

Dear colleagues,

I suggest you analyze the article Li L, et al. "Normal Anorectal Musculatures and Changes in Anorectal Malformation", published in the journal "Pediatric Surgery International" in January 2020 [1]. Very true the statement of the authors that "Understanding the anorectal musculatures is crucial for surgical correction of anorectal malformations (ARM) [1]. Unfortunately, all other statements require detailed analysis.

Introduction.

The authors of the article claim that Stepfens "...firstly described that the puborectalis constituted the only potential sphincter available for continence following pull-through operation..." The fact that puborectalis muscle (PRM) is an important element in stool retention has been known for a long time. Stepfens showed for the first time that PRM located between the rectum and the anal canal and that its upper edge corresponds to the location of the pubococcygeal line (P-C line). Based on this study, ARMs were divided into low type if the distal gut is below the P-C line, intermediate type if it was at the level of the line, and high type if the gut is above the P-C line. It was meant that with a low type of ARM there is an anal canal that should have been preserved in order to get the best functional result [2].

Further, the authors of the article claim: "In 1982, Alberto Pena highlighted the importance of muscle complex, which was defined by a vertical group of striated muscle fibers between the levator musculature and the fibers about the anal dimple. According to this theory, he proposed the posterior sagittal anorectoplasty (PSARP)". How can it be called a "theory" the fact that all 3 parts of the external anal sphincter (EAS) were called the muscular complex? In fact, in this article, he stated that during operating on patients with an ARM through the posterior sagittal approach, he could not identify PRM, and therefore he sees

no reason to consider PRM as a sphincter, as previously thought [3]. This is an unsubstantiated statement, which contradicts scientific facts, was the rationale for pull-through operation through the posterior sagittal approach, accompanied by the intersection of the PRM. At the end of this article was a discussion by leading pediatric surgeons who have expressed doubt or disapproval of the proposed operation. However, since then, for the last 3 decades, PSARP has become mainly of surgical technique, although it, unlike other pull-through methods, damages the PRM. The reason for the success of the method is that since then scientific discussions have been suppressed.

The authors of the article apparently decided that before them no one had studied the muscles of the pelvic floor in ARM. In any case, the article does not contain a single reference to such studies.

Material and methods

“Pelves of 50 neonates died of ARM-unrelated disease and 16 patients with anorectal malformations (8 high, 5 intermediate, and 3 low ARMs) were dissected and sectioned” [1]. This work is difficult to analyze because there is no important information about patients with ARM: (a) at what age and from what reason did the children die? (b) before or after surgery? (c) on what basis the ARM type was established. The article presents a histological examination, which is interpreted by pediatric surgeons. I am not able to evaluate the quality of histological examination. The conclusions reached by the authors cause serious objections.

In conclusion, it states: "In ARM, the IAST and the inner longitudinal muscular fibers of the rectum is absent, the LMT only consists of the longitudinal striated muscle from the levator ani muscle. The LMT in ARM can be divided into the pelvic LMT and the perineal LMT. The pelvic LMT is displaced anteriorly just posterior to the neck of bladder and posterior urethra in high ARM or to the terminal rectal pouch and rectobulbar fistula in intermediate ARM. The perineal

LMT is fused to form a vertical column in both high and intermediate ARM. In ARM, the LMT is a closed muscular tube, which could be possibly dilated to widen leading the rectum pull-through to establish the normal anorectal function” [1]. The authors use anatomical designations that are not accepted in anatomy studies, and therefore are not always understood. For example, called the internal sphincter tube (IAST), they probably mean the internal anal sphincter (IAS). A longitudinal muscle tube (LMT) may correspond to a longitudinal muscular layer, which interposed between the IAS and the external anal sphincter (EAS). The longitudinal anal muscle is mainly composed of outer striated muscle fibers and small numbers of inner smooth muscle fibers [4]. Interesting, the authors describe a typical picture of the longitudinal anal muscle but did not notice IAS and EAS. It is completely not clear what the authors mean by the term “rectal pouch”, where it is located and how it differs from the rectum.

