Diagnosis of anorectal malformation in newborns. Literature analysis.

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Recently, many articles have appeared on the advantages of ultrasonography in diagnosing anorectal malformations (ARMs) in newborns compared to radiographic examinations. I analyze the article by Takada et al., which reflects all the recent trends in the diagnosis of ARMs in neonates [1]. This article represents a rare, if not the first, case in which diagnosis and surgical planning are consistent with the Wingspread classification and posterior sagittal anorectoplasty (PSARP) is not used for ARM correction. This choice was made based on a comparison of the results of different approaches to pull-through surgery. The authors emphasize that the classification into low and high ARM types has important clinical significance, as it determines the choice between primary anoplasty and staged correction with a diverting colostomy in the neonatal period. To differentiate between low and high types of ARMs, the pouch-perineum distance upper limit of low type was 10 mm. A postnatal examination was used to make the decision. However, institutional practice has shifted toward repeating PPD measurements immediately before surgery to account for physiological changes. The final diagnosis was established by direct perineal exploration during definitive surgery. An analysis of this method is shown in Figure 1.

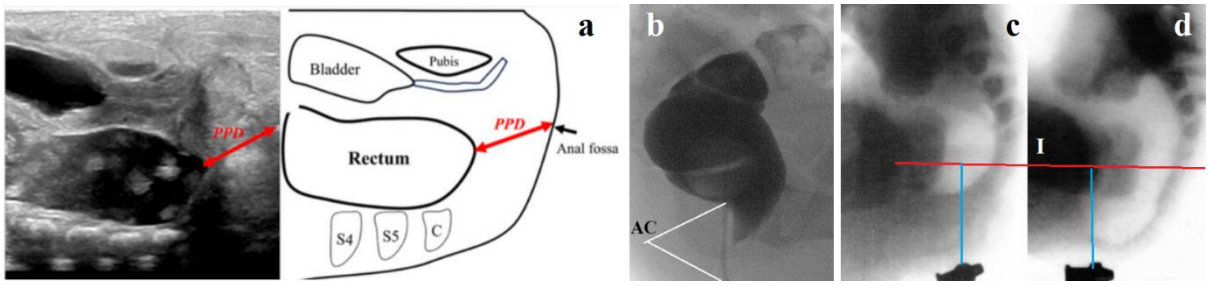

Figure 1. (a) From Takada et al [1]. In this case, a postnatal examination was used. An angular protrusion of the rectum toward the PPD is detected. This is relaxation of the internal anal sphincter (IAS), which is accompanied by contraction of the puborectalis muscle (PRM) and the external anal sphincter (EAS). **(b)** In a healthy child, the anal canal (AC) contracted around the catheter through which the rectum was filled. Anterior to the catheter, contrast medium penetrated into the upper part of the anal canal because of relaxation of the IAS. The posterior wall of the anal canal is pressed against the catheter by the contracted PRM. (**c-d**) Newborn without visible fistula. When the rectal pressure, caused by the volume of gas and meconium raises the rectal pressure to a level that excites the defecation reflex, the levator plates open the anal canal, and the contents of the rectum approaches the subcutaneous tissue opposite the anal

dimple [2,3,4]. Radiographs (c-d) show successive stages of anal canal opening located distal to line (I) (red), drawn at the border between the distal 1/3 and proximal 2/3 of the ischium, which has a pear-shaped form [5]. The true length of the anal canal (blue line) in all full-term newborns is 1.7 cm [2].

The idea that the PPD is a contracted anal canal, the image of which can be fixed at any time during its opening, explains the following facts:

1. Large variation in PPD length: low type - 6.5 (3.5–7.6) mm, high type - 15.0 (14.0–19.0) mm [1].

2. It was shown that the PPD on the next day (mean ± SD, 9.37 ± 4.89 mm; range, 2.1–20.9 mm) was significantly shorter than on the day of birth (15.75 ± 6.67 mm; range, 8.1–37.2 mm; P = .001) [6]. This is because on the next day, additional gas and meconium arrive, leading to increased pressure in the rectum, resulting in an increased incidence of anal canal opening.

