
Kyrklund et al. [36]	85	15	0
de la Fuente [37]	90	8	2
Nixon [3]	90	?	?

These observations confirm the assumption that the unsatisfactory results of the cutback procedure may be since the operation was performed with an already developed megarectum with damage to the RPM function (descending perineum syndrome).

II. Reliability of basic provisions regarding the anatomy and physiology of the ARM in Group 2.

A) Posterior sagittal anorectoplasty In 1982, deVries and Peña published an article sharing the use of pull-though procedure through posterior sagittal approach, called posterior sagittal anorectoplasty (PSARP) which described 34 cases [38]. Two months later, an article by Peña and Devriese was described 54 cases of PSARP application [39]. Surprising is the inexplicable appearance of an additional 20 observations. In this article, statements are made that deVries, known for his research, could not endorse.

In it, Peña stated: "Our incision exposes all the muscle structures in the region. We do not deny the existence of the sling, but we have not been able to identify it. We also do not understand why it should be considered more important than the rest of the muscle structures that we found"; "probably nobody has actually seen it (PRM) by means of the conventional incisions" [39]. This article does not contain information about the presence of the anal canal in 7 patients with a low type. Peña consider "that the outstanding advantage offered by this technique is the extensive expo sure provided and the meticulous repair of the structures for achieving optimum continence".

This article served as the basis for the subsequent denial of (1) the important role of PRM in fecal retention and (b) the assertion that patients with ARM do not have an anal canal [40]. The authors did not produce any studies confirming these conclusions and did not refer to other sources. Returning to Peña's article [39], to which all pediatric surgeons refer, I inevitably ask questions to determine the cause of the contradictions in it.

1. Why Peña, despite the wide opening of the perineum, could not identify the PRM, if all pediatric surgeons, including deVries (not Devriese), successfully detected it and inserted the intestine inside it [1,2,3,4,5,6,11,34,35,36,41]? For example, Bielowicz-Hilgier used “the pull-through procedure advocated by Stephens and Rehbein for ARM in surgery of high-type (above the puborectalis sling). The sacrococcygeal route of Kraske, adapted by Stephens for operative treatment of anorectal malformations, gives approach from behind to the supralevator space for identification of puborectalis sling. The muscle is separated from urethra or vagina, and gradually the tunnel through the sling and external sphincter is created” [41].

2. Maybe before deVries showed him PSARP he had no experience with ARM? This answer is likely, as Peña writes about the possibility of good functional outcomes but does not compare PSARP results with previous experience. Meanwhile, according to Mulder et al “The PSARP for high and intermediate anorectal malformations does not give better functional results than the pull-through operation” [42].
3. Why does he doubt that someone could have identified the PRM during the operation? Is he not aware of literature or does he not trust scientific sources?
4. If he does not see the PRM and the anal canal during the operation, then why does he claim that the final diagnosis is made by the surgeon during the operation [7].
5. What Peña calls the outstanding advantage of the technique, i.e. the extensive exposure provided and the meticulous repair of the structures for achieving optimum continuity, is detrimental to anorectal function, because muscle denervation disrupts their reflex connections, and hence the function of striated sphincters [29,30,31]. The IAS, which provides 50% of the tone of the anal canal, Peña also does not see and therefore excised it.
6. Are these contradictory observations of Peña during surgery scientific evidence that can refute the numerous scientific studies of pediatric surgeons and pathophysiologists? Not!

There is only one explanation that answers all the questions that have arisen: Peña deliberately distorted the truth, stating without any reason that patients with ARM do not have an anal canal and there is no need to preserve the PRP, because with PSARP you have to cross the PRM and to destroy the anal canal, as it turned out not only with visible fistulas, but also with urethral fistulas in males and vaginal fistulas in females.

B) Persistent cloaca.

Peña based on "a retrospective review of all girls with anorectal malformations treated from 1980 through September 2000 and on the pertinent literature” diagnosed cloaca in 42 (6.8%) females who had previously been diagnosed with vaginal fistula. "Of the 617 patients his identified, only 6 were found to have a true rectovaginal fistula, an incidence of 1%" [43]. **Figure 7** shows patterns of ARM with vaginal fistula, which Peña came to call persistent cloaca without any studies of genitourinary and anorectal function.

