Pathological physiology of the anorectal malformations without visible fistula. A review.

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All information contained in this review has been repeatedly sent to leading pediatric surgeons, who have published their work on the diagnosis and treatment of anorectal malformations over the past 10 years. At the forum, which assumed an open discussion, there was not a single objection or attempt to correct my views on the pathological physiology of anorectal malformations with visible fistulas. The new view of the pathophysiology of ARM opens great opportunities for improving the surgical correction of this pathology.

Until 1982, pediatric surgeons came to a consensus that in patients with anorectal malformations (ARM), the intestine that is located caudal to the pubococcygeal line is the anal canal and, in order to achieve the best functional result, it must be preserved during surgery. Simultaneously with the publication of posterior sagittal anorectoplasty (PSARP), it was stated that except for patients with rectal atresia and anal stenosis, patients with ARMs are born without an anal canal. It was believed that the rectal pouch or fistula should be removed, which was the justification for the use of PSARP in all types of ARM. **Purpose,** analysis of the literature to define anatomy and physiology of ARM without visible fistulas. **Results.** Analysis of the literature indicates that most males with ARM without visible fistula and females with two openings in the vaginal introitus have a functioning anal canal, which ensures normal fecal retention, and defecation. Females with a single opening with a formed urethra refer to recto-ano-vaginal ectopy, most of which have a narrow vagina with a rigid wall. Most of them have a normal urinary system. This form of defect has been little studied. **Conclusion.** New concepts of the pathological physiology of ARM without a visible fistula allow the use of surgical treatment that ensures the preservation of the urinary system and anorectum in patients with a low type of ARM. Studies of the function of the pelvic floor in girls with a single opening in the introitus are needed.

Keywords: anorectal malformations, anorectal physiology, cloaca, pathophysiology anorectum, vaginal fistula, without fistula.

Introduction In 1953, Stephens proposed the concept of a pubococcygeal (P-C) line, which runs from the lower limit of the pubic bone to the distal coccygeal vertebra. He showed that this line corresponds to the location of the puborectalis muscle (PRM), which is located between the rectum and the anal canal. If the blind end of the intestine is located above this line, these cases are considered a high type of anorectal malformations (ARM), if at the level of this line it is an intermediate type, and low type if the intestine is located more caudally of this line [1]. This understanding of the pathological physiology of ARM was reflected in the Wingspread classification (1984). Since then, it was believed that if the gut is located below the P-C line, it means the patient has an anal canal that needs to be preserved during surgery [2]. Such a subdivision was of great practical importance. In high types of ARM, the pull-through surgery was performing, and with low types, perineal rectoplasty was performed. "The results in low deformities operated by perineal rectoplasty showed "good" clinical scores, good preservation of the rectoanal reflex, and good electrical activity of the external sphincter [3].

In 1982, deVries and Peña published their experience in the use of pull-through operation through the posterior sagittal approach proposed by Jean Zulema Amussat in 1835 [4,5]. The endearing feature of posterior sagittal anorectoplasty (PSARP) is that "in perineal and vestibular fistulas, the rectum is immediately visible" after separating the superficial portion of the external anal sphincter (EAS) from the coccyx, after the incision of muscle complex (which consists from the puborectalis muscle (PRM), deep and superficial portions of EAS) and levator muscles in the midline" [6]. Two months after the first article [4], Peña publishes an article, exclusively based on materials from Mexico City, in which the following provisions are put forward without any justification. [5].

1) PRM is not a sphincter, which means it is not involved in the fecal retention.

2) Most patients with ARM do not have an anal canal.

3) the rectum has a common wall with the bladder in females and with the urethra in males [5]. It is currently believed that "except for patients with rectal atresia and stenosis, patients with ARMs are born without an anal canal". The intestinal segment located caudal to the P-C line is called the rectum, rectal pouch, or fistula [6]. Therefore, the Krickenbeck classification (2005) was adopted, which is a listing of the main types of ARM without a division into high and low types [2]. The main idea, proposed by Dr. Peña and accepted by the community of pediatric surgeons, is that in ARMs the anal canal is absent, and the rectal sac or fistula is so different from the rectum that it cannot be used for defect correction. PSARP is claimed to be an ideal operation. For defects with a good prognosis, a posterior sagittal approach is sufficient, and if the rectum is difficult to find, an additional abdominal approach is used. In such cases, tissue dissection is more extensive, so they have a poor prognosis. The long-term results of ARM treatment indicate that "A large proportion of the patients have persistent fecal incontinence, constipation, and sexual problems that have a negative influence on their quality of life" [7].

The aim of this study was to analyze articles to elucidate the pathological anatomy and physiology of ARMs without visible fistulas.

Material and methods. The article analyzes 50 studies related to the anatomy and physiology of the pelvic floor and the pathological physiology of ARM, which indexed in PubMed, SCOPUS, and EBSCO. To understand the pathological physiology of ARM, it is necessary to use generally accepted anatomical designations and to know the normal anatomy and physiology of the anorectum.