Children with ARM are different from healthy ones, but the conclusions cited by the authors of the peer-reviewed article contradict the anatomical studies of other authors, the results of a study of the pathological physiology of ARM, as well as the functional results of operations, which preserve the anal canal.

Anatomical studies:

(1) Lin and Chen described fourteen patients with a rectourogenital fistula which “... were treated with posterior sagittal anorectoplasty using the fistula end as the neoanus (internal sphincter-saving)” [5].

(2) In the article AbouZeid and Mohammad [6] (the mid-sagittal T2WI), a closed anal canal is seen in all boys with low ARM (**Figure 1**).

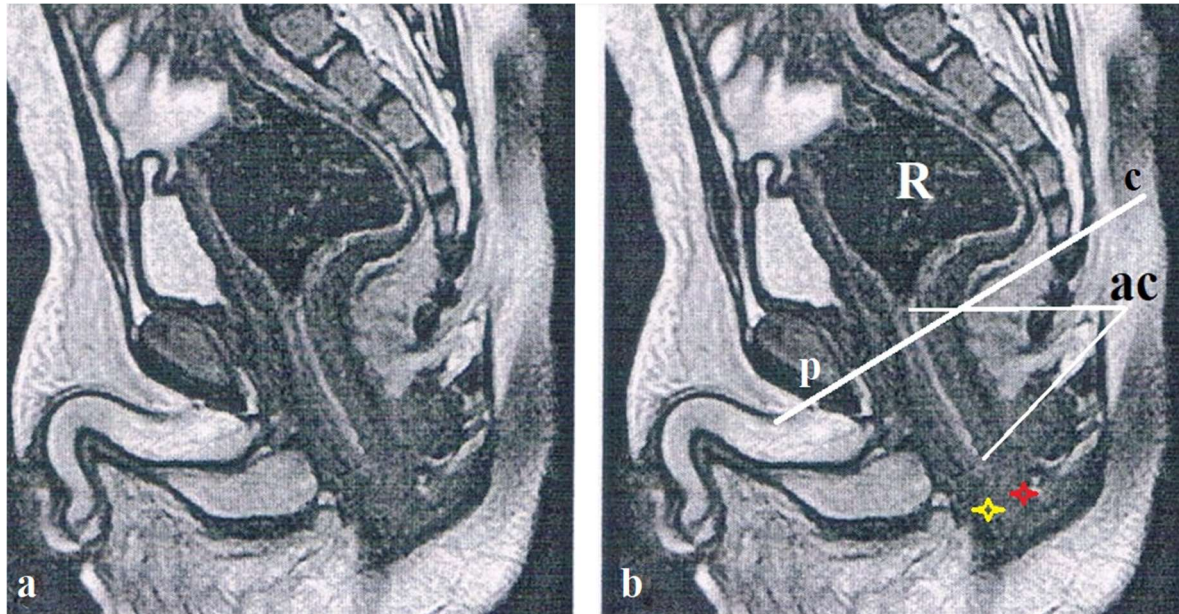


Figure 1. Demonstration of the pelvic MRI anatomy at a male with perineal fistula (a), and its a circuit (b). The anal canal (ac) is caudal to the rectum (R) and pubococcygeal line (p-c line). The mucous layer of the anal canal in the form of a white line is visible in its center. The internal anal sphincter and the longitudinal muscle layer are around it. A red asterisk corresponds to the location of the anal dimple. The yellow asterisk corresponds to the location of the fistulous opening.

(3) “In the animal models a clinically important discovery has been the character of the rectourogenital or perineal communication; the fistulous communication is actually an ectopic anus. This ectopic anus has the characteristics of normal anal canal including a distal zone of transitional epithelium, anal glands, and the internal anal sphincter...” [7].

Pathological physiology of the ARM.

1) In infants with visible fistulas (perineal, vestibular), there is normal fecal continence, and if the ectopic opening is wide enough and does not prevent defecation, the defect can be detected by chance. During an X-ray examination after the administration of a contrast medium through the fistula into the rectum, a long channel is determined, closed around the catheter. The walls of this channel are in constant contraction and do not pass the contrast medium from the rectum

to the outside. The length of this channel is equal to the length of the normal anal canal (**Figure 2**).

