Why can't refer to Peña's experience?

Recently, an article by Morandi et al ". Labeling male anorectal malformations: objective evaluation of radiologic imaging before surgery" [1], in which the authors argue that the PC and I lines, as well as the "dove sign", are very useful for better clarifying the anatomy before surgery in boys with anorectal malformations without visible fistula.

What is the pubococcygeal line?

Stephens's research showed that the puborectalis muscle, located between the rectum and the anal canal, is located at the level of the pubococcygeal line (P-C), which runs from the middle (or lower) of the pubic bone to the last coccygeal vertebra. If the distal intestinal wall is located below this line, then we are talking about a low type of ARM. A low type of ARM indicates the presence of an anal canal, which must be preserved to achieve the best functional result [2,3]. This study was the basis for the adoption of the Wingspread classification (1984), in which anal stenosis, perineal and vestibular fistulas were classified as low types [4].

What does the "I" line mean?

The authors of the peer-reviewed article describe "the ischiatic (I) line", traced parallel to the PC line and passing through the lowest visible ischiatic point **(Figure 1),** without citing the authors. This line was proposed by Cremin et al for the case when it is impossible to accurately determine the bony landmarks on the invertogram. They proved that a horizontal line drawn between the distal 1/3 and the proximal 2/3 of the pear-shaped ischium corresponds to the location p-c line [5]. Thus, the line "I" is an analogue of the pubococcygeal line.

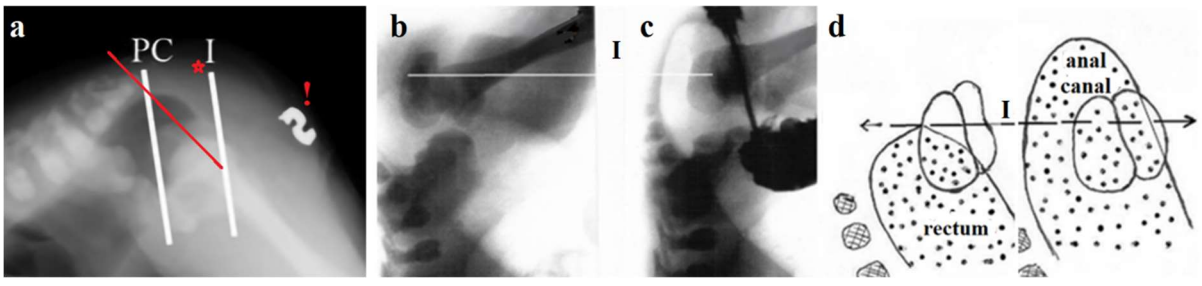

Figure 1. (a) Figure 1 from the article by Morandi et al. [1]. The authors mistakenly drew a line (PC) between the last coccygeal vertebra and the iliac crest. I drew a red line through the lower third of the pear-shaped ischium. This is the "I" line, which corresponds to the p-c line. The gas, is caudally to this line, is in the anal canal during relaxation of the internal anal

sphincter (IAS). The rest of the anal canal is in a closed state. The curved contrast shadow, which probably should be near the anal fossa, is in fact on the thigh. I marked the site of the anal fossa with a red star. **(b-c)** Study of the newborn without visible fistula. **(c)** After 24 hours, in a resting state the blind end of the intestine is at the level of the p-c line, i.e. in the rectum. The surgeon wanted to inject the contrast agent into the rectum but injected it near the rectum. During the straining, when the rectal pressure was increased, the anal canal opened, and the gas approached the skin (see diagram **d**).

In **Figure 2** from the peer-reviewed paper, the authors show the gas in the shape of a bird's beak.