3. Therefore, the conclusion about the type of ARM based on a postnatal examination is erroneous.

Pediatric colorectal surgery is part of general medicine. The same muscles, reflexes, and laws operate in children as in adults. Leading pediatric surgeons likely isolated pediatric colorectal surgery from general surgery at some point to strengthen their position among practicing pediatric surgeons. Pediatric colorectal surgery developed without relying on advances in physiological science. A clear example is the article by Wangensteen and Rice, who proposed an invertogram to determine the distance from the rectum to the anal pit. They suggested holding the newborn upside down to allow gas in the rectum to rise, thereby revealing the distance between the rectal wall and the anus. However, in the human body, it is not physical but physiological laws that operate. Movement in the digestive tract occurs through peristalsis. This is described by the "law of the intestine" (Bayliss and Starling), which states: "Excitation at any point of the gut excites contraction above, inhibition below." [7]. Secondly, they write about the presence of the anal canal, but call it the rectum, not considering that the length of the anal canal has long been known from anatomical studies, and the anal canal is in constant contraction in all people, except for a few seconds a day, during defecation. This means that gas from the rectum cannot penetrate the anal canal unless conditions are created to provoke defecation, regardless of body position.

Following Stephens' research, ARMs with visible fistulas were considered low-type ARMs, as they were proven to have an anal canal. For several decades, pediatric surgeons with perineal ectopy have used a cutback procedure, which involved severing the rigid and narrow ectopic anus along with the subcutaneous portion of the EAS [8,9,10,11]. Some surgeons used this procedure for vestibular ectopy [8]. Ninety percent of patients had good functional results after the cutback procedure.

Due to unreliable invertogram results, ARMs without visible fistulas were considered high-type ARM, and these patients underwent pull-through surgery. 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. The gentle and in proper plane pull of the bowel through an undamaged puborectalis muscle gives the possibility to save its function as rectal sphincter and receptor as well. This delicate procedure cannot be performed on newborns without damaging vital structures. Therefore, a colostomy is performed on newborns, and reconstruction is postponed until a later date. [12,13].

In 1982, Peña, describing PSARP, asserted that the role of the PRM is clearly overestimated. He reached this conclusion because he could not identify the PRM during surgery. This suggests that in his practice, he did not operate on patients with ARMs while preserving the PRM. He claimed good results after PSARP, during which he transected the PRM. However, he had no basis for this assertion, as he did not compare his results with those after other surgeries. My analysis of this article revealed several other false statements by Peña, which together raise doubts that he had experience with other surgical methods [4].

Despite Stefans's research on the presence of an anal canal in low-type ARMs, he stated without evidence that the anal canal is absent in all ARMs except rectal atresia [14]. He trained an entire generation of pediatric surgeons who believed his "experience," even though all his statements contradicted reliable scientific research, the laws of physiology, and common sense. In the article by Kraus et al., dedicated to the augmented-pressure distal colostogram, Peña stated, “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…” [15]. From anatomy it is known that there are no muscles around the rectum; muscles are located around the anal canal, and they are in constant contraction,

performing the function of fecal retention. Thus, Peña described absolutely accurate anal canal function in boys with urethral fistulas, which were previously considered high-type ARMs because they allegedly lacked an anal canal. The fact that Peña called the anal canal the rectum does not make it the rectum. But this error leads to the prescription of PSARP or other pull-through surgeries. To understand what happened to these patients, regardless of approach, it is necessary to turn to pathophysiological analysis of PSARP: 1) Approach to the rectum is opened after transecting the puborectalis muscle, which plays an important role in fecal continence; 2) The deep and superficial parts of the external anal sphincter (EAS) are intersect. The subcutaneous part of the EAS, which is 10 times smaller than the other parts and does not play a significant role in fecal continence, remains intact; 3) The internal anal sphincter, which performs 50% of fecal continence, is removed, and the avascular and denervated rectum is inserted in its place. The rectum, unlike the anal canal, plays the role of fecal accumulation and has a different innervation. 4) During dissection of the rectum, the levator plates, which open the anal canal during defecation, are torn away from rectum. This means that when trying to defecate, the rectum at the site of the anal canal does not open and does not reduce resistance to the passage of feces. 5) With extensive dissection of the perineal tissues, numerous invisible nerve connections between the organs responsible for pelvic floor reflexes (anorectal, genital and urological) are severed. Thus, instead of a normally functioning anal canal, a perineal fistula arises. These patients inevitably suffer from chronic constipation, fecal incontinence, urological and sexual problems, which worsen with age.

Holschneider et al, found absence or sharp decrease in ganglion cells in the rectal sac and concluded that the rectal sac cannot be used to correct ARM [16]. Following Peña's example, they called the anal canal the rectal sac, believing that it should have the same innervation as the rectum. They did not know that the fistula (rectal pouch, rectal sac) has all the characteristics of the anal canal, including the absence of an intermuscular plexus [17, 18]. As shown by den Hollander et al., all patients with ARM, regardless of type, have a rectoanal inhibitory reflex, which disappears after correction [19]. How did it happen that Peña'sstatements, contrary to the anatomy and physiology of the anorectal region, became generally accepted "laws"?