1. Normal anatomy and physiology of the anorectum.

Stool accumulates in the rectum, where pressure on the intestinal wall stimulates reflexes, which play an important role in retaining feces and defecation. The anal

canal, located distal to the pubococcygeal line, is constantly closed. Its length as a zone of high pressure in comparison with the pressure in the rectum increases from 1.7 cm in newborns to 4 cm in adults [8,9]. The same figures were obtained during barium enema, where the anal canal is defined as the distance from the rectum to the contrast marker at the anal dimple along the posterior contour of the enema tip [10] (**Figure 1,** a).

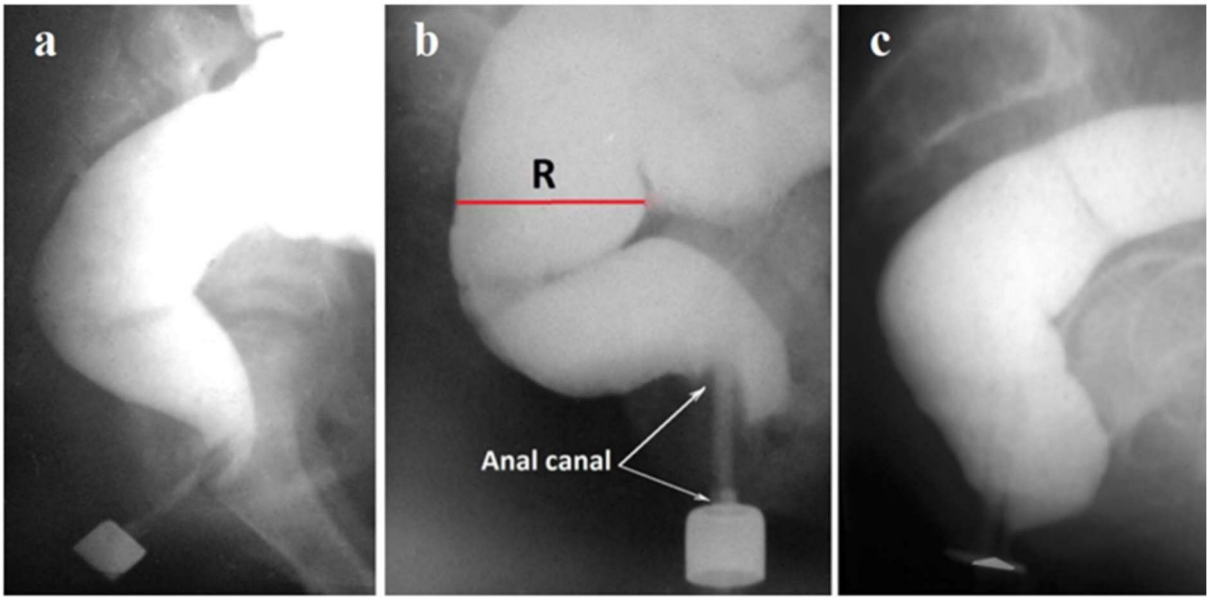

Figure 1. Lateral radiographs of anorectum in different patients without digestive system pathology. (a). In a 12-year-old patient, the length of the anal canal is 3.4 cm. As a result of the contraction of the PRM, the axis of the anal canal is displaced forward relative to the axis of the rectum with the formation of an acute rectoanal angle. (b). In a 9-year-old patient, the anal canal is 3.3 cm long. As a result of IAS relaxation, barium penetrated the upper part of the anal canal in front of the enema tip. The posterior wall in the upper part of the anal canal is pressed against the tip of the enema by contracted PRM. This is a picture of the rectoanal inhibitory reflex. Despite the relaxation of the IAS, barium (fecal) retention occurs because of the contraction of the PRM and EAS. (c). Under the influence of high pressure in the rectum, a wide opening of the anal canal occurred, which is due to the contraction of the levator plates. This attempt defecation was prevented by voluntary contraction of the subcutaneous portion of the EAS.

Table 1 shows the length of the anal canal and the width of the rectum in children of different ages without pathology of the digestive tract [8,9].

Table 1. The normal size of the rectum and anal canal in different ages.

Ages	The width of the rectum (cm)	The length of the anal canal (cm)
5 days – 11 months	1.3 – 3.0 (2.24±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 (3.95±0.07)	3.1 – 3.9 (3.43±0.10)
23 – 64 years	3.5 – 4.8 (3.95±0.21)	3.4 – 4.2 (4.08±0.07)

Fecal Retention. At rest, the IAS and striated muscles of the pelvis floor are in a state of tonic contraction. They help to support the pelvic organs and participate in the continuous retention of feces. When the fecal bolus penetrates from the sigmoid colon into the rectum, it stretches the wall of the rectum and rectal pressure increases. It causes a reflex relaxation of the IAS and contraction of the EAS and PRM (anorectal inhibitory reflex). The PRM pulls forward the upper part of the anal canal. In front, inside the PRM loop the anal pressure decrease because of the IAS relaxation. Between the rectum and anal canal, there is a narrow opening through which the gas and liquid feces can penetrate the upper part of the anal canal. In the mucosa at this level, there are sensors that allow distinguishing the liquid from the gas. During anorectal inhibitory reflex, the formed stool remains in the rectum due to the acute anorectal angle and the narrow hole between the rectum and anal canal. After a few seconds, the rectum adapts to the new rectal volume and relaxes. After entering the rectum of another bolus of feces this picture (anorectal inhibitory reflex) is repeated. This picture can be observed up to seven per hour. During IAS relaxation, its muscle fibers restore contraction ability. In this period, the fecal retention is performed by the

PRM and EAS contraction. During the rise of the intra-abdominal pressure (rise from the spot, cough, etc.), the reflex contraction of all sphincters occurs.

