Dear colleagues!

I propose an analysis of the article by Agarwal et al “Rethinking Fistula Preservation in Anorectal Malformation Surgery: A Histopathological Perspective”, published in the February issue of the European Journal of Pediatric Surgery [1]. In it, the authors presented a histological study of 0.5 to 2.0 cm of the most distal part of the rectal pouch that was resected in 65 boys with anorectal malformation (ARM) with urethral fistulas. They did not find the internal anal sphincter. Signs of inflammation and fibrosis were found in the mucosa, and adequate ganglion cells were seen only in 6 (25%) samples. Therefore, they concluded that the rectal pouch does not have normal features of the anal canal and should not be preserved during ARM reconstruction. The topic of the article is of crucial importance for children with anorectal malformations (ARMs), because depending on ideas about the anatomy and physiology of the defect, surgeons choose the treatment tactics.

Based on the scientific research of Stephens (1953) it was established that if the blind end of the intestine is located caudal to the pubococcygeal line, this indicates the presence of an anal canal, which must be preserved to obtain a good functional result [2]. Peña, without scientific justification, began to argue that in ARMs the anal canal is absent. This statement justified the excision of the internal anal sphincter (IAS) and the reduction of the bloodless and denervated rectum in its place during posterior sagittal anorectoplasty (PSARP). Since then, the distal part of the intestine in ARMs has been called a rectal pouch or fistula [3]. That this substitution had no scientific meaning is proven by the article by Peña et al., which states the following: - “It is extremely important in this regard to understand that the lowest part of the rectum is usually collapsed due to the muscle tone of the funnel-like striated muscle mechanism that surrounds the rectum in 90% of cases…” [4]. That article talked about augmented-pressure distal colostogram in patients with urethral fistulas. From anatomy it is known that there are no muscles around the rectum. The part of the intestine that is permanently closed at low rectal pressure due to the contraction of the IAS is the anal canal. Contraction of the anal canal prevents the leakage of intestinal contents located in the rectum. At high pressure, as is normal, the anal canal opens, and the contrast agent approaches the mark glued to the anal dimple. Why do the authors of the peer-reviewed article propose removing the distal part of the intestine, which functions as the anal canal, and replacing it with the rectum, which accumulates feces and expels them during peristalsis? [5].

The only argument that supposedly justified the removal of the "fistula" was histological studies of the anal canal wall, which revealed pathological changes in the form of inflammatory and fibrous changes, as well as the absence of ganglionic cells or hypoanagliosis {1, 6,7,8,13,14,16,17}. These studies, as in the reviewed article, were made with serious violations of scientific methodology. Firstly, beginning with the work of Duhamel (1969) and his associates, it was proven that the normal anal canal, unlike other parts of the digestive tract, does not have an intermuscular nerve plexus [6]. This was confirmed by other researchers in children {3,11}, and in an experiment on neonatal pigs with ARM {4, 18 (All references in this paragraph (except Duhamel) are taken from the article under discussion).

The authors who claimed that the rectal sac (i.e. anal canal) was pathological did not compare the histological findings in the “rectal sac” with the condition of the normal anal canal but mistakenly believed that in the normal anal canal the intermuscular ganglion plexus should be the same as in the rectum. For example, Xiao et al., unlike other authors, compared the results of studies of the remote "fistula" with the control group. However, "The tissue specimens were the rectum that 2 to 4 cm above the dentate line in the control group" [7]. In these cases, the biopsy was taken above the anal canal, the length of which in adults is 3.1 – 3.9 (3.43 ± 0.10 cm) [8]. Secondly, these authors examined the wall of the IAS removed at different times after birth, since in the case of invisible fistulas, a colostomy is first performed. Inflammatory and fibrous changes in the anal canal wall, which are registered several months after birth, are the result of stenosis of the ectopic opening and dilation of the rectum. These are acquired complications that are not observed in newborns [9,10], and which can be prevented by early intervention. Thirdly, any intestinal structure that has a mucous membrane has a muscular layer, which, which, as shown above, is in a contracted state. If the authors of the peer-reviewed article did not recognize it, then either they chopped up tissues that they did not need in parts, or did not know that the thickness of the functioning IAS in infants is less than 1 mm. This figure is approximate, since in the literature there is a statement about the thickness of the anal smooth muscle of adult men. It is equal to 3 ± 0.7 mm and includes both the circular (IAS) and longitudinal layers [11].