In 2005, Peña organized an international conference in the city of Krickenbeck, to which he invited 26 pediatric surgeons. By this time, he had published 42 articles in which he described his experience. Of the 26 invited, 23 participants did not have articles on ARMs, and most of them had no publications at all. Peña had their addresses after they completed his training

course. Three participants (Holschneider, Iwai and Rintala) had 2-3 articles each that were indirectly related to the problems of ARMs, in which they agreed with Peña's ideas [20]. Thus, out of 26 conference participants, 22 (85%) were extras. However, since then the decisions of this conference have been considered the international standard for scientific publications and evaluation of treatment outcomes. This means that by manipulating classification, he secured a ban on the publication of materials contradicting his "experience." The articles, demonstrating that fistulas are an anal canal that should be preserved, were not published in pediatric journals. This created the false belief that all scientists agree with Peña's experience.

Peña never published a single scientific study, but all his statements became widely accepted ideas. For example, all authors began to assert that ARMs arise in the embryonic period because of impaired cloacal development. However, it is known that this long-standing idea is inconsistent with existing scientific evidence. All types of imperforate anus result from impaired development of the exodermal portion of the anal canal, as evidenced by the absence of anal openness. They normally form in the postcloacal period, when the IAS penetrates the anal canal in a craniocaudal direction. Without encountering the exodermal portion of the anal canal, it either penetrates the subcutaneous tissue and skin within the subcutaneous portion of the EAS, creating a narrow, rigid fistula (anal stenosis), or turns forward and upward, creating ectopic anuses from the perineum to the urethra in boys and to the vestibule and vagina in girls. Thus, by the time ectopic anus develops, in both low and high ARMs, a normally functioning anal canal already exists. The canal is blocked only between the IAS and the perineal skin, and its length in newborns is 2 mm [21].

To make sure that all types of imperforate anus have an anal canal, which is 1.7 cm long in all full-term newborns and approximately 1.5 cm in premature infants, it is necessary to create enough pressure in the rectum to open the anal canal, as during defecation. This examination should be performed no earlier than 30 hours after birth to allow sufficient gas and meconium to accumulate in the rectum. If gas is detected at a great distance from the anal dimple during fluoroscopy, pressure should be applied to the infant's abdomen. This is usually sufficient to cause the anal canal to open, which is manifested by the rapid approach of gas to the anal dimple. The approach of gas to the perineum in the restless newborn is a known phenomenon. In the literature, it is mistakenly explained 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 [14,22]. However, from physiology it is known that during an increase in intra-abdominal pressure, the muscles

of the pelvic floor do not descend but rise [23]. This contradiction explains another false claim by Peña, that the final diagnosis is made during surgery. In four newborns consecutively admitted to the Belarusian Center for Pediatric Surgery in Minsk with anorectal malformation without visible fistula, I detected opening of the anal canal during abdominal compression. In each case, the operating surgeons discovered a rectum 2 cm from the anus. This occurred because, under anesthesia, the pressure in the abdominal cavity and rectum decreased, leading to contraction of the anal canal, which was undetectable during dissection [24].

I am writing this analysis, firstly, so that pediatric surgeons and radiologists who performed X-ray and ultrasound studies to determine PPD can verify and confirm that these studies are misleading for all participants in the treatment of newborns with ARMs and lead to destruction of the anal canal. Secondly, it is impossible to change the situation to save the lives and health of children without hard work to correct these mistakes. It is time to abandon blind faith and return to the fold of science.

To preserve the anal canal in newborns with an imperforate anus without a visible fistula, I performed a perforation of the perineum procedure (PPP).