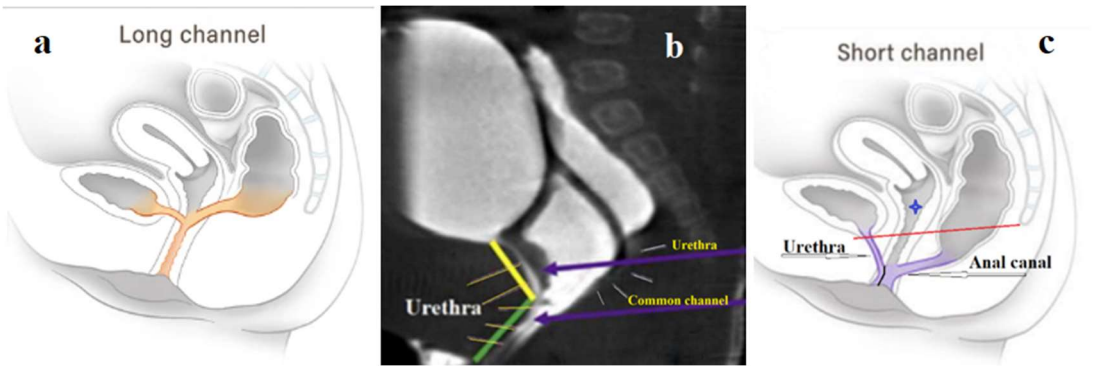

Figure 7. (a & c) Schemes from Stafrace et al [7]. (a). The diagram shows the formed urethra, the vagina, which is narrow in the distal part, and the anal canal. The true length of the urethra is shown in the Figure 7.b; (b). Study from Wood et al [44]. The purple arrows are drawn by the authors of this article. Where "common channel" is indicated, two channels are visible, separated by a gray strip, which represents the walls of the adjacent urethra and the narrow part of the vagina. As in normal, the urethra approaches the vagina and continues its course to the vulva next to the vagina. I marked it with yellow lines. Behind the vagina, a well-contrasted rectum is visible, ending at the level of the PRM (pubococcygeal line). Below it, a poorly contrasted closed anal canal (white lines) is visible. (c). The true length of the urethra finished drawing by me with a black line.

First, since these patients have a urethra, vagina, and anal canal, this means that in the embryonic period they successfully passed the cloacal stage and therefore cannot be considered a cloaca. Secondly, what Peña and his followers call the common canal suggests that after the creation of the urethra, vagina, and anal canal, these formations supposedly again merged into a single canal, which is contrary to the laws of embryology. Thirdly, the presence of an anal canal makes it possible to attribute this type of ARM to low types and to perform operations that preserve the anal canal.

Calling vaginal fistula cloaca, Peña and followers began to apply correction of the urinary system as if it were a real cloaca [45,46]. The functional results of urogenital separation and total urogenital mobilization in persistent cloaca are almost the same as in the classical cloaca, which is diagnosed less than once a year and in which the urethra is absent [47]. Peña and followers justify the poor results of operations by arguing that if the urethra is <1.5 cm long, its function is insufficient to hold urine. This is incorrect for two reasons. First, as shown above, the urethral measurement was incorrect. Second, if 29% of healthy adult women have a urethral length of 2 cm [48], how can a urethra of less than 1.5 cm in infants not be good enough?

The female urethra is the internal sphincter. If it is isolated together with the urogenital complex [46], then it and the bladder lose their innervation and, therefore, the urethra does not open when the bladder overflows, which requires continuous or intermittent catheterization. If it is isolated from the surrounding tissues [45], then it loses not only innervation, but also vascularization, turning into a fibrous tube.

The claim that sacral ration, as a reflection of spinal cord anomalies, is the cause of poor results has no evidence. Below are two pieces of evidence that no concomitant anomalies impair urological and anorectal function in ARM, erroneously called persistent cloaca.

1. Before the change name and treatment, neither journal articles nor textbooks make any difference in the functional results of pull-through operations in patients with vaginal fistulas (read persistent cloaca) compared with other types of high defects. In the literature, there is no information about problems with the urinary system in patients with rectovaginal fistula until most cases were called the cloaca.

2. In articles about persistent cloaca, where surgeons limited themselves to only correcting rectal fistula with the addition of introitoplasty and/or dilatation of a narrow canal, all patients had an intact urogenital complex and none had documented recurrent urinary tract infection until adolescence [49]. Similar results are described by AbouZeid [50]. This means that the poor results of the operation are due to the destructive effect of the operation itself. After surgery, the urethra very often does not function even for experienced surgeons: “When the common channel is shorter than 3 cm, about 20% of the patients will require intermittent catheterization to be able to empty their bladders. On the other hand, when the common channel is more than 3 cm, 80% of the patients require intermittent catheterization” [51]. According to Warne et al up to 50% of patients may have urinary incontinence or may be dependent on clean intermittent catheterization after cloaca repair [52].