Physiological studies prove that the distal part of the intestine in ARMs functions as an anal canal, in low (visible) fistulas, and in high (invisible) fistulas. This is evidenced by the basal pressure characteristic of a normal anal canal, as well as the presence of a positive rectoanal inhibitory reflex [12,13,14]. X-ray studies also prove the presence of a normally functioning anal canal (**Figure 1**).

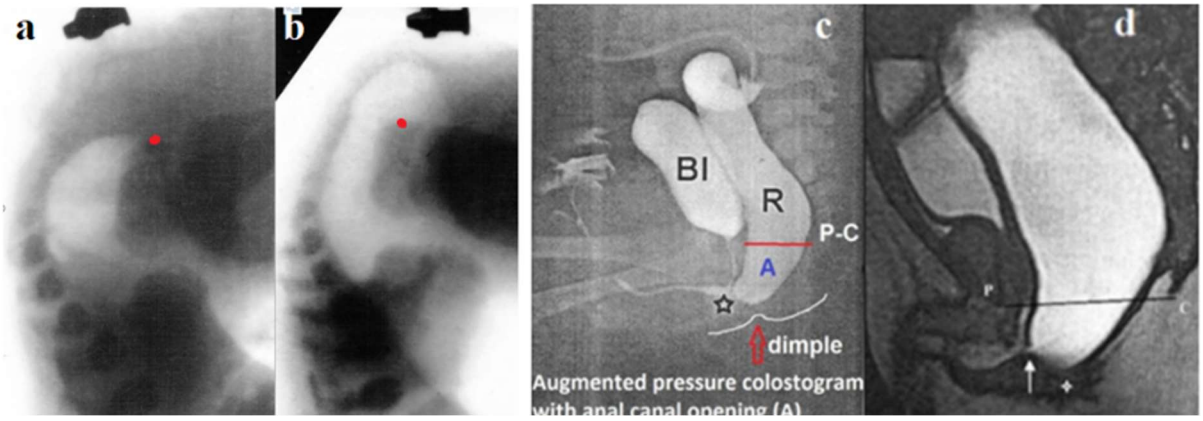

Figure 1. Radiologic studies in males with ARMs without visible fistulas. **(a).** On the radiograph of a newborn 30 hours after birth, the gas reaches the caudal part of the ischium (red dot), **(b).** In the same patient, during abdominal compression, the gas penetrated the anal canal and approached the contrast marker glued to the anal dimple. **(c, d)** Augmented-pressure distal colostograms: (c-X-ray) and (d-MRI). In both cases, under the high pressure, the anal canal (caudal to pubococcygeal line – red) opened, and its caudal wall approached the anal dimple.

The authors' reference to the article by Lombardi et al. is incorrect for the following reasons. The article by Lombardi et al. showed that the use of IAS in low types of PSARP leads to severe constipation not less than 61.4%. During histological examination of the resected 3 cm of the "fistula", they found pathological changes, on the basis of which they considered it necessary to remove the distal 3 cm. Like other authors, they did not know that IAS does not have an intermuscular nerve plexus, and the acquired changes in its wall were accepted as its congenital pathology. In the series of 42 patients followed up after removal of 3 cm of the "fistula" at least after 3 years of age, 40 cases (95.2%) showed postoperative good continence without the use of laxatives [15]. This triumphant picture contradicts the data of Levitt et al., who showed that after PSARP the morbidity of constipation included fecal impaction, megacolon, incontinence and performance of unneeded surgery, regardless of the height of the ARMs [16]. According to Senström et al., among adolescent and adult females with different types of ARMs, fecal incontinence is reported by 40–67% and lack of voluntary bowel control by 15–30% [17]. During ultrasound examination of the neoanus, the most frequent finding was fragmented IAS. In some females, diastases in both the deep and superficial component of the EAS were identified. A majority of the patients had a gap of >5mm from the rectum to the skin anteriorly where also a lack of tissue under the superficial EAS and the skin was found [17]. These data, firstly, indicate a significant percentage of fecal incontinence and chronic

