Dear Colleagues,

I offer you an analysis of the article by Shandilya et al "Evaluation and management of "low" anorectal malformation in male children: an observational study", published in the Journal Pediatric Surgery International in 2022 [1]. I chose this article for analysis because it reflects all the problems of pediatric colorectal surgery.

First, the article describes two types of ARM (with perineal fistula and without fistula), which do not differ in any way in children of both sexes. Therefore, there is a suspicion that the diagnosis and treatment of these defects in girls was different - which is surprising.

Secondly, in the introduction, all links to articles are not related to scientific research, but only to links to other articles. For example, a clinical article by Lombardi et al describes severe constipation after PSARP. Rather than abandon this destructive operation, they proposed removing 3 cm of the distal colon [2]. They didn't do any research on the sphincter mechanism. Therefore, referring to this article about the presence of a sphincter mechanism is not acceptable. Because of such references, myths are born in the scientific space. For example, Peña and Levitt published an unsubstantiated claim that there is no anal canal in ARM [3]. Although it contradicted scientific research, numerous references to these authors have turned this myth into a generally accepted theory. It took 40 years before the decision of the European consortium appeared: - “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” [4].

Third, the authors state that they "followed the Krickenbeck classification of ARM in this study". This statement has no rational explanation, because scientific work is carried out with the aim of approaching the truth and no classifications can regulate the search for scientists. The aim of this study was "evaluate LARM in male patients, emphasizing the role of various factors on the outcome and follow-up" [1]. However, in the conclusion, the authors did not present anything they had planned, except for meaningless phrases: - "LARM in male patients may have a diverse presentation. The associated anomalies need proper assessment. Awareness may avoid delayed presentation and unwanted complications. When managed by an expert, the condition can be effectively managed. Regular follow-up is important" [1]. This pointless conclusion can complete any article.

Fourth, the merit of the authors of the article is that, in the interests of their patients, they performed the cutback procedure contrary to the

Krickenbeck classification protocol. Thus, they retained all the elements of the anal canal. Their article should have brought to reason those pediatric surgeons who continue to perform PSARP, destroying the anal canal. But due to the poor quality of the article, this did not happen.

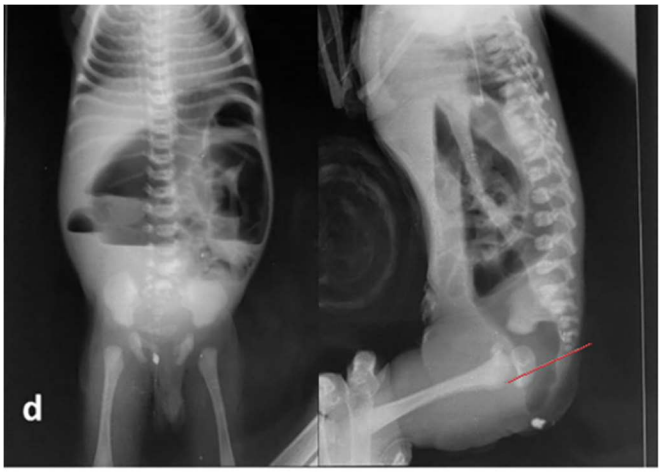
For diagnosing ARM without fistula, they used cross-table, prone position X-ray in lateral position and diagnosed a low-level if "the distal-most gut has at least partially passed through the sphincter mechanism, i.e., if "the distance of most distal bowel gas and the perineal skin marker was less than 1 cm". At the same time, they are "attempted to palpate the rectal pouch at the anal dimple, especially when the baby cried, thereby having the effect of increased abdominal pressure" [1]. The method of the anal canal opening by provoking the anal reflex has not been published anywhere before. However, the authors did not explain its physiological essence and significance. To determine its reliability, it is advisable to compare this method of provocation of high abdominal pressure with the method of the abdominal compression [5].