Why don't pediatric surgeons who perform operations on the so-called cloaca ask themselves the question: was Peña right in changing the name of this pathology without any examination of the function of the bladder, urethra and anal canal? Do children benefit from the operations proposed to them? After all, the answers to these questions are obvious. No and no.

Except for the so-called cloaca (ARM with vaginal fistula), all other patients undergo pull-through operations with different accesses, the results of which differ little from each other. At the same time, the results are not very good for both defects with good and bad prognosis. The most reliable results are reported in a systematic review by Springford et al. "The prevalence range for long-term active problems was fecal incontinence 16.7% to 76.7%; chronic constipation 22.2% to 86.7 %; urinary incontinence from 1.7% to 30.5%; ejaculatory dysfunction - from 15.6% to 41.2%; erectile dysfunction - from 5.6% to 11.8%" [53].

Compare these figures with the results of treatment of patients after operations that preserve the anal canal in Table 2. When offering the parents of an infant

surgical treatment, the surgeon must offer the parents a choice: either a beautiful ass, or a normal function of holding urine and feces, and defecation.

III. The program of radiological examination of children with ARM, considering their pathological physiology.

A) ARM with visible fistula.

1) Newborns with visible fistula

Since all newborns with a visible fistula always have an anal canal, X-ray examination, CT and MRI is not useful. Ultrasound is necessary to determine the condition of the kidneys and exclude hydrocolpos. To preserve the function of fecal retention and defecation and prevent the development of a megarectum in infants with anal stenosis, perineal and vestibular fistulas, it is enough to cut the narrow rigid anus in two places and insert a tracheostomy tube into the rectum to 7-10 days, which will be fixed with balloon inflated in rectum. Aesthetic correction for vestibular fistulas is possible at puberty. In newborns with a single hole, a colostomy should be done.

2) In infants and children with constipation, a lateral radiograph of anorectum should be taken after injection barium into rectum to determine the degree of rectal dilatation versus normal (see Figure 1). This is necessary to monitor the timely emptying of the rectum, after crossing the stenotic ring, as described above.

B) ARM without visible fistula.

1. Newborns without visible fistula

Fluoroscopic examination in a horizontal position on the side should be performed 30 hours after birth. By this time, enough meconium and gas have accumulating in the rectum to cause a defecation reflex, which is manifested by a wide opening of the anal canal and the approach of gas to a marker glued to the anal dimple. Observation of the radiologist is necessary because the opening of the anal canal is quickly replaced by its contraction and the return of gas into the rectum. Compression of the abdomen between the two palms of the doctor from the back and abdomen provokes a more stable opening of the anal canal. In this case, the following situations are possible:

a) If, during abdominal compression the gas approached the perineum, but immediately disappeared, and the width of the rectum decreased, then there is a previously undiagnosed fistula.

b) If, during compression, the gas approached the perineum and lingered there until abdominal compression continued, and the width of the rectum did not change, then there is an anal canal without a functioning fistula (see Figure 4).

c) If, despite compression of the abdomen, the gas does not approach the perineum, but the width of the rectum is < 2 cm, then there is too little content in the rectum to cause a bowel movement and the examination should be repeated after 4 hours.

d) If the gas does not approach the perineum, but the rectum is wide, then the child has a high type of ARM.

2. **Research in the presence of a colostomy.**

If, after the introduction of the diluted paint into the rectum, it does not appear in the urine, then the patient does not have a functioning fistula, which is observed in almost 100% of cases. Therefore, the purpose of the study is to establish the presence or absence of the anal canal. For the correct execution of the augmented-pressure distal colostogram, it is necessary to create a high uncontrolled pressure in the rectum. However, in such cases there is a risk of rupture of the intestine. Therefore, most researchers try not to create high pressure, but in this case, they do not achieve the desired effect [27]. Secondly, by creating very high pressure, they stretch the non-functioning fistula, turning it into a functioning one. Therefore, this study has no functional justification. The diagnostic method proposed by Pakarinen and Rintala is completely safe and can also be used for surgical treatment. The anal canal is intraluminally visualized using retrograde flexible endoscopy through the previously performed sigmoid mucous fistula. The distal termination of the anal canal is clearly identified as by convergence of the anal columns. Bright translumination of the endoscop light the anal canal to the anal dimple within the external anal sphincter indicates a low malformation [54]. Perforation perineal procedure, without suturing the wall of the anal canal to the skin, preserves all the elements of the anal canal, ensuring the normal function of fecal retention and defecation [19].

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