constipation in the late period after PSARP. Secondly, the release of the "fistula" from the surrounding tissues leads to damage to both the IAS and the EAS. It should also be considered that fecal incontinence is largely due to the intersection of the puborectalis muscle. In addition, constipation occurs due to the release of the rectum from the surrounding tissues, because of which it is torn away from the levator plates. After this, this muscle mass does not open the neoanus when trying to defecate, which leads to a sharp resistance to the movement of feces outward. Anorectal surgery with preservation of the IAS, which also includes limited PSARP, leads to chronic constipation because the IAS, isolated from the surrounding tissues, loses blood supply and nervous regulation. When stretched, it ruptures and hemorrhages. It loses reflex connection with other muscles. Preservation of the anal canal differs from the use of IAS in that the anal canal remains intact and therefore functions normally. Simple dissection of the narrow ectopic anus (cutback procedure), which ensures normal emptying of the rectum, never leads to fecal incontinence, and constipation, which can occur if megacolon has already developed by the time of surgery, disappears over time [18,19,20]. The remarkable results after the cutback procedure provide irrefutable evidence that the distal portion of the intestine between the rectum and the subcutaneous fat tissue above the anal dimple is a normally functioning anal canal.

Thus, at low rectal pressure the anal canal is in a contracted state, preventing involuntary defecation. And when the pressure in the rectum reaches the level that causes the defecation reflex, the anal canal opens wide to reduce the resistance to the passage of feces. This is how the normal anal canal functions. The rectum cannot replace the IAS, since it performs the function of storing feces, and relaxes, adapting to its volume [5]. The evidence described above are consistent with a recent statement by pediatric surgeons from the ARM-Net Consortium: “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” [21].

The above evidence allows us to compare them with the statements based on Peña's "experience".

1. Levitt and Peña's statement about the absence of sensitivity in the rectal pouch [22] is false, since the presence of the rectoanal inhibitory reflex, its opening under high pressure in the rectum, and normal functioning after the cutback procedure, indicate the presence of a normal anal canal, and therefore normal sensitivity of its wall.

2. Peña's statement that since he did not find the puborectalis muscle (PRM) during the operation, it cannot be significant for fecal continence, is false for the following reasons: (1). It contradicts common sense, since it does not follow the reasoning - there is no logic; (2). All surgeons before him, starting with Stephens, found the PRM from the sacrococcygeal approach to passing the rectum inside its loop [23]. This suggests that Peña's experience is not reliable, or he lied to justify crossing the PRM during PSARP. (3) This claim by Peña is refuted by numerous studies by physiologists who have shown that the PRM plays an important role in fecal continence [24,25].

From 1970 to 2020, Peña has published the most articles (64) on ARM, followed by Levitt (53). This list, published in the journal "Chldren", contains only 10 most productive authors publishing on ARM [26]. All of them are Peña's supporters, because other authors are not published, if their opinion do not correspond to Peña's experience. In all his articles, Peña shares his experience, but none of them contain any scientific studies or references to them. For 5 decades, no scientific studies have been published on the anatomy and physiology of the anorectal zone, as well as on the pathological physiology of ARM, since PSARP was and still is considered the ideal operation. There have been long-standing discussions in journals about which of the pull through operations is better: from the posterior sagittal approach, the anterior sagittal approach, or using laparoscopy. Since all these operations destroy the anal canal, the difference in functional results between them is insignificant. The problem is that all the authors are obsessed with surgical skill and perceive Peña's experience on the anatomy and physiology of the ARM as something scientific. However, as shown above, this is not the case. I propose a theoretical justification for a new surgical tactic for ARM with invisible fistulas [27].

Conclusion Children with ARMs have a normally functioning anal canal, which must be preserved. The results described in the peer-reviewed article were obtained with significant methodological errors. The terminal portion of the intestine that functions as the anal canal should be compared with the normal anal canal, not with the rectum. To exclude the influence of surgical dissection and secondary changes associated with stenosis of the ectopic opening, the study should be performed soon after birth and before surgery. If this is not available, the study as in this case was meaningless. Therefore, the authors' conclusion contradicts the known scientific data.

Several generations of pediatric surgeons grew up on a false idea of ARMs. Ignorant of the anatomy and physiology of the anorectum and, believing Peña’s «experience", they were

destroying the anal canal. They were brought up to ignore scientific research as the main evidence of truth. They are unfamiliar with the methodology of science, i.e., the logic of scientific analysis. Now they unite to defend their faith. The truth will ultimately prevail.

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