Defecation. When the need for a bowel movement coincides with the possibility of its implementation, a contraction of the abdominal wall and diaphragm increases the rectal pressure to the threshold defecation pressure. A strong peristaltic wave of the rectum expels stool through the open anal canal. The wide opening of the anal canal is due to the relaxation of the IAS, PRM, and EAS, with a simultaneous contraction of the LAM. Any of the pressure levels depend not only on the volume of feces but also from the tone of the rectum. During the opening of the anal canal, its wall is stretched at the level of deep and superficial portions of the EAS. The subcutaneous portion is relaxed, but it is not connected with LAM and therefore does not stretch. Therefore, during the evacuation of soft feces, it forms a tape, the diameter of which depends on the viscosity of the feces [10, 11,12,13].

II. Pathological anatomy and physiology of ARM without visible fistulas

(females and males without fistula; males with urethral fistula).

Histological studies in ARM. In a study by Holschneider et al. it was shown that in patients with ARM "Classical aganglionosis was found in 31% of the rectal pouch specimens, hypoganglionosis in 38%, neuronal intestinal dysplasia (NID) type B in 14%, and dysganglionosis in 10%". In the authors' opinion, "...the recommendation to use the distal rectal pouch and parts of the fistula in the reconstruction of anorectal malformations should be reconsidered" [14]. These histological results were confirmed by other researchers, who believed that the histological structure of the rectal pouch should be consistent with the structure of the rectum [15,16]. Xiao et al., also concludes that the distal rectal pouch has distinct defects in the neuromusculature and need to be respected for better functional outcomes of the remaining gut [16]. First, like the previous authors, they compared the tissue specimens of the rectum, taken from 2 to 4 cm above

the dentate line in the control group, with the tissue specimens, taken in the ARMs group from 0.5 to 2.0 cm of the most distal part of the rectal pouch, i.e. compared the samples from the anal canal with the rectal samples. Secondly, they did not consider that in the postnatal period there are changes in the terminal part of the intestine, associated with chronic constipation due to stenosis of the ectopic anus.

Alamovich et al. investigated the innervation of the normal IAS. This study shows that the internal sphincter itself has no autonomous innervation unlike the rest of the digestive tube [17]. Thus, the research of Holschneider and other authors proves that what they called the "rectal pouch" is the anal canal. Lambrecht and Lierse in neonatal pigs with ARM found that the proximal region of the fistulae in ARM has most features of a normal anal opening: (1) it is surrounded by an internal sphincter, (2) the rectal pouch in the region of the internal sphincter as well as the fistulae are hypoganglionotic, (3) the proximal fistula region is lined by transitional epithelium, and (4) it contains anal glands. They, therefore, consider that the fistula should be designated as an ectopic anal canal. The most important result was the demonstration of a normal internal sphincter even in high and intermediate types of ARM [18]. Rintala et al have shown that in anorectal malformations the distal rectal pouch with the fistulous connection is an anal canal ectopy [19]. Articles that use anatomical designations that are not accepted in scientific research, in which the conclusions are the same for defects of different levels, do not inspire confidence and therefore they are not given in this work.

Manometric study. In 5 infants with anorectal malformations (high type 2, intermediate type 3), for whom colostomies had been performed as newborn, a preoperative manometric study at the rectal end was performed with a probe introduced from the distal colostomy. A preoperative manometric study of the

rectal end showed the presence of rhythmic activity in all and positive reflexive pressure fall by rectal distension in 4 [20].

X-ray examinations.

In newborns first day of life, the rectum with gas is located at the level of the pubococcygeal line, regardless of the location of the baby's body. The idea that the gas rises upward and more accurately determines the location of the intestine on the invertogram is erroneous. Intestinal contents move as a result of peristalsis. In patients with a low type of ARM, the anal canal, as well as in healthy newborns, is in a closed state. For it to open, the pressure in the rectum must rise to the threshold pressure for defecation. This happens when meconium and swallowed air enter the rectum. This condition usually occurs 30 hours after birth. For example, Hosokawa et al on the sonograms found, that the pouch-perineum distance on the next day (mean ± SD, 9.37 ± 4.89 mm; range, 2.1-20.9 mm) was significantly shorter than on the birthday (15.75 ± 6.67 mm; range, 8.1-37.2 mm; P = .001) [21]. Since bony landmarks (pubis and last coccygeal vertebra) are often poorly defined on lateral neonatal radiographs, Cremin et al suggested drawing a horizontal line (M) between the middle and distal third ischium, which has a typical pear shape. According to their data, this line corresponds to the pubococcygeal line [22] **(Figure 2).**