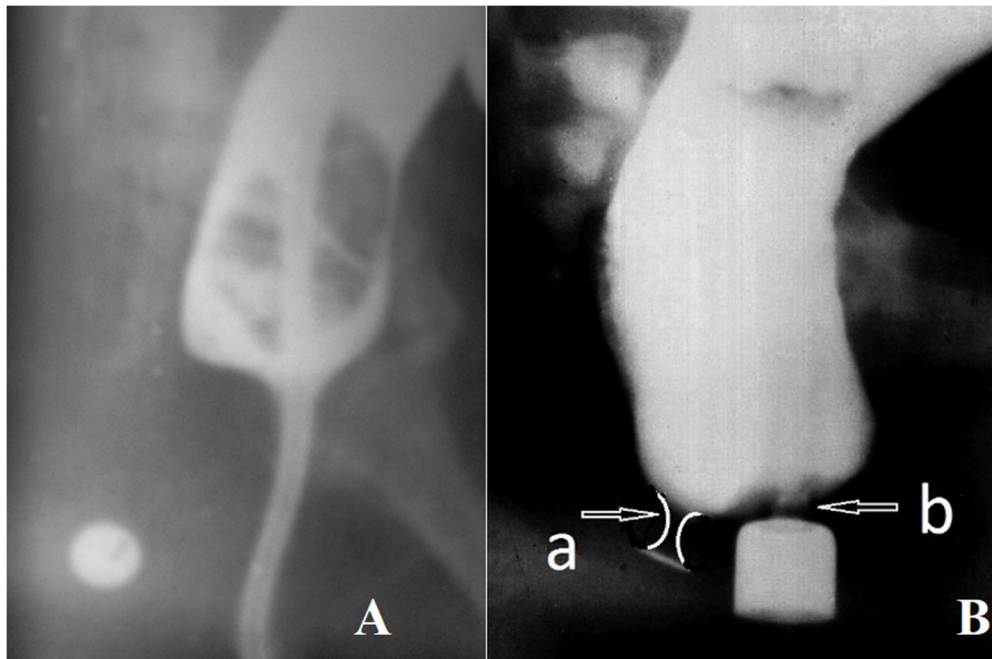


Figure 2. A female with a vestibular fistula. **(A)** Under the age of one year. Barium was injected into the rectum through a catheter inserted into the rectum from the fistulous opening. The button glued to the anal dimple. **(B)** At the age of 1.5 years, during the barium enema, a wide opening of the anal canal occurred **(a)** the distance from the wall of the anal canal to the anal dimple is $\approx 4\text{-}5\text{ mm}$; **(b)** the length of the narrow fistulous opening that was blocked by the tip of the enema is $\approx 2\text{-}3\text{ mm}$. The true width of the marker near the fistula is 1.6 cm.

This is a typical x-ray picture of patients with visible fistulas, which indicates the presence of IAS since only IAS can be in continuous contraction. As shown by manometric studies, all patients with visible fistulas have normal basal pressure and recto-anal inhibitory reflex [8, 9]. This is scientific evidence of the normal innervation of the rectum and anal canal, as well as the presence of functioning puborectalis muscle (PRM) and EAS. The wide opening of the anal canal during an attempt to defecate indicates the normal function of the levator ani muscle (LAM). Scientifically, this is a low type of ARM. From the point of view of pathological physiology, it is vestibular ectopy of the anal canal.

The authors cite an old concept about the role of LAM during defecation. If it relaxes during defecation, as it is written in the article, then it should be suspected that during fecal retention it should be in a contracted state. Firstly, striated muscles cannot be in a state of prolonged contraction. Secondly, LAM fibers at the periphery attach to the posterior and lateral surfaces of the pelvis, and the other side to the longitudinal muscle layer of the anal canal. Since the muscle shortens during contraction of muscle fibers, then when the LAM is contracted, the anal canal opens to pass stool [10, 11].