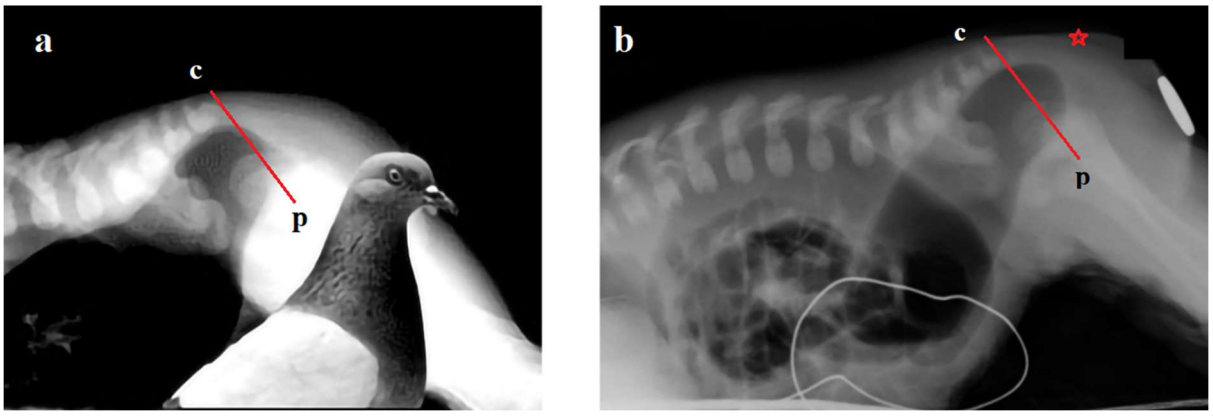

Figure 2. (a-b) Caption from the article: “Pigeon sign” is highlighted. (a) Presence of the “pigeon sign”. **(b)** No “pigeon sign”. I have drawn a red line “I”. It is known that relaxation of the IAS occurs about 18 times per minute. During this time, fecal retention is achieved by contraction of the external anal sphincter (EAS) and the puborectalis muscle (PRM), which pulls the posterior wall of the anal canal forward. This radiographic picture is a manifestation of the anorectal inhibitory reflex [6,7]. **(b)** This radiograph was taken at one of the moments of the anal canal opening.

The same unexplained errors are shown in Figure 3a.