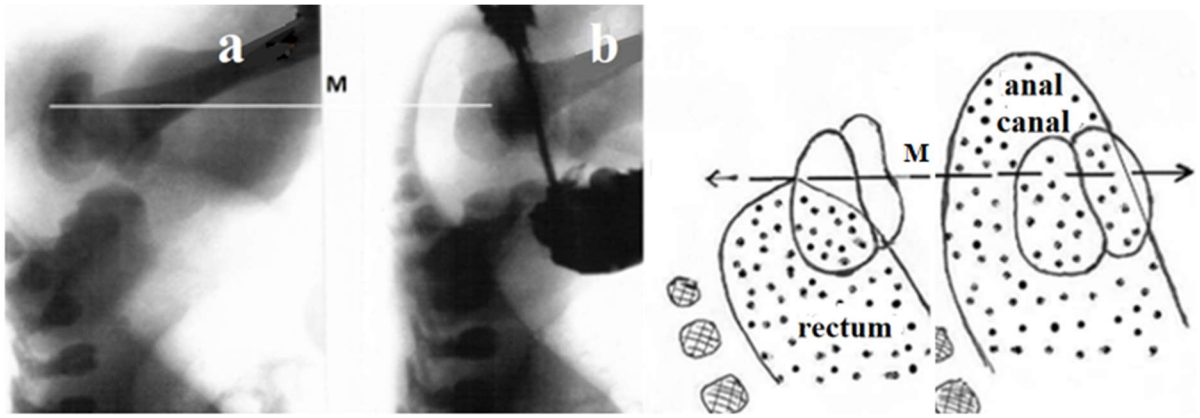

Figure 2. Radiographs of a newborn with ARM without a visible fistula. (a) At rest. The distal contour of the rectum is located on the "M» line (see scheme). (b)

After the erroneous introduction of contrast medium into the perineal tissue (instead of to the rectum), the anal canal opened, and gas is visible close to the perineal skin. Line "M" was drawn through the border of the middle and distal third of the shadow of the sciatic bone, corresponding to the pubococcygeal line.

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 leads to a reflex contraction of the anal canal and the displacement of gas from the anal canal into the rectum. In the process of increasing the volume of rectal contents, this situation is repeated several times. Therefore, the production of an X-ray even 30 hours after birth does not guarantee that the opening of the anal canal will be recorded exactly at the time of the shooting. The compression of the abdomen increases rectal pressure and causes the anal canal to open at the time of fluoroscopy (**Figure 3)**.

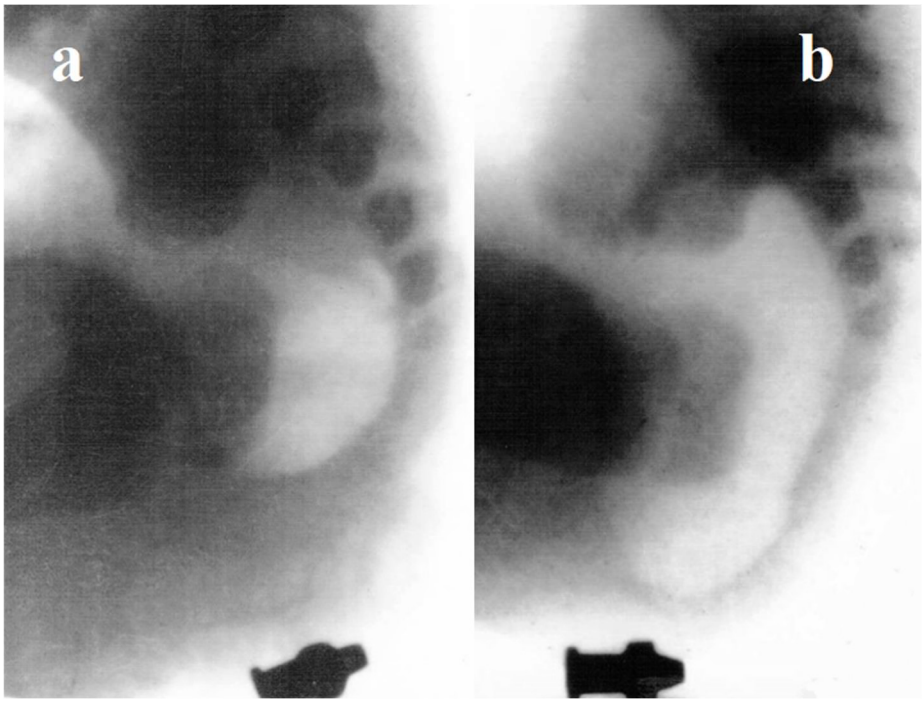

Figure 3. Radiographs of a newborn with ARM without fistula performed horizontally. A radiopaque marker was glued to the anal dimple. (a). At rest. (b). During abdominal compression, the gas approached the marker. The distance between the marker and the intestine is the thickness of the skin and subcutaneous tissue.

In Levin's article, it was shown that newborns who were diagnosed with low type ARM on X-ray examination using abdominal compression, during surgery, the rectum was found 2 cm from the perineum. On this basis, a diagnosis of high-type ARM was established, and pull-through surgery was done [10]. The approach of gas to the perineum in the restless newborn is a known phenomenon. In the literature, it is associated with the descending perineum during an increase in intra-abdominal pressure. It is considered a mistake to assess the level of ARM by the descending perineum since in a calm state the perineum returns to its place [1].

First, from the point of view of formal logic, the perineum of a newborn cannot be displaced by 1 cm. Second, as is known from physiology, during an increase in intra-abdominal pressure, the muscles of the pelvic day do not descend but rise [13].