2) Most newborns with ARM without a visible fistula (urethral, vaginal, without fistula) have a functioning anal canal. On the first day after birth, it is usually in a closed state, as in healthy children. The anal canal opens only when sufficient volume of gas and meconium accumulate in the rectum to raise the pressure to a certain (defecation) level. For example, in patients with ARM who undergone sonography on both the birthday and the next day, the pouch-perineum distance on next day (mean \pm SD, 9.37 ± 4.89 mm; range 2.1-20.9 mm) was significantly shorter than on the birthday (15.75 ± 6.67 mm; range, 8.1-37.2 mm; $P = .001$) [12]. Nagdeve et al described 12 neonates male with high ARM who on invertogram showed a well-descended rectum with the lower limit of the rectal gas bubble at or below the ossified fifth sacral vertebra. Among them, there were eight patients with recto-bulbourethral fistula and four neonates with recto-prostatic urethral fistulas. Referring to Peña, the authors call this state "low-lying rectum". Results of the repair were "...without significant morbidity and good continence" [13].

I find it necessary to repeat the results of the Stepfens study: the rectum is always located above the pubococcygeal line. It is fixed in tissues and does not move. That intestine, which is located below the pubococcygeal line, is called the anal canal. Previously produced an invertogram, assuming that light gas is moving up. In fact, gas and meconium move in the gut with a peristaltic wave.

Gas enters the open anal canal when the pressure in the rectum rises to a certain level. This pressure is created with a certain volume of gas and meconium. 30 hours after birth, such a volume most often appears. Abdominal compression usually helps to open the anal canal (**Figure 3**).

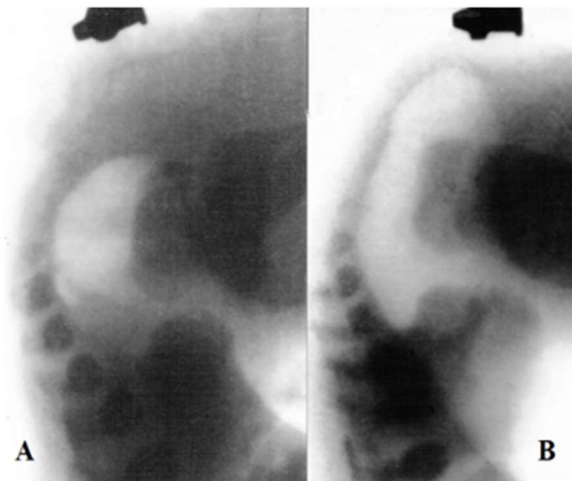


Figure 3. Radiograph of a newborn with ARM without visible fistula. (A) before the abdominal compressing. (B) during compression of the abdomen. Gas penetrated from the rectum into the open anal canal. The distance between the blind end of the intestine and the marker near the anal dimple is 2-3 mm and equal to the thickness of the skin and subcutaneous tissue.

The most convincing evidence of a functioning anal canal in 90% of patients with urethral fistula is presented in an article by Pena et al [14]. They showed that with a high-pressure distal colostogram, it is necessary to create sufficiently high pressure in the rectum so that the contrast medium penetrates the urethra. The author 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...". It is known there are no muscles around the rectum. The rectal function is to accumulate the feces. The rectum is in the open state around the clock and contracts only during defecation, when its strong peristaltic wave expels the feces through the open anal canal. In the description of Figure 3, the authors explain the closure of the "rectum" by the contraction of the "rectal sphincter". However, only three sphincteric structures are described in the anatomy of the pelvic floor: IAS, PRM, and EAS. A rectal sphincter does not exist. Thus, the terminal part of the intestine, which is in a constant contraction, is surrounded by sphincters,

including the IAS and the PRM, which opens in response to high pressure, it corresponds to the existing ideas about the anal canal.