Without understanding the physiology of the anorectum, the authors make two mistakes that drastically reduce the accuracy of this method. At low pressure in the rectum, the anal canal is in a closed state. Therefore, the gas is in the rectum, i.e., above the pubococcygeal line. In newborns, the true distance between the rectum and the anal fossa is equal to the length of the anal canal (1.7 cm in term, 1.5 cm in premature). The opening of the anal canal occurs at a certain pressure. It is possible only 30 hours after birth, when the volume of meconium and gas in the rectum can provoke this pressure. However, after opening the anal canal, the rectum adapts to this volume and relaxes. The pressure in it decreases, which leads to a contraction of the anal canal, which squeezes the gas into the rectum. To increase the likelihood of relaxation of the anal canal, one should increase rectal pressure by compressing the abdomen, or induce an anal reflex. Irritation of the anal fossa causes contraction of the subcutaneous part of the external sphincter and the defecation reflex (relaxation of the superficial and deep parts of the external anal sphincter, as well as the puborectalis muscle, with simultaneous contraction of the rectum. So, some animals (cats) lick the anus of newborn cubs to cause a defecation reflex. Thus, to detect the opening of the anal canal, it is necessary: (a) to carry out the study no earlier than 30 hours after birth,) b) to take a picture at the moment of opening of the anal canal during fluoroscopy.

It is known that the sphincter complex consists of the three parts of the external anal sphincter, and puborectalis muscle. Each of them has a different attachment, different nervous regulation, and a different function.

Therefore, when stimulating the contraction of the subcutaneous portion of the external sphincter, one cannot call it a sphincter complex, especially since at that time other muscles are not contracted but relaxed.

The opening of the anal canal occurs within a few seconds, and the radiograph takes place within milliseconds. Therefore, different positions of the distal wall of the anal canal can be recorded on the radiograph: from complete closure to varying degrees of opening. In fact, with full opening of the anal canal, its distal wall in newborns is located 2 mm from the anal dimple. Between it and the marker is only the skin and subcutaneous tissue. Thus, the boundary between the lower and high types of ARM cannot be 1 cm since the length of the anal canal in full-term newborns is 1.7 cm [6]. This can be seen in one of the radiographs given in the article.



fistula. **d** Cross-table X-ray in a prone position of the same patient showing gas shadow within 1 cm of perineal skin marker

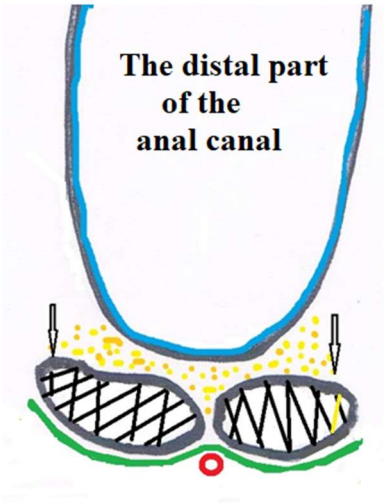


Figure d. In a newborn with ARM without a fistula, with late admission, in a vertical position, an x-ray picture of intestinal obstruction is visible. The wide fluid levels in the colon are indicative of high intra-intestinal pressure. In a horizontal position on the side, the gas penetrated the open anal canal (caudal to the pubococcygeal line - in red). It is located 2 mm from the marker in the anal fossa. In the diagram of the distal anal canal, the subcutaneous part of the external sphincter (arrows) is located under the skin (green). Between the internal anal sphincter (blue) and the subcutaneous part of the external sphincter is fatty tissue. Contrasting marker in the anal dimple (red ring).

It can be said without exaggeration that all ARMs without a fistula are of the low type. And the idea that there may be high types, i.e., without an anal canal, is due to erroneous X-ray examination, when radiographs are

taken before 30 hours after birth, or are made at the moment when the anal canal is closed or not fully opened.

At the beginning of embryological development, the internal anal sphincter migrates caudally within the sphincters as normal. However, without encountering the exogenous anlage of the anal canal on its way, it reaches the subcutaneous tissue and either stops, forming an ARM without a fistula, or penetrates the subcutaneous part of the external sphincter in the form of a congenital rigid stenosis, or shifts anteriorly and opens with a rigid fistula to the perineum, into vestibule or urethra. However, in any of these options, during the anal canal opening, its caudal wall is located opposite the anal dimple at a distance equal to the thickness of the skin and subcutaneous tissue.

As can be seen from a typical X-ray examination, the subcutaneous portion of the external anal sphincter is located under the skin in the gap between the blindly ending internal anal sphincter and the skin. Its thickness is not more than 2 mm, which is less than one tenth the length of other sphincters. This explains the fact known in the literature that the cutback procedure, in which an incision is made from the ectopic opening in the perineum to the middle of the anal fossa, leads to good results. If the operation is done in a timely manner, then despite the intersection of the ring of the subcutaneous part of the external sphincter, fecal retention and defecation are normal in such children.