Some authors call this phenomenon «well-descended rectum" [Nagdeve], ignoring the fact that the rectum is fixed in the tissues of the pelvis and cannot move. The fistula with a urinary tract was found in 11 of 12 patients (seven had fistula to the bulbar urethra and four to the prostatic urethra) with a lower limit of the rectal gas bubble at or below the ossified fifth sacral vertebra They concluded that "repair of high ARM in male neonates with a well-descended rectum is feasible without significant morbidity and good continence" [23].

Kraus et all 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 (ARM without visible fistulas) is usually collapsed from the muscle tone of the funnel-like striated muscle mechanism that surrounds the rectum in 90% of cases…" [24]. However, it is known from anatomy that there are no muscles around the rectum. Muscles surround the anal canal, participating in fecal retention and defecation. These authors in principle refer to the anal canal as a "rectum" "fistula" or "rectal pouch" to avoid the accusation that during

surgery they remove the IAS and destroy the anal canal. On the other hand, this statement suggests that at least 90% of boys with ARM without a visible fistula have a functioning anal canal.

During surgery, the pressure in the rectum is low, so the anal canal is closed. The wall thickness of the IAS in newborns is about 1 mm. A closed IAS both in norm and low ARM is a canal that looks like a fistula. Koga et al proposed a method for measuring the urethral fistula during surgery to remove it without residue [25]. However, «the internal anal sphincter is currently regarded as a significant contributor to continence function" [26].

Examination through a colostomy after administration of a contrast medium under high pressure confirms the presence of an anal canal in patients with urethral fistulas (**Figure 4).**

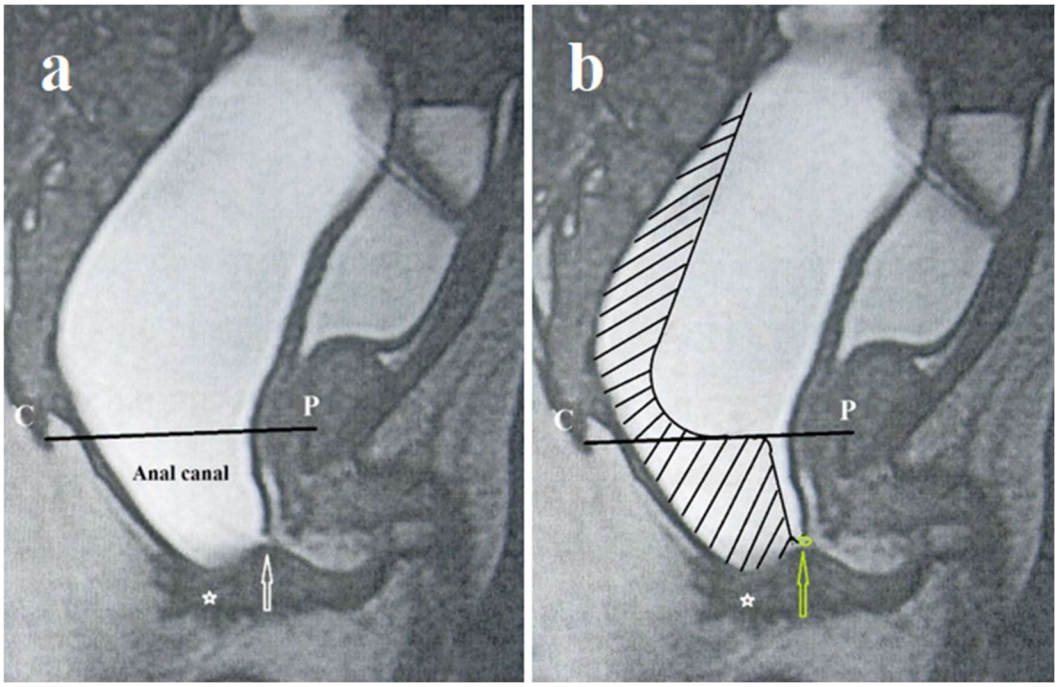

Figure 4. (**a**). MRI imaging during augmented-pressure distal colostogram in a male with recto-bulbar fistula (arrow). Distal to the pubococcygeal line (p-c), a wide-open anal canal is visible. Its blind end is located ≈ 2-4 mm from the proposed site of the anal fossa (asterisk). (**b**). Defect reconstruction scheme with low rectal pressure. The anal canal is closed. Conclusion: Ectopia of the anal canal into the bulbar part of the urethra.

Low types of ARM occur in the post-cloacal period when the IAS migrates caudally and does not meet the ectodermal part of the anal canal on its way, as evidenced by the absence of the ectodermal part of the anal canal. Having reached the subcutaneous tissue, it deflects forward and, most often, penetrates either through the skin or into some organ, depending on the degree of deviation. Gans et al showed that in almost all cases, IAS penetrates somewhere, but it does not always function since it is either too narrow or is represented by a fibrous cord [27]. Since the embryo-pathogenesis of ARM does not have gender differences, vaginal fistulas should be attributed to low ARM without visible fistula, which in frequency should be commensurate with urethral fistulas **(Figure 5).**