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

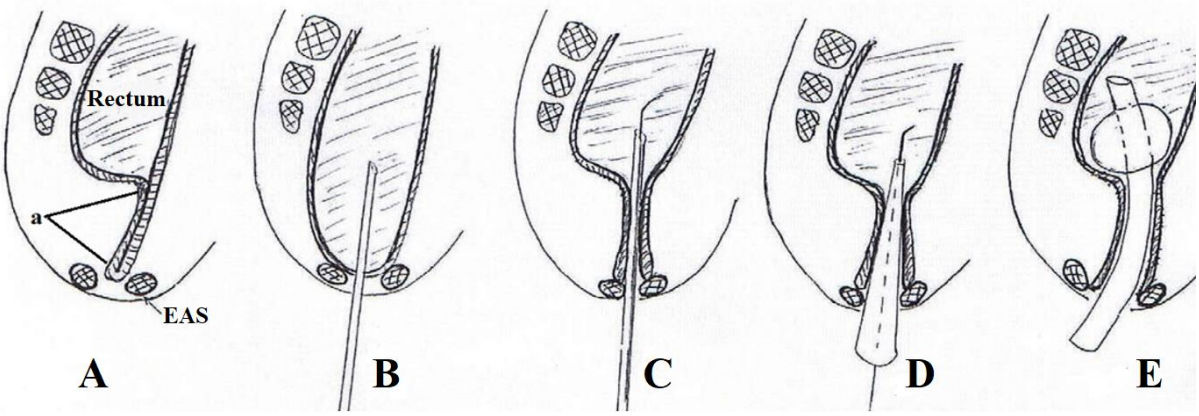

Figure 2. The PPP scheme. Lateral view. (A) The anal canal (a) is closed. The "EAS» is the subcutaneous part of the external anal sphincter. (B). The anal canal is open during abdominal compression. Under fluoroscopy, a needle is inserted into the anal canal. (C). A guidewire for vascular catheterization is inserted into the rectum through a needle. (D). A conical dilator is inserted over a guidewire to widen the newly created channel in the skin and subcutaneous tissue. (E). After removing the dilator, an 8 mm diameter tracheostomy tube is inserted into the rectum through the guidewire. The balloon of the tube is inflated in the rectum to fix the tube in the anal canal.

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

I completed the PPP for four newborns with ARM without visible fistulas. In one of them, ARM was combined with esophageal atresia. He died after correction of the esophagus from aspiration pneumonia. In three patients the postoperative period was uneventful. They were called for examination at the ages of 1.5, 1.9 and 2.2 years, and in each of them the functions of fecal continence and defecation did not differ from the norm [24]. In one case, no gas escape was heard during anal canal perforation. Therefore, doubt arose that the needle was in the intestinal lumen. To confirm the needle was in the rectum, I injected a contrast agent. **(Figure 3).**

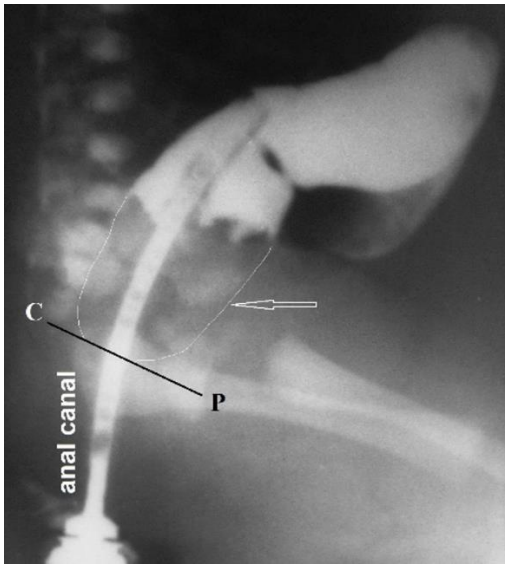

Figure 3. A tube was passed over the guidewire, through which a contrast agent was injected. It turned out that there was a large amount of meconium in the rectum (arrow). Because of this, there was a small volume of gas in the rectum.

Conclusion

1. The Krickenbeck classification is based on false ideas about the pathophysiology of ARMs. All its postulates contradict scientific facts. The truth lies where it is irrefutably proven. Only by following the truth can one help the patient.

2. In all types of imperforate anus, a normally functioning anal canal is present. Therefore, all types except anal stenosis should be referred to as anal ectopy, with indication the location of the ectopic anus. For example, anal ectopy into the urethra.

3. Since the anal canal length in all newborns is 1.5-1.7 cm, conventional diagnostic tests (US, X-ray, etc.) are meaningless.

4. In cases of visible fistulas, the cutback procedure has significant advantages over pull-through surgery, regardless of the approach.

5. In case of invisible fistulas, X-ray examination should be performed no earlier than 30 hours after birth using abdominal compression.

6. Preserving the anal canal can make a patient completely healthy. I have theoretically and practically justified a perineal perforation procedure that preserves the anal canal in cases

where gas penetrates the anal canal during an X-ray examination and does not disappear during compression.

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