However, with late presentation, retention of feces over the narrow, rigid ring leads to the development of a megarectum and secondary damage to the pelvic floor muscles, including the puborectalis muscle. This is evidenced by the case described in the peer-reviewed article.

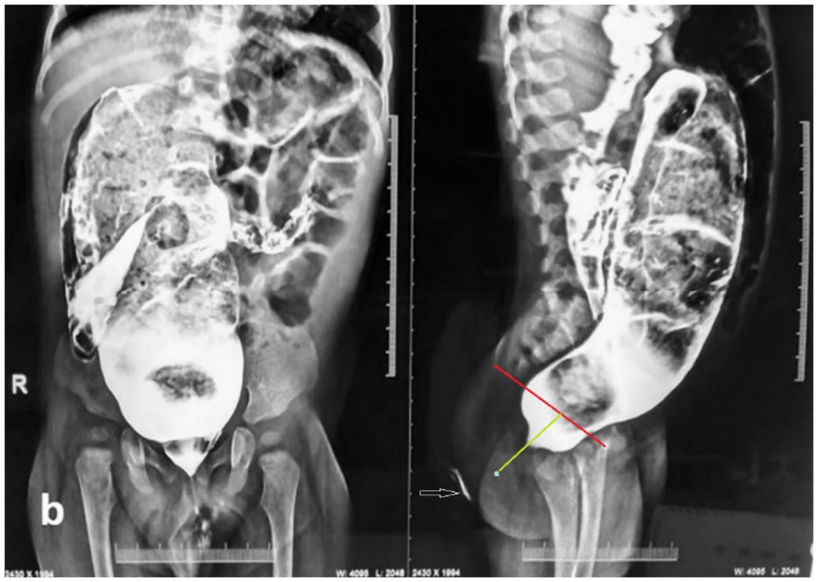


Figure b. “Delayed presentation of a patient of perineal fistula. Watersoluble contrast enema of the same patient showing gross colonic dilation” (caption from article). Judging by the development of the pelvis and joints, this boy is about three years old. On the lateral radiograph, the arrow shows the artifact, which is located outside the buttocks. With a blue dot, I marked the approximate location of the anal fossa, which is located 2.8 cm from the pubococcygeal line (red line). The normal anal canal is located between the anal fossa and the pubococcygeal line (green line). However, the upper half of the anal canal does not work to the feces retention, as the pelvic floor muscles, including the puborectalis muscle, are stretched and weakened by wide fecal masses, which the contracting rectum pushes and cannot push through. Thus, this patient developed megarectum, and descending perineum syndrome. Therefore, despite a well-performed cutback procedure, this patient will have chronic constipation and encopresis.

Obviously, to evaluate the effectiveness of the cutback procedure, it was necessary to study the long-term results of patients where the operation was performed before the development of megacolon, because unsuccessful results in late cases are due to secondary damage of the rectum, and anal canal due to stenosis of the ectopic anus.

It is impossible to evaluate the results of treatment for the following reasons:

1. Because the authors describe complications (constipation, soiling), regardless of the time of operation and type of operation.

2. Since the results of treatment 5 years after surgery do not reflect the full picture, which, for example, after PSARP worsens at a later age, lasts a lifetime, and requires constant monitoring, repeated surgery, antegrade enema, etc.

3. Impossible to assess the degree of chronic constipation without objective studies. In order to accurately know the state of the anorectal zone and take the necessary therapeutic measures in a timely manner, it is necessary to know the exact width of the rectum, which in its size forms the diameter of the stool, as well as the maximal throughput of the anal canal. If the width of the rectum is greater than the maximum possible width of the anal canal, this discrimination will only increase with age and lead to the disability of the child. The width of the rectum and the degree of megacolon can only be measured with a barium enema [6], and the width of the anal canal by the size of the Hegar dilator.

Young children have a chance to become practically healthy if doctors (parents) take measures for a timely bowel movement for a long time to prevent further expansion of the rectum. With the age of the child, the diameter of the anal canal increases, and a situation may occur when the width of the anal canal becomes the same as the width of the rectum.