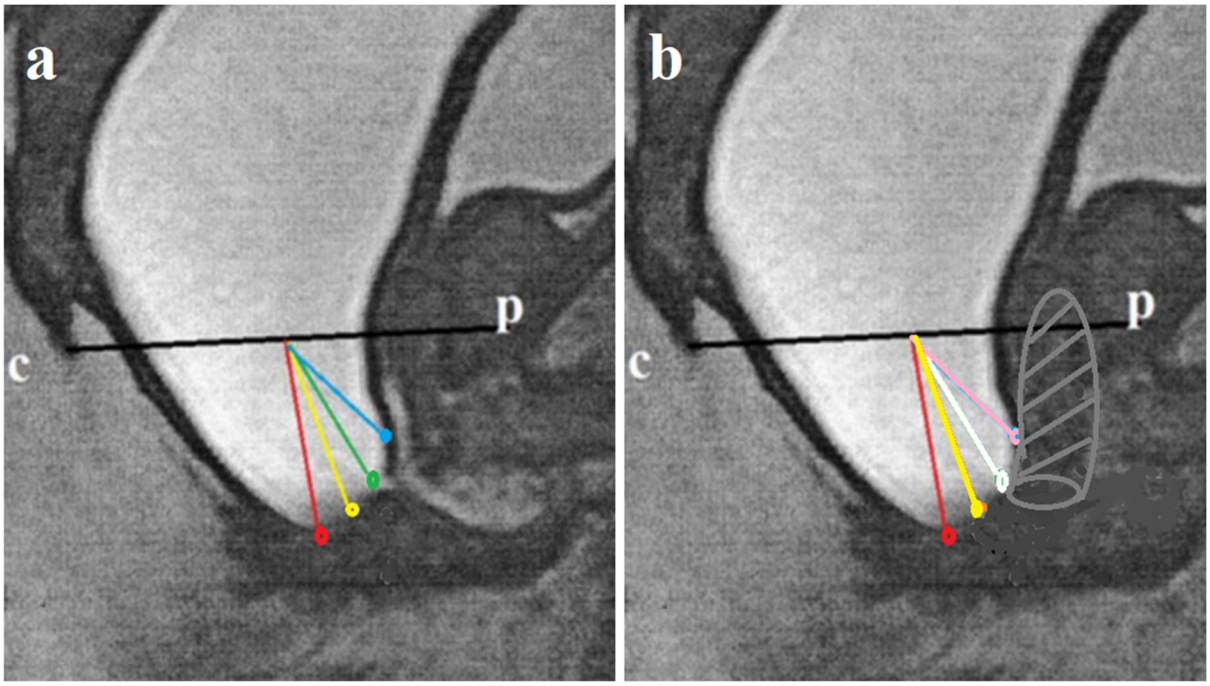

Figure 5. Schemes of a low type ectopia on the different levels. The open anal canal has the same shape and size in different low types of ARM. However, during its contraction, the length of the anal canal is shorter, the higher the anal ectopy. (a). In males: norm -red line; perineal fistula-yellow; recto-bulbar fistula-green; recto-prostatic fistula-blue. (b). In females: vestibular fistula- white; vaginal fistula -purple.

Diagnosis of ARM in newborns without a visible fistula should solve 2 problems: 1) determining the level of the fistula (high or low); 2) determine if there is a functioning fistula. The wider the cavity into which the ectopic anal

canal flows, the wider the ectopic opening. Therefore, the vaginal fistula always functions and is manifested by the discharge of feces from the vagina. Since the ectopic opening is always narrower than the normal anus, abdominal compression allows differentiation between low and high anomalies. The diameter of the urethra in newborns is about 1 mm [28]. The urethral circumference is 3 mm. The fistulous fistula occupies only its posterior wall. Moreover, it is represented by a narrow and rigid ring. Therefore, in many males, urethral fistulas do not function. If a newborn does not have traces of meconium in the urine by 30 hours of life, and when the abdomen is compressed, the gas enters the anal canal and does not disappear, and does not decrease in volume, this case should be considered as ARM without fistula.

Discussion. The ARMs without visible fistula include males with urethral fistulas, children of both sexes without fistulas, and girls with vaginal fistula. From the point of view of Krickenbeck classification, these patients are born without an anal canal. When using PSARD or the laparoscopic method, the end segment of the intestine, called the rectal pouch or fistula, is resected and the rectum is relegated to this place. An analysis of the literature shows that in at least 90% of boys with urethral fistulas, the distal intestine is located below the pubococcygeal line, surrounded by muscles that compress it, preventing the passage of contrast agent or gas. Histological examinations reveal all the signs of IAS. A manometric study reveals a decrease in pressure in this segment in response to an increase in rectal pressure. This is a rectoanal inhibitory reflex, which indicates a normal function of the IAS in ensuring the fecal retention. The increase in rectal pressure because of abdominal compression causes the wide opening of the distal bowel, which is evidence of a normal defecation reflex. All these data indicate the presence of a functioning anal canal, which performs the functions of fecal retention and defecation. It differs from the normal anal canal that its distal segment is outside the subcutaneous portion of the external anal

sphincter, 2-4 mm from the anal dimple. The outlet (anus) is displaced forward and is represented by a narrow rigid ring. At urethral fistulas, displaced anus often does not function and thus functionally does not differ from ARMs without fistula.