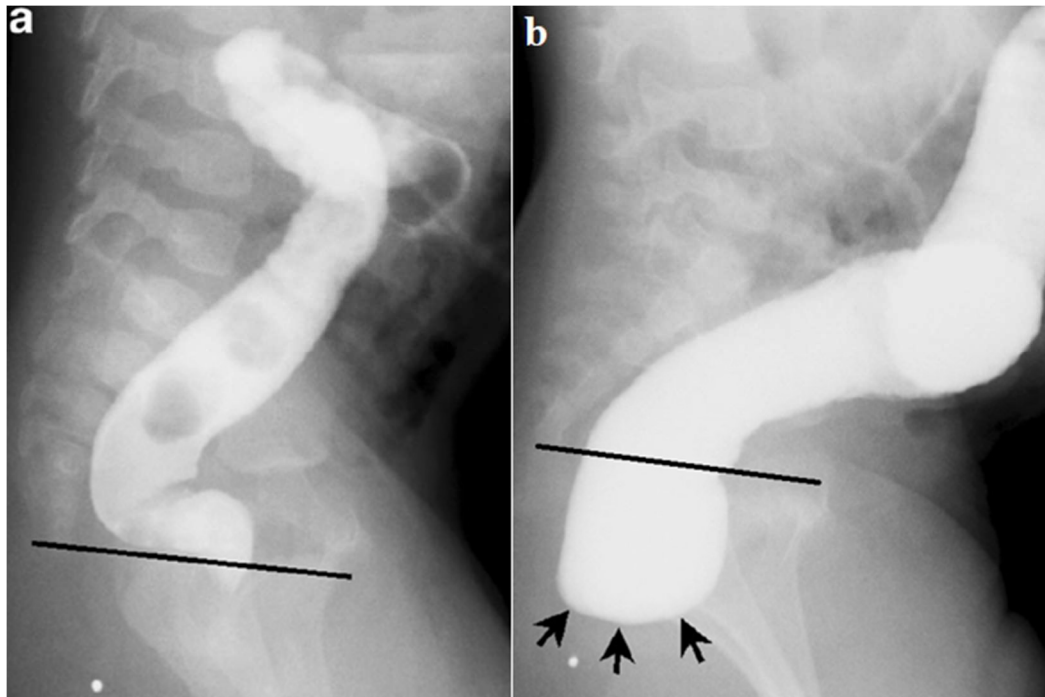


Figure 4. Distal colostograms of different patients from an article by Kraus et al [14]. (a) “Distal colostogram in a 4-month-old boy performed with inadequate pressure”. The distal part of the rectum is shifted forward as a result of the PRM function. The anal canal is closed since the rectum is not sufficiently high pressure. (b) “Distal colostogram in a 10½-month-old boy”. High pressure in the rectum led the opening of the anal canal, located below the pubococcygeal line (black line). Since the length of the anal canal at this age is approximately 2.3 cm, the distance from the distal wall of the anal canal to the marker in the anal dimple is 4 mm - this is the thickness of the skin and subcutaneous tissue.

An analysis of the literature and my research indicate that:

(a) all patients with visible fistulas and most patients ($\approx 90\%$) without a visible fistula have a functioning anal canal (IAS, EAS, PRM, and LAM, which are involved in recto-anal inhibitory and defecation reflexes.

(b) the described cases do not relate to high, but low types of ARM. The authors of the peer-reviewed article claim that "In high and intermediate ARMs (really low), the terminal rectum does not pass through the pelvic floor to the perineal skin". The rectum cannot move at all. When Peña uses various terms about a

moving rectum, rectal pouch or fistula, he means the anal canal, but he always uses coined names to avoid the question: why do you destroy the anal canal?

(c) The authors claim that «The perineal LMT is fused to form a vertical column in both high and intermediate ARM. In ARM, the LMT is a closed muscular tube, which could be possibly dilated to widen leading the rectum pull-through to establish the normal anorectal function" (**Figure 5**).