Fifth, the authors write that "are attempted to palpate the rectal pouch at the anal dimple", this indicates that they do not understand that they are talking about the anal canal. The rectal pouch is something that does not exist, the physiology of which is not known, while the anal canal has a certain length, known components and obeys known reflexes [7]. This misunderstanding explains the following contradiction: only low-type ARMs were selected for the article, i.e., where "gut has at least partially passed through the sphincter mechanism". How did patients appear in the article who had fistula was not within the muscle complex»? [1]. Why was limited anterosagittal anorectoplasty performed on them?

In these 14 patients, without a visible fistula, stimulation of an anal reflex and high abdominal pressure during x-ray study caused opening of the anal canal. Therefore, gas entered the anal canal and approach the marker in the anal dimple. This was the basis for the diagnosis of low-type ARM. During the operation under anesthesia, when the pressure in the rectum was low, the anal canal was in a closed state and therefore was not found during the operation.

In these patients, the results of treatment cannot be good, because the release of the intestine leads to the intersection of invisible nerve connections, without which the reflex reaction of all sphincters is impossible. Devascularization of the intestine, its tension and suturing cause an inflammatory reaction and the formation of sclerotic tissue, leading to re-stenosis. Surgeons differ from tailors by understanding of these processes. In order not to destroy the anal canal, it is necessary to enter the needle into it during its opening in fluorography. And in order not to cause inflammation and stenosis, it is necessary to insert a tracheostomy tube into the rectum for 7-10 days. A inflated balloon in the rectum will fix it in this place and ensure the evacuation of feces [8].

Conclusion. I cannot imagine that such an article could be accepted for consideration in a scientific journal. This article does not carry any scientific information because the authors did not use scientific tools. It is based on false ideas about the anatomy, physiology, embryology, and pathology of the anorectum. Her methodology defies common sense. It looks so primitive that I suspected that the reviewer forced the authors of

the article to accept his corrections if they want the article to be published. I contacted the authors of the article by e-mail so that they would send me the version of the article that I assumed they sent to the editor. But I no received response from them.

This article has been rated by at least two reviewers. Sane people could not recommend this article for publication. At the final stage, the editor had to read it. This is clear evidence of the terrible state of pediatric colorectal surgery, because of the false teachings of Albert Peña, who preached his experience instead of scientific research. He created a mafia structure of reviewers in collaboration with the editors of pediatric journals. As a result of their criminal activities, children's colorectal surgery as a science was destroyed, and for almost 40 years, children born with ARM were subjected to an operation that destroys the anal canal and urinary system, including patients with the so-called cloaca. Because of this, in children who might be healthy after a simple operation, most deceived pediatric surgeons produce PSARP, which destroys the anal canal and disables the children for life. It is impossible to change anything for the better without destroying this criminal system. It is impossible to prevent this in the future without punishing the perpetrators.

References

1. Shandilya G, Pandey A, Pant N, et al. Evaluation and management of "low" anorectal malformation in male children: an observational study. Pediatr Surg Int. 2022 Feb;38(2):337-343. doi: 10.1007/s00383-021-05035-5.
2. Lombardi L, Bruder E, Caravaggi F, et al. Abnormalities in "low" anorectal malformations (ARMs) and functional results resecting the distal 3 cm. J Pediatr Surg. 2013 Jun;48(6):1294-300. doi: 10.1016/j.jpedsurg.2013.03.026.
3. Levitt MA, Peña A. Anorectal malformations. Orphanet J Rare Dis. 2007 Jul 26;2:33. doi: 10.1186/1750-1172-2-33.
4. Amerstorfer EE, Schmiedeke E, Samuk I, et al. 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.
5. Levin MD, Averin VI, Degtyarev Y.G. Pathological physiology of anorectal malformations (ARM) without visible fistulas. Review. Novosti Chirurgii (Belarus) 2022; 30(3): 105-12. DOI: 10.18484/2305-0047.2022.3.298.

6. Levin MD. Radiological anatomy of the colon and rectum in children. Gastroenterology & Hepatology. 2019; 10 (2):82-6. Open access.
7. 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
8. Levin MD. Anorectal malformations. https://www.anorectalmalformations.com/_files/ugd/4d1c1d_0961be5f16f34858bd226cc847191b83.pdf

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