With low types of ARMs, the rectum is located above the pubococcygeal line, and below it is the anal canal. Discussion on this matter is impossible since there is no scientific evidence that contradicts this concept. For a correct understanding of the pathological physiology of the defect, it is necessary to restore the previous names: ectopia of the anal canal into the urethra, ectopia of the anal canal into the vagina, ectopia of the rectum into the bladder and low type ARM without fistula [29].

In numerous articles constantly quoted a statement about “the presence of an extensive common wall between the rectum and urethra in males or vagina in females” [30,31], which contradicts scientific evidence. Firstly, in 90% of boys, the urethral fistula is located below the pubococcygeal line, so the wall of the anal canal, and not the rectum, is tightly adjoined to the urethra. Secondly, the presence of the urethra, vagina, and anal canal is the result of dividing the cloaca into 3 separate structures, each of which has its own walls. Third, a study by Pakarinen et al proved that "the distal termination of the rectum, we did not find either intimate contact between the urethra and the distal termination of the rectum in low forms or a common wall between the 2 structures in high forms of imperforate anus without fistula" [32]. Therefore, a reasonable question arises, why should a well-functioning anal canal be removed with great effort and with the danger of damaging the urethra and vagina?

Wingspread classification subdivided the ARMs into high and low types. This was of great diagnostic value since in low type there is an anal canal, which was preserved during the correction of the defect. At present, ARMs are subdivided into good and not good function prognosis” [31]. Differentiation is made based

on the results of the operation. If the intervention was limited to PSARP, a good result is expected, but if it was necessary to supplement with an abdominal approach, then the function prognosis is less favorable. It is presumed that the more extensive the tissue dissection, the worse the functional result.

Analysis of the literature suggests that functional outcomes after PSARP are independent of any circumstance. First, although the outcome of low anomalies (with good function prognosis) has traditionally been considered good, recent more critical long-term follow-up reports show a different picture. Chang et al showed that constipation occurs shortly after operation [33]. According to Lombardi constipation in "low" ARM has been reported in 42%-70% of cases [34]. A study by Daher et al showed that functional prognosis in patients with low-type lesions is no better than in intermediate and high lesions [35]. Second, for many decades, newborns with ARM have undergone colostomy. When the patient reached 3-6 months, the defect was corrected. The colostomy was closed after the successful expansion of the neoanus. Such a 3-stage treatment corresponds to the axiom that the older the patient is, the easier it is for him to endure the operation, and the easier it is for the surgeon to perform anatomically accurate correction [1-6]. In recent years, more and more pediatric surgeons perform operations on newborns in one stage, i.e. without colostomy stage. It has been stated that the primary repair of ARMs with recto-urinary fistula is a feasible, safe, and effective technique in the neonatal period. [36,37,38].

The pathological physiology of ARMs allows us to understand why after PSARP "the range of reported prevalence of long-term active problems was as follows: 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%" [39].

The PSARP implementation includes the following steps: 1) Separation of the superficial portion of the external anal sphincter from the coccyx. This fixation

of EAS to the coccyx contributes to the wide opening of the distal part of the anal canal during defecation [11]. 2) Intersection in the midline of a puborectalis loop, deep and superficial portions of the EAS. In the surgical wound, it is impossible to distinguish these muscles. Therefore, it is not realistic to sew dissected parts of the puborectalis muscle in the final stage of the operation. Therefore, after surgery, the PRM is not involved in the fecal retention. 3) The IAS резецируется, без которого нормальная функция удержания кала невозможна [13, 40, 41]. 4) Separating the rectum from the levator plates. As a result, levator plates do not open the neoanus during an attempted bowel movement, which leads to chronic constipation. 5) The denervated and devascularized rectum is placed in the wound channel of the pelvic floor muscles, but not in the place where the IAS was located. 6) Transection of the nerves to the levator ani, of the pudendal nerve, inferior rectal nerve, middle rectal plexus leads to denervation of the pelvic organs, which leads to the disappearance of the inhibitory reflex and the defecation reflex. An injured EAS can function in fecal retention, however, due to the lack of nerve connections with the rectum, it does not relax during an increase in pressure in the intestine, which is another cause of chronic constipation.

"The anorectum is a region with a very complex structure, and surgery the anorectum is impossible without accurate anatomical knowledge" [41]. The PSARP destroys this structure, so it makes no difference when to perform surgery or how intensively the distal gut is mobilized.

ARMs with vaginal fistulas also belong to defects without a visible fistula since the ectopic anus is located above the vestibule. For many decades, the number of such patients significantly outnumbered the number of patients with cloaca. In 2002, an article was published that dramatically changed the view on the pathogenesis of the cloaca [42]. The explanation is as follows. "A retrospective review of all girls with anorectal malformations treated by the senior author from

1980 through September 2000 was performed, and the pertinent literature was reviewed". As a result, only 6 (1%) of the 617 patients were found to have a true rectovaginal fistula. A total of 139 of the 617 patients were referred after a previous repair. Of these, 42 had a diagnosis of recto-vaginal fistula originally. The diagnosis was incorrect in all 42. Twelve patients had a rectovestibular fistula, and 30 had a cloaca [42]. There is no scientific research or literature in this article that would allow the senior author to change the understanding of the pathophysiology of ARM in girls without visible fistula. Meanwhile, это "discovery" also contradicts scientific evidence like the previous unsubstantiated assertions of this author: PRM does not play a significant role in the fecal retention; most patients with ARM are born without an anal canal; the rectum has a common wall and the urethra and boys and the vagina in girls, etc.