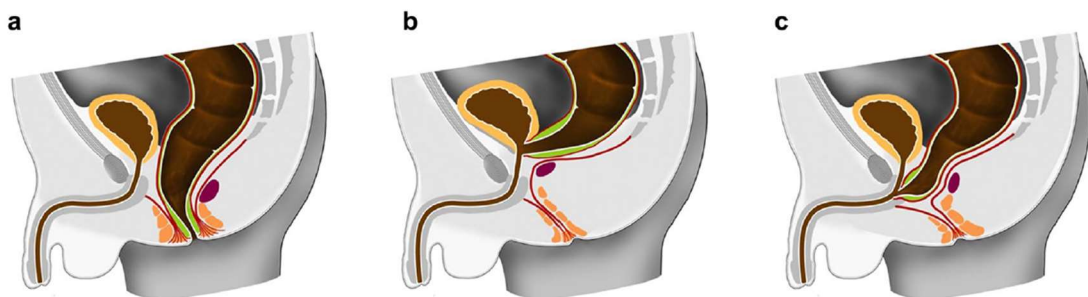


Fig. 6 Diagrams of the anorectal musculature in normal neonate (a), rectobladderneck fistula (b) and rectobulbar fistula (c). a showing that the longitudinal muscle tube (LMT) comes from the longitudinal smooth muscle of the rectum and longitudinal striated muscle of the levator ani muscle. b showing the LMT comes from the levator ani

muscle and is displaced upwards and anteriorly just behind the posterior urethra. c showing the LMT comes from the levator ani muscle, the pelvic LMT is widened by the rectal pouch and the perineal LMT is fused to form a column

In schemes from an article by Li et al (**Figure 5**), all proportions are violated. There are no designations. The anal canal is not closed, but not open. If IAS is marked in green, then its length in (a) occupies only a third of its actual length. With ARM (b, c), there is no IAS between the fistula and perineum, which contradicts anatomical examination (Figure 6). On CT with augmented-pressure distal colostogram it can be seen that longitudinal muscle cannot be fused. The closed anal canal has no volume. The thickness of IAS in newborns is about 1 mm. It is impossible to penetrate the lumen of the closed anal canal during surgery. In any case, the need to enter the center of the muscular complex past the anal canal leads to the excision of IAS and damage to the muscles of the pelvic floor.

The rectum lowered instead of IAS is torn off from blood vessels and nerves, as well as from contact with LAM. Because of this, the neural connection of the rectum with reflex arches is lost. Therefore, after pull-through operations, in

response to an increase in pressure in the rectum, the EAS and PRM residues do not contract, and during an attempt to defecate, the LAM does not relax. Thus, instead of a functioning anal canal, after a pull-through operation, regardless of access (anterior sagittal, posterior sagittal or with laparoscopic technique), a fistulous passage forms in the perineum. If the fistula is narrow constipation occurs if it is wide, fecal incontinence prevails.

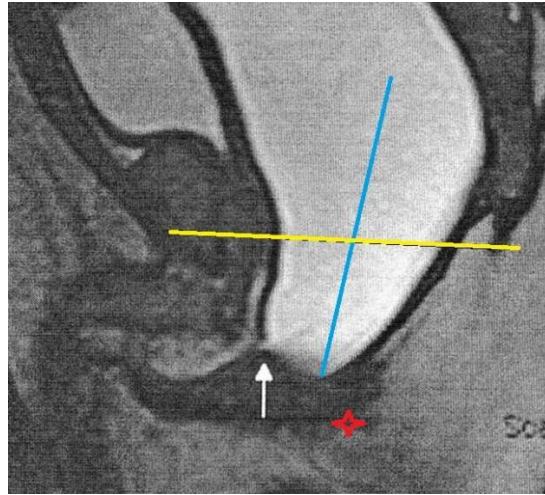


Figure 6. CT with augmented-pressure distal colostogram. The intestine located below the pubococcygeal line (yellow line) is an open anal canal. The fistulous opening (not the channel) between the anal canal and the urethra is indicated by an arrow. A red asterisk indicates the approximate location of the anal dimple.

At low pressure in the rectum, the anal canal closes and resembles a fistulous canal, the length of which, according to Koga et al, varies from 5 to