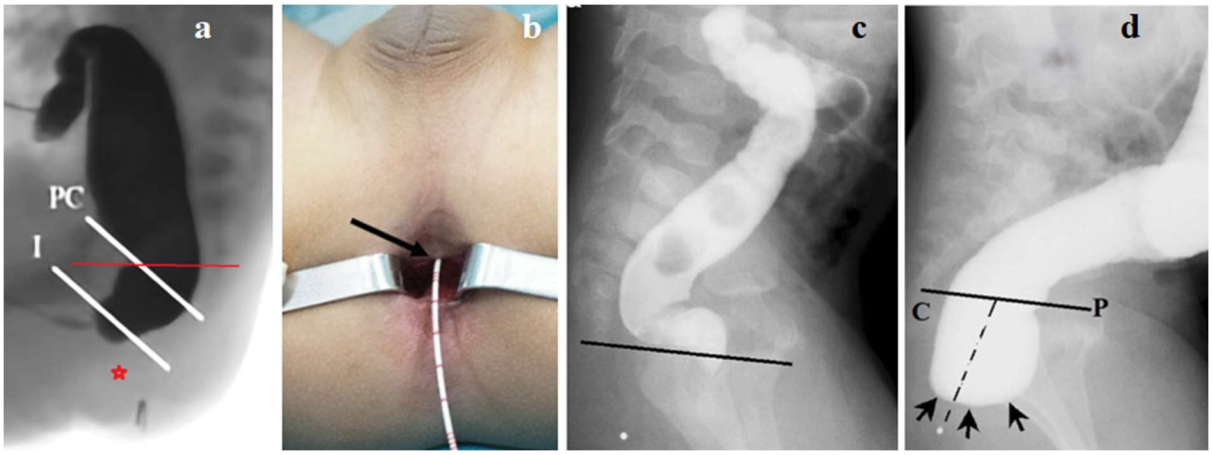

Figure 3. (a) Caption to Figure 3 from the peer-reviewed article: - "This colostogram shows the presence of a fistula with the urinary tract below the I line, consistent with a rectobulbar fistula" [1]. However, the PC line is not horizontal. It is not drawn from the distal coccyx and continues in an unknown direction. The "I" line is not associated with any bony landmarks. I have drawn the p-c line in red from the distal coccygeal vertebra to the pubis, which is at the same level as the head of the femur. I have marked the anal fossa with a red asterisk because it is located between the buttocks close to the wall of the patent anal canal. The distal portion of the bowel located below the p-c line is the anal canal. An ano-urethral fistula is an opening between the anal canal and the urethra.

(b) The photograph a black arrow shows the opening into the urethra from the anal approach [8]. Although the authors state that it was located 1 cm from the anal verge, they did not use any obvious landmarks or instruments for precise measurement.

(c) Radiograph from the article by Kraus et al (Levitt, Peña) on the high pressure colostogram [9]. The horizontal rectal branch is the result of contraction of the PRM. Below the p-c line, the relaxation of the IAS is visible (beak). The remainder of the anal canal is closed. The authors explain that (rectal pouch) "…is closed because of inadequate pressure" [8].

(d) From the same article, with adequate rectal pressure, the opening of the anal canal is visible, the wall of which has approached the marker in the anal dimple. The authors 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…" [9].

Every educated person knows that there are no muscles around the rectum, that the lowest part of the intestine, around which there are muscles that are in constant contraction and prevent

involuntary defecation, is the anal canal. This primitive lie is aimed at justifying posterior sagittal anorectoplasty, which destroys the anal canal. However, the evidence presented above shows that not only ARMs with visible fistulas have an anal canal, but all types of ARMs are ectopia anus. For example, members of the Arm-Net Consortium stated that "According to present knowledge, the “fistula” in ARM represents an ectopic anal canal and should be preserved as far as possible to improve the chance for fecal continence" [10].

The Krickendeck classification, adopted at an international conference in 2005, is not a scientific document in principle. In addition,of the 26 participants invited by Peña to vote, only three (Holschneider, Iwai and Rintala)had 2-3 articles each that were indirectly related to the problems of ARM. For example, Holschneider and histologists (1996) concluded that the recommendation to use the distal rectal pouch and fistula portions in the reconstruction of anorectal malformations should be revised, since abnormal innervation patterns were found in 96% of cases [10]. What Peña's predecessors called the anal canal, Peña, without any research, began to call the rectal pouch. Perhaps this is why Holschneider et al believed that the innervation of the rectal pouch should be the same as in the rectum. However, numerous studies have shown that epithelial and ganglionic distribution was similar in the distal rectal end of ARMs and in a normal anal canal [12,13,14, 15]. All other participants had no publications on ARMs by the time of the conference. Most of them have not published any publications since the conference to this day. Peña deliberately invited pediatric surgeons who were not involved in scientific research. Thus, the division into high and low types disappeared from the classification, since Peña presented PSARP as an ideal method for treating all types of ARM. Since then, all scientific studies have been discontinued or not published, as they contradicted the Krickendeck classification. And articles that contradict scientific studies, but confirm Peña's false concept, are free to be published. For example, Skaba et al (2024) found that A total of 65 (70.7%) patients presented an aganglionic segment in perineal fistula, and 27 patients presented hypoganglionosis. The median length of the resected fistula was 25 mm [16]. Instead of a cutback procedure, which preserves the anal canal, they performed PSARP, destroying the anal canal.