First, by many generations of anatomists, embryologists, surgeons, it has been proven that "Persistent cloaca results from the total failure of the urogenital septum to descend and therefore it occurs a very early stage of development (10 mm stage). This means that there was no division of the cloaca into 3 channels (urinary, vagina, and anal canal). In 1997, this author described the total urogenital mobilization to repair cloaca [43]. In this article, a diagram of the alleged cloaca shows the urethra and the anal canal that penetrates the vagina (Figure 6.a). At the Belarusian Center for Pediatric Surgery, a cloaca was diagnosed in one (2%) of 48 consecutively examined girls with ARM [44] **(Figure 6.b).**

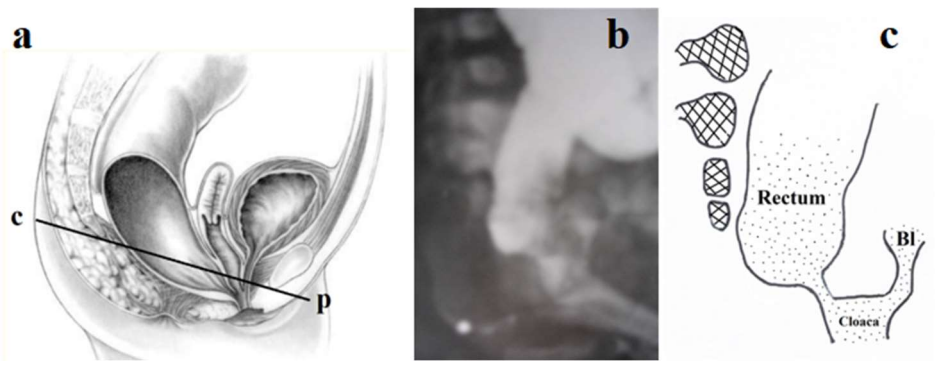

Figure 6. (a). Scheme of the persistent cloaca by Peña. It shows three channels. The orifices of the urethra and intestine are located caudal to the pubococcygeal line (P-C), which indicates the presence of the urethra and anal canal. (b). The radiograph of the cloaca and scheme to it (c). The contrasting of the rectum and bladder, communicating with the wide cloacal cavity, is determined [44].

Firstly, those forms of ARM, which since 1997 Peña began to classify as the cloaca, have nothing to do with cloaca. The canal, which the authors call the common canal, it is the vagina. Secondly, the presence of the urethra indicates the presence of an internal urethral sphincter. It is highly likely that such patients also have an external urethral sphincter, but no studies of the sphincters have been performed. And the function of the urinary system was not studied before the operation. Third, what practical benefits did this innovation bring? It became possible to easily catheterize the bladder. The author claims: "when the common channel is longer than 3 cm, 70% of my patients need intermittent catheterization" [43]. However, this is a clear result of surgery with denervation of the pelvic organs (PSARP and total urogenital mobilization), because after pull-through surgery, the function of the urinary system is not affected [45,46].

According to Kittur et al, total urogenital mobilization in infancy is a difficult procedure and even in the best of hands can be followed by serious complications such as urethral stenosis, complete vaginal and anal closure, tight introitus, neurogenic bladder, and urinary incontinence. Up to 50% of patients may have urinary incontinence or may be dependent on clean intermittent catheterization after cloaca repair. They have done a follow-up study of 9 female patients born with a cloaca, who had rectal pull-through alone in infancy, leaving the urogenital region untouched. The common channel after full growth was used as a vagina. Five patients got married. Three have reported regular sexual intercourse and one has conceived. Three patients, who are not married, had introitoplasty done for free menstrual flow. Four patients needed introitoplasty beyond 20 years of age. With these measures, the common channel was used as a vagina for regular

intercourse. Only one patient out of nine reported urinary and fecal incontinence which was associated with a poorly developed sacrum. All the others were continent for urine and stool. The authors concluded that there is a strong case for not subjecting cloaca patients to TUM which has a sizeable potential of developing urinary incontinence [45].

Unfortunately, there are no studies of the function of the urinary system and urethral sphincters in girls with a so-called cloaca before surgery in the literature. There are no high-pressure colostomy examinations to determine the level of the defect. All authors refer to Peña, who also did not do any research. In the absence of the anus, the diagnosis is based on an overview of the perineum: three openings - vestibular fistula; two openings - rectovaginal fistula or vestibular fistula with vaginal agenesis; single opening - cloaca. Nevertheless, some conclusions from the analysis of the literature can be drawn already now.

1. ARMs with a formed urethra are not cloacal malformations, but vaginal ectopy. 2. Most of these patients have normal urinary function, as evidenced by the results of pull-through surgery [45,46]. Urinary incontinence after PSARP and TIM is due to iatrogenic denervation of the pelvic floor. 3. Most patients have a narrow vagina with rigid walls. This problem is solved by gynecologists at any age. 4. Probably some patients have an anal canal, as evidenced by Peña's scheme, the use of anoplasty [46], and study in dogs [47]. Further functional studies are needed to develop optimal treatments.

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