Analysis of the article "Anorectal Malformations"

by Wood and Levitt

The article by Wood and Levitt “Anorectal Malformations” [1] until recently could be considered the head of a textbook on colorectal pediatric surgery. It contains the postulates that most pediatric surgeons still adhere to. It should be noted that most of them contradict the knowledge that was adopted before the enchanting victory of posterior sagittal anorectoplasty (PSARP). So, what was it - the Renaissance or the Middle Ages? Let's turn to the facts.

The authors write that: "It is important to not make any decisions regarding the surgical management before 24 hours of life because significant intraluminal pressure is required for the meconium to be forced through a fistulous tract, which helps to establish the diagnosis". "If the neonate has signs of a rectoperineal fistula, primary surgery in the form of an anoplasty, without a diverting colostomy, can be performed then or in the first few months of age (Fig. 1)" [1].

![Image](image_url)

**Fig. 1**

Cross-table lateral film in two newborns. (a) In a reachable rectum, a newborn anoplasty can be performed. (b) In a distant rectum, a colostomy is required.

1. Pathophysiology of the low-type ARM.

First, the authors argue that in one of the two cases of perineal fistula there was a low location of the rectum (a), and the other had a high location (b), which influenced the choice of treatment tactics. This example contradicts the authors' statement that "The Wingspread classification (1984), which described malformations as low, intermediate, and high, does not help predict the type of
surgery required or the clinical outcome." Meanwhile, the Krickenbeck classification is a simple enumeration of different forms of ARM, which, unlike Wingspread classification, can in no way help in choosing treatment tactics.

Secondly, according to the Wingspread classification, all perineal fistulas are low types of ARM. This means that the distal intestine located lower from the pubococcygeal line. And as was proved by the Stephens study, the intestine located caudal to the pubococcygeal line is the anal canal [2]. From this, it follows that x-ray studies in newborns with perineal fistula do not make any sense, therefore they are harmful. Figure 1 analyzes these radiographs.

**Figure 1.** (a). A wide opening of the anal canal with simultaneous peristaltic contraction of the rectum is determined. The anal canal is located below the P-C line (black). As always, the blind wall of the internal anal sphincter (IAS) is located 2 mm from the anal fossa. The red line shows the width of the anal canal, which the authors call the fistula. Real fistula is located between the wall of IAS and the skin of the perineum (white arrow). X-ray picture of the normal defecation reflex, which is excited at high pressure in the rectum. (b). The white dot, which should be located near the anal fossa between the buttocks, is actually located on the buttock. The approximate location of the anal fossa is marked with a red asterisk. The rectum, located above the P-C line (black), appears wider than the anal canal in the image (a). A comparison of the height of the vertebrae in two newborns indicates that the image (b) taken with a large projection magnification. The correction factor is 0.6. Therefore, the actual width of the rectum is much
smaller than in the picture (a). The yellow arrow shows the penetration of gas into the upper part of the anal canal with an offset forward. This is a typical manifestation of a rectoanal inhibitory reflex. Gas from the rectum entered the anal canal because of IAS relaxation. The contraction of the puborectalis muscle (PRM) shifts the upper part of the anal canal forward. The anal canal is closed because the pressure in the rectum is lower than the threshold pressure of the defecation reflex.

An analysis of the radiographs from the article Wood and Levitt shows that the intestinal segment below the pubococygeal line is in a closed state. When the rectum expands to a certain size (pressure), IAS relaxes with a simultaneous contraction of PRM and EAS (rectoanal inhibitory reflex). With a larger expansion of the rectum (pressure), the distal segment opens wide, which is accompanied by the strong rectal peristalsis and its contents are forced out. The normal anal canal functions in the same way.

Secondly, the results of this analysis completely coincide with the results of a barium enema in patients with visible fistulas (perineal and vestibular [3] (Figure 2).

![Figure 2](image)

**Figure 2.** Lateral radiographs of the anorectum, made in the same female with vestibular fistula at different times. (a). At the age of 3 months. The rectum is filled with barium through the catheter, conducted through the fistula. Button is located near the anal dimple. The distal intestine, with the length equal to the length of the anal canal, contracted around the catheter, preventing leakage of
barium. (b). In 9 months. During a barium enema, involuntary defecation occurred with a wide opening of the anal canal. The distance from the button near the anal dimple to the distal wall of the open anal canal is ≈ 4 mm.

In the human body, there are no analogs of anorectum. Therefore, its function causes numerous disputes. The pathophysiology of anorectum with ARM manifests itself in these radiographs.

1. The intestine located below the pubococcygeal line is in constant contraction and successfully performs the fecal retention (barium).

2. During bowel movements, it opens wide to the width of the rectum. In this case, its width is 4 cm with an average norm of 2.2 cm (magarectum). Since there are no voids in the muscles of the pelvic floor, the appearance of the intestine with a diameter of 4 cm is possible only as a result of the formation of the channel by the contracted pelvic muscles. In accordance with the place of attachment, it can only be levator plates. One end they are attached almost along the perimeter of the lower edge of the rectum and are associated with the longitudinal layer of the anal canal. Another end is attached to the pelvic ring. The lowest wall of the opened anal canal is always opposite the anal fossa. The fistulous opening is shifted forward and upward. Since IAS is fixed to the fistulous opening, during the contraction of the anal canal, its length decreases. The greater the displacement, the higher it is, and during the contraction of the anal canal its length becomes shorter (Figure 3).

3. In all children with visible fistulas during a manometric examination before the surgery, the normal basal pressure and rectoanal inhibitory reflex were found, which also confirms the presence of the anal canal [3,4].
4. As shown in a study by Kyrklund et al, "All males treated for low ARMs (rectoperineal fistula) with cutback anoplasty, incision of anocutaneous membrane, or dilatations had voluntary bowel movements; 98% of patients were socially continent (p = NS); 67% of patients and 64% of controls were totally continent (p = NS)." Constipation amongst patients (33 vs 3% in controls; p < 0.0001) declined significantly with age. Outcomes by bowel function scores were good at 85% and satisfactory in 15%" [5]. Thus, the long-term results of operations and procedures that fully preserve the anal canal indicate normal function fecal retention. Constipation amongst 33% of patients, which declined significantly with age, can be explained by the fact that these children were operated on after they developed a megarectum.

Based on the foregoing, the authors of the article have no reason to state that "Except for patients with rectal atresia and stenosis, patients with ARMs are born without an anal canal, and therefore sensation does not exist or is rudimentary" [1]. An analysis of the article and the literature convincingly proves that at low type ARMs there is a functioning anal canal, the preservation of which provides the normal function of fecal retention and defecation.

This truth was considered an axiom until 1982. In combination with new ideas about the function of the anal canal, it changes the tactic and method of diagnosis of the ARM.

Firstly, ARM's with visible fistulas are a low abnormality and do not need x-ray examination. Secondly, the cross-table film determines the opening of the anal canal at a certain threshold pressure (defecation) in the rectum. This pressure most often appears at the end of the first day. However, after the opening of the anal canal, the rectum adapts to increased pressure, the tone of the rectum decreases, and the pressure in it decreases. As a result of this, the IAS contracts and gas is squeezed into the rectum. This situation can continue several times [6]. There is no guarantee that cross-table film is produced at the time of the anal canal opening. This confirms the sonographic measurement of the pouch-perineum distance. It on the next day (mean ± SD, 9.37 ± 4.89 mm: range, 2.1-20.9 mm) was significant shorter than on the birthday (15.75 ± 6.67 mm; range, 8.1-37.2 mm; P = 0.001) [7]. Therefore, this method is considered uninformative. Compression of the abdomen in the horizontal position of the newborn 30 hours after birth increases intra-abdominal and rectal pressure, provoking the opening of the anal canal.
2 About cloaca.

The authors of the article claim that "Cloaca, in the past, was considered a rare defect, whereas rectovaginal fistula was reported commonly, but the converse is true, as cloacas comprise the third most common defect in females (after vestibular and perineal fistulas)" [1]. This proposal there is no sense. The authors refer to an article by Rosen et al in which there is no evidence other than links to experience by the senior author (Peña) [8]. By many generations of anatomists, embryologists, surgeons, it has been proven that "Persistent cloaca this is a rare condition that occurs in female infants. It results from the total failure of the urogenital septum to descend and therefore occurs a very early stage of development (10 mm stage). “A persistent cloaca is a symptom of a complex anorectal congenital disorder, in which the rectum, vagina, and urinary tract meet and fuse, creating a cloaca, a single common channel” [Wikipedia] (until 2017). In 2017, Levitt replaced this article on Wikipedia. All other anorectal malformations occur in the post-cloacal period.

![Persistent cloaca](persistent_cloaca.png)

Figure 4. Scheme of the persistent cloaca from an article by Kraus et al [9]. The type of abnormality that the authors call the "cloaca" arisen in the post-cloacal period since there is already a urethra and anal canal below the pubococcygeal line. This means that the cloacal membrane has already divided the cloaca into urogenital and rectal parts.

Serious urological problems (urinary incontinence, infections, etc.) are observed after repair of the rectal component in patients with "persistent cloaca". Dr. Peña and his followers convince pediatric surgeons that PSARP is an ideal surgery. Therefore these symptoms are due to the urogenital sinus. Они рекомендуют total urogenital mobilization (TUM) in infancy. Unlike Dr. Peña, I trust only reliable facts.

Firstly, the coup in the understanding of “cloaca”, as well as with respect to the anal canal at low ARM, was carried out on a hunch, i.e. without any research or evidence from literary sources. This can be seen from the discussion:

Dr. Herldren (Boston, MA) asks:
"Do you think the external sphincter is of any importance in these patients? And if there is any importance to it, do you think that this mobilization risks any injury to the external sphincter?"

Answer of Dr. Peña:

"Concerning the external sphincter, my experience in the management of cloacas is that the girls who suffer from urinary incontinence don’t suffer from the lack of urinary sphincter but rather because of lack of contractility of the bladder. When the common channel is longer than 3 cm, 70% of my patients need intermittent catheterization." "I don’t believe that the external sphincter plays an important role in urinary control in general." [10]. Until then, when dr. Pe Serious urological problems (urinary incontinence, infections, etc.) are observed after repair of the rectal component in patients with "persistent cloaca". Dr. Peña and his followers convince pediatric surgeons that PSARP is an ideal surgery. Therefore these symptoms are due to the urogenital sinus. Они рекомендуют total urogenital mobilization (TUM) in infancy. Unlike Dr. Peña, I trust only reliable facts.

Firstly, the coup in the understanding of “cloaca”, as well as with respect to the anal canal at low ARM, was carried out on a hunch, i.e. without any research or evidence from literary sources. This can be seen from the discussion:

Dr. Herldren (Boston, MA) asks:

"Do you think the external sphincter is of any importance in these patients? And if there is any importance to it, do you think that this mobilization risks any injury to the external sphincter?"

Answer of Dr. Peña:

"Concerning the external sphincter, my experience in the management of cloacas is that the girls who suffer from urinary incontinence don’t suffer from the lack of urinary sphincter but rather because of lack of contractility of the bladder. When the common channel is longer than 3 cm, 70% of my patients need intermittent catheterization." "I don’t believe that the external sphincter plays an important role in urinary control in general." [10]. Until then, when Dr. Peña published his experiments considering that he was all getting away with, there were no such serious complications in girls with ARM.

Secondly. PSARP destroys the anal canal at low types of ARA and denervates the pelvic organs the more, the stronger the dissection of the organs.

Thirdly. None of the articles devoted to the description of the cloaca does not contain the accepted anatomical designations and information on the physiology
of urination. It is known that the retention of urine is provided by three muscles: the internal urethral sphincter, the external urethral sphincter, and the pubourethralis muscle, as part of the puborectalis loop. These sphincters connected by nerve paths constitute a highly organized system [11]. The authors did not study these reflexes either before or after surgery. They never even mentioned the sphincters. They proclaim a completely fantastic goal of the operation: lengthening the urethra by creating a channel in the tissues, which supposedly can improve urine retention.

**Fourth,** 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. There is a strong case for not subjecting cloaca patients to TUM which has a sizeable potential of developing urinary incontinence. Furthermore, the common channel can be used as a vagina along with introitoplasty and dilatation [12].

The “revolution” of Dr. Peña and Dr. Levitt, based on their faith, has no scientific justification. There is no evidence that their treatment of “persistent cloaca” improves the function of the urinary system. On the contrary, the destructive effect of their operations can only worsen the function of the pelvic floor organs.

**Diagnostics.** With ARM with one opening on the perineum, it is important to determine the presence or absence of the anal canal. If during abdominal compression gas from the rectum approaches the skin of the perineum, the anal canal ectopy is diagnosed. Perineum perforation allows you to save the function of the fecal retention and defecation. Laparoscopic closure of the fistula can be done after creating the anus of a normal size. If the anal canal is missing, cystourethrography is necessary [13]. Detection of the urethra precludes persistent cloaca. The least traumatic method of the reconstruction is laparoscopic anorectoplasty. Urological and gynecological problems should be eliminated by appropriate specialists.

**3. Analisis of PSARP.**

The authors of the article write: "The incision includes the skin and subcutaneous tissue and separates the parasagittal fibers, muscle complex, and levator muscles in the midline". Such a description of the operation is not permissible. To
understand the importance of each section, surgeons should use anatomical terms.

The anorectal anatomy diagram (Figure 5) shows that in order to detect the rectum, the surgeon must first separate the superficial portion of the EAS from the coccyx (red star). It is assumed that the fixation of EAS to the coccyx contributes to the wide opening of the distal part of the anal canal during defecation [14]. Muscle complex consists 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.

It is further written: "In perineal and vestibular fistulas, the rectum is immediately visible and multiple silk sutures are placed on the rectum to apply uniform traction to facilitate the safe separation from adjacent structures". As shown above, in perineal and vestibular fistulas there is always a functioning anal canal. During PSARP, the anal canal is in a closed state, so it is almost impossible to detect the thin wall of the IAS. During the dissection and traction of the rectum, it is separated from the levator plates and the IAS is imperceptibly cut off from the rectum. The denervated (after transsection of the nerve to levator anii, pudendal nerve, inferior rectal nerve, middle rectal plexus) and devascularized rectum is placed in the wound channel of the pelvic floor muscles, but not in the place where the IAS was located.

4. Anorectal function after PSARP.
1. The intersection of the above nerves interrupts the reflex connections of the rectum with the muscles of the pelvic floor.

2. Cutting off of the superficial part of the EAS from the coccyx and separating the levator plates from the rectum excludes the possibility of a wide opening of the new anal channel during the passage of feces, which sharply increases the resistance to emptying.

3. The posterior sagittal anorectoplasty (PSARP) allowed surgeons easily to view the rectum but does not allow to see the anal canal. In all cases of ARM, regardless of the level (low or high), a PRM dissected and an "anal canal" is created from the rectum. The rectum located at the site of the IAS cannot perform its function because: (1) its function normally differs from the function of the IAS; (2) it is denervated and devascularized; (3) it is located in the wound channel, which heals with inflammation reaction with the formation of fibrous tissue around the gut.

4. The PRM is not involved in the fecal retention, firstly, because its loop is dissected. Secondly, it does not receive information from the denervated rectum.

5. Denervated EAS is in constant contraction since it does not respond to rectal distension with fecal masses. The strength of its contraction is weaker than normal and depends on the degree of its damage during PSARP. For example, forty females with different subtypes of ARM, median age 13 (4–21), were followed up regarding bowel symptoms. During an ultrasound study in 11/17 of the examined females, the rectum did not reach the skin and a subsequent pocket under the skin and superficial EAS was found. The most frequent finding was fragmented IAS. In 11 females diastases in both the deep and superficial components of the EAS were identified. Fecal incontinence (Krickenbeck 1-3) was reported by 30/40 (75%) and constipation by 37/40 (93%) without bowel management. The use of regular bowel management with oral laxatives and/or enemas was reported by 31/40 (78%) with the aim to treat constipation and secondary overflow incontinence [15]. The stitches on the background of devascularization and denervation were the cause of wound dehiscence in 17 (43%) without colostomy [16], which precedes the fibrosis of the tissue and the formation of stenosis.

Thus, instead of a functioning anal canal, a non-functioning canal arises after PSARP. This perineal fistula does not provide normal emptying of the rectum, because of which large amounts of feces accumulate in the rectum, causing
megarectum and megacolon. The more damage to the pelvic floor muscle, the more likely that fecal incontinence will prevail after surgery.

To objectively know the result of the operation, you need to make a barium enema and determine the width of the rectum and the length of the perineal channel.

5. The results of surgical treatment of ARM

The authors argue that: - "Long-term results are good provided there is an accurate anatomical reconstruction and a focus on maximizing of functional results." And they add: - "There appear to be three factors that affect the outcomes in these patients. (1) The type of malformation, (2) sacral development or lack thereof as suggested by the sacral ratio, and (3) spinal abnormalities all have an influence of the potential for bowel control" [1]. There is no scientific evidence in the literature on the effect of congenital spinal pathology on pelvic floor function. Such an assumption was made to justify the poor results of pull-through surgery. But not a single article on this subject has a scientific justification. At the same time, pediatric surgeons do not pay attention to the fact that pull-through surgery, regardless of the approach to the rectum, occurs with denervation of the pelvic organs. The higher the defect, the more extensive denervation.

Studies of newborns (radiographs, ultrasound, CT, or MRI), which are performed only to predict a poor result, make no sense, since they do not change the treatment tactics. Most patients with ARM have a functioning anal canal. Study Hashish et al "showed that stooling patterns are perceived to worsen with age" [17]. "A large proportion of the patients have persistent fecal incontinence, constipation, and sexual problem [18]. Interestingly, these same authors, who perform the “accurate anatomical reconstruction” of the ARM, created a postoperative support program for their patients, in which "Professional and facility charges for all services over the 12-month period totaled $ 3,033,176 "[19].

6. Conclusion

1. Wood and Levitt's article contains misconceptions about the pathological anatomy and physiology of ARM.

2. The described diagnostic methods do not make any sense.

3. The touted PSARP destroys the anal canal in patients with low malformations that make up the majority of ARM.
4. Diagnosis and surgical treatment of persistent cloaca have no scientific justification.

5. Very poor long-term outcomes are explained by the false statement about the absence of an anal canal in these patients and the use of PSARP, which destroys the anal canal.

6. Analysis of the article and literature prove that the preservation of all elements of the anal canal can significantly improve the results of treatment.

Michael D. Levin, nivel70@hotmail.com;

https://www.anorectalmalformations.com

References