The authors of the peer-reviewed article, following false ideas about the anatomy and physiology of the ARM Peña, using incorrect labeling of the anorectal zone, came to the false conclusion that “ARM was defined as rectobulbar when the rectal pouch was below the I line, rectoprostatic when between PC and I lines, and rectovesical when above the PC line” [1]. All ARMs, except for the true cloaca, are formed in the postcloacal period, when the IAS, moving

in the craniocaudal direction, does not encounter the exodermal rudiment of the anal canal, as evidenced by the absence of the anus. Therefore, the IAS continues to move, shifting forward and upward. This is how ectopy of the anus is formed (perineal, bulbar, prostatic, and bladder-neck). This means that by the time the IAS penetrates the urethra, regardless of the height of penetration, the distal wall of the anal canal is 2-4 mm from the anal fossa [17] (see Figure 3).

Additional claims made by the authors of the peer-reviewed article that are based on Peña's experience but contradict scientific facts.

1. Traditionally, a male patient born with ARM, in the absence of an evident orifice as for perineal fistulas, is first studied with prone cross-table lateral x-ray (CTLxR) after 24–48 h of life. This examination is meaningless, since at rest and with insufficient volume of gas and meconium in the rectum, the anal canal, as in the norm, is in a closed state. In order to open the anal canal, fluoroscopy should be performed no earlier than 30 hours, and abdominal pressure should be increased by compressing the abdominal wall betweenbetween two palms для increase rectal pressure to the threshold level of defecation. Lateral radiograph should be done at the moment of opening the anal canal. This is the only way to be sure of the presence of the anal canal.
2. Peña claims that this is a false lowering of the perineum in response to the baby's tension, since after the pressure decreases, the "rectum" returns to its original place. This explanation does not stand up to criticism, since during tension the pelvic floor does not drop, but rises.
3. The authors refer to Peña's assertion that surgery accurately determines the diagnosis. First, during the operation, intraabdominal and rectal pressure decreases, causing the anal canal to always contract and the IAS to appear as a narrow canal, which pediatric surgeons, ignorant of the anatomy and physiology of the ARM, call a fistula and remove it. Second, Peña stated that during the operation, he could not identify the PRM, so he does not believe that it plays an important role in fecal continence. However, predecessors, knowing the important role of the PRM in fecal continence, discovered of the PRM during operation and pulled out the rectum through its loop. Thus, this statement by Peña also has no scientific support.
4. Functional prognosis is influenced by the type of ARM, the quality of the sacrum, and the presence of spinal defects. Literature analysis shows that the function of fecal continence and defecation depends only on the degree of damage to the anal canal (the higher the ectopia, the more extensive the dissection, the worse the functional results).

In a review article by Levitt et al., it was shown that «At present no solid evidence supports the concept that tethered cord by itself affects the functional prognosis of patients with anorectal malformations» [18]. It is also known that spinal defects do not affect the good functional results of the cutback procedure. Subsequently, Peña et al. without evidence began to argue that the poor results are associated with spinal defects, and not the destructive operation - PSARP.

5. **Aaugmented-pressure distal colostogram**. A water-soluble contrast is injected in the Foley catheter under direct radioscopic control. When the distal pouch is visualized, the contrast injection is continued to fully distend the distal colon and rectum. With adequate pressure, the distal pouch overcomes the strength of the pelvic muscles and assumes a roundshaped profile. In addition, the high pressure will open and highlight the presence of a possible urinary fistula [1]. This recommendation is not feasible because there is no limit (to full dilation). When the abdomen is compressed (described above), a physiological process of opening of the anal canal occurs, which contracts again after the pressure is decreased. The recommended by Peña augmented-pressure distal colostogram [9] occurs with uncontrolled hydrodynamic pressure, which leads to mechanical opening of the anal canal and often causes intestinal ruptures. Therefore, most surgeons, fearing perforation, do not create high pressure and this study turns into a fiction.
6. The authors described 13 (62%) cases that received both CTLxR and high-pressure distal colostogram. The described studies did not diagnose the presence of the anal canal, did not prevent its destruction, and did not influence surgical treatment at all.

The presence of a normally functioning anal canal in most children without a visible fistula, as well as the location of the distal wall of the anal canal in newborns at 2 mm from the anal dimple, allows to perform perforation of the perineum at the time of opening of the anal canal [19]. As shown in the study by Gans et al., an ectopic anus almost always opens into the urethra, but its opening is very small and most often does not function, since the lumen of the fistula is completely closed by fibrous tissue [20]. If there are no signs of meconium in the urine and gas does not disappear from the anal canal during abdominal compression, then the urethral fistula is most likely not functioning. In such cases, perforation of the perineum is indicated, and the functional result will not differ from the norm. If a functioning urethral fistula is suspected, it is advisable to perform a colostomy and perforate the anal canal using an endoscope inserted through the colostomy [19].

Conclusion. Until 1982, studies on the normal anatomy and physiology of the anorectal area, and anorectal malformations had a scientifically demonstrative nature. In 1982, deVries PA, Peña A. published an article. "Posterior sagittal anorectoplasty". Two months later, Peña A, Devries PA published an article. "Posterior sagittal anorectoplasty: important technical considerations and new applications", in which Peña used the name of a respected pediatric surgeon but changed the spelling of his last name. In this article, the number of patients operated by Peña increased by 20. This is the first article that was published outside of Mexico, and it shows the methods Peña uses. From 1982 to 2005, when he organized a conference of pediatric surgeons, he published 42 articles on ARM. The articles claimed remarkable results after PSARP, even though the results were not compared with other methods. It was a massive advertising campaign aimed at convincing pediatric surgeons that PSARP was the ideal operation. To justify resection of the IAS, dissection of the EAS and PRM, separation of the rectum from the levator plates, disruption of the blood supply to the rectum, and dissection of the invisible nerve fibers that provide the reflexes of retention and defecation Peña put forward theoretical concepts that contradicted already known scientific facts. The advertising campaign worked. Even though Peña did not publish a single scientific study, most pediatric surgeons cite his experience.

Followers of Peña, Levitt and Wood in 2024 published one article every 2 months (M.A. Levitt) and one article each month (J. Wood), which represent a continuation of the advertising campaign in support of PSARP to prevent scientific research to improve the treatment of patients with ARM. The sooner pediatric surgeons abandon Peña's practice, the sooner the inevitable revival of ARM science will occur. These children can be healthy.

About classification

The Wingspread classification (1984) defined perineal and vestibular ARMs as low types in which there is an anal canal that must be preserved to obtain better functional results. The cutback procedure was widely used for this purpose.

The Krickenbeck classification (2005) was proposed by Peña and was adopted with the help of specially selected pediatric surgeons who had no experience with ARMs. Despite the known indisputable scientific evidence, they rejected the idea of the existence of an anal canal and recommended performing posterior sagittal anorectoplasty in all cases of ARM, which is supposedly the ideal operation, although in fact it destroys the anal canal and causes severe complications (fecal incontinence, severe chronic constipation, sexual and urological problems).

Since then, despite the resistance of Peña and his followers, it has been established that the anal canal is present in almost all ARMs. And the distal portion of the intestine that was removed under the names of the fistula or rectal pouch is a functioning anal canal.

Based on modern concepts of normal anatomy and physiology of the anorectum, as well as pathological anatomy and physiology of ARM, I propose a classification that facilitates the diagnosis and treatment of ARM **(Table 1).**

Table 1. Classification of anorectal malformations

Females	Congenital stenosis of the anus	**Males**
	Ectopy of the anus:	
Perineal	With visible fistula	Perineal
Vestibular		
Vaginal	Without visible fistula	Bulbar
Wide vagina (low or high)		Prostatic
Narrow vagina (low or high)		Neck - bladder
	All types of anal ectopy with open anal canal	
	Atresia (stenosis) of the anal canal	
	True cloaca	
	Other	

Explanations

1. The names should correspond to the embryology, anatomy and physiology of the defect and not be ambiguous. For example, by “congenital anal stenosis” many pediatric surgeons refer to stenosis of the anal canal. Based on this, they remove the internal anal sphincter and destroy the anal canal. However, the anal canal in these patients is normally developed. The narrow rigid canal in newborns and children of the first year of life occupies only 2-4 mm from the anal canal to the anal dimple and its length depends only on the thickness of the skin and subcutaneous tissue. Simple dissection of stenosis will preserve the anal canal. The correct diagnosis is “Congenital stenosis of the anus”.

2. The names “Rectourethral fistula” or “rectovaginal fistula” et al, erroneously imply that between the rectum and the anal dimple there is a fistula or rectal pouch, which contradicts manometric, radiological, histological and embryological scientific evidence. Based on this

misconception, surgeons destroy the anal canal created by nature instead of preserving it. The correct diagnosis is “Ectopy of the anus”. But not anal ectopia, since the anal canal is in place.

3. Most urethral fistulas are very narrow and do not function. If there are no signs of meconium in the urine and gas does not disappear into the urinary tract during abdominal compression, this means that the fistula is not functioning, so there is no point in distinguishing separately "ARM without fistula". Diagnostic methods allow us to accurately divide fistulas into functioning and non-functioning. The colonography, which is performed at high hydrodynamic pressure, cannot be used, since, firstly, it is dangerous due to the possibility of intestinal perforation. If low pressure is used, it does not bring any benefit. Secondly, high pressure can turn a non-functioning fistula into a functioning one, which leads to more difficult surgical manipulations. A multi-country analysis showed that "The first week mortality proportion was 12.5%, 3.2%, 28.3%, and 18.2% among all, isolated, multiple, and syndromic cases, respectively" [21]. It is obvious that the less surgical trauma, the greater the chances of survival of the newborn. In this sense, the cutback procedure has an advantage over the pull-through, regardless of the access.

4. If women have a urethra and an anal canal, then a narrow vagina is not a common canal, and the pathology is not "Persistent cloaca". This is anovaginal ectopy with a narrow vagina.

5. The defect that was considered atresia or stenosis of the rectum is in the anal canal at 1 cm from the anal verge. It should be called atresia/stenosis of the anal canal. Removal of the membrane is available from the anal approach, because of which the anal canal is preserved, and the operation is better tolerated by patients.

6. The defect that was called "H-type ARM" is different types of ectopy of the anus with an open anal canal. This anal canal is not normal, as some authors write.

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References

1. Morandi A, Maestri F, Ichino M, Pavesi MA, Macchini F, Di Cesare A, Leva E. Labeling male anorectal malformations: objective evaluation of radiologic imaging

before surgery. Front Pediatr. 2023 Aug 3;11:1224620. doi: 10.3389/fped.2023.1224620

2. Stephens FD. Imperforate rectum. A new surgical technique. Med J Australia. 1953;**1**:202.
3. Stephens FD, Smith ED. Pediatric surgery. Pediatr Radiol. (1986) 92(3):200–5. doi: 10.1016/j.suc.2012.03.018
4. Gangopadhyay AN, Pandey V. Anorectal malformations. J Indian Assoc Pediatr Surg. 2015 Jan;20(1):10-5. doi: 10.4103/0971-9261.145438.
5. Cremin BJ, Cywes S, Louw JH. A rational radiological approach to the surgical correction of anorectal anomalies. Surgery. 1972 Jun;71(6):801-6.
6. Levin MD. Anatomy and physiology of anorectum: the hypothesis of fecal retention, and defecation. Pelviperineology 2021;40(1):50-57. DOI: 10.34057/PPj.2021.40.01.008
7. Levin MD. Pathological physiology of the anorectal malformations without visible fistula. A short review. Pelviperineology 2023;42(2):74-79. DOI: 10.34057/PPj.2022.41.02.2021-9-1.
8. Huang X, Chen Y, Pang W, Peng C, Wu D. Transanal fistulectomy for postoperative persistent rectourethral fistula in patients with ARM: is simple resection enough? BMC Surg. 2021 Apr 2;21(1):179. doi: 10.1186/s12893-021-01186-3.
9. Kraus SJ, Levitt MA, Peña A. Augmented-pressure distal colostogram: the most important diagnostic tool for planning definitive surgical repair of anorectal malformations in boys. Pediatr Radiol. (2018) 48(2):258–69. doi: 10.1007/s00247-017-3962-2
10. Amerstorfer EE, Schmiedeke E, Samuk I,et al, Arm-Net Consortium. Clinical Differentiation between a Normal Anus, Anterior Anus, Congenital Anal Stenosis, and Perineal Fistula: Definitions and Consequences-The ARM-Net Consortium Consensus. Children (Basel). 2022 Jun 3;9(6):831. doi: 10.3390/children9060831.
11. Holschneider AM, Ure BM, Pfrommer W, Meier-Ruge W. Innervation patterns of the rectal pouch and fistula in anorectal malformations: a preliminary report. J Pediatr Surg. 1996 Mar;31(3):357-62. doi: 10.1016/s0022-3468(96)90738-1.
12. Duhamel B. Physio-pathology of the internal anal sphincter. Arch Dis Child. 1969 Jun;44(235):377-81.
13. Howard ER, Nixon HH. Internal anal sphincter. Observations on development and mechanism of inhibitory responses in premature infants and children with

Hirschprung's disease. Arch Dis Child. 1968 Oct;43(231):569-78. doi: 10.1136/adc.43.231.569.

14. Weinberg AG. The anorectal myenteric plexus: its relation to hypoganglionosis of the colon. Am J Clin Pathol. 1970 Oct;54(4):637-42. doi: 10.1093/ajcp/54.4.637.
15. Uemura K, Fukuzawa H, Morita K, Okata Y, Yoshida M, Maeda K. Epithelial and ganglionic distribution at the distal rectal end in anorectal malformations: could it play a role in anastomotic adaptation? Pediatr Surg Int. 2021 Feb;37(2):281-286. doi: 10.1007/s00383-020-04786-x.
16. Skaba R, Dotlacil V, Fuccillo P, Rouskova B, Pos L, Rygl M. Perioperative Histologically Controlled Fistula Resection in Patients with Imperforate Anus and Perineal Fistula. Eur J Pediatr Surg. 2024 Oct;34(5):418-422. doi: 10.1055/s-0043-1777101.
17. Levin MD. Embryology of ARM. Hypothesis. chrome-extension://efaidnbmnnnibpcajpcglclefindmkaj/https://www.anorectalmalformations.com/_files/ugd/4d1c1d_df7a24edff4048458ea5a87eb43ceb0c.pdf
18. Levitt MA, Patel M, Rodriguez G, Gaylin DS, Pena A. The tethered spinal cord in patients with anorectal malformations. J Pediatr Surg. 1997 Mar;32(3):462-8. doi: 10.1016/s0022-3468(97)90607-2. PMID: 9094019.
19. Levin MD. Theoretical Basis of New Surgical Tactics for Anorectal Defects without Visible Fistulas. Novosti Khirurgii. 2023. 31 (5); 397-404. (DOI: https://dx.doi.org/10.18484/2305-0047.2023.5.397.
20. Gans SL, Friedman NB, David JS. Congenital Anorectal Anomalies: Changing Concepts in Management. Clin Pediatr (Phila). 1963 Nov;2:605-13. doi: 10.1177/000992286300201105.
21. Kancherla V, Sundar M, Tandaki L, et al. Prevalence and mortality among children with anorectal malformation: A multi-country analysis. Birth Defects Res. 2023 Feb 1;115(3):390-404. doi: 10.1002/bdr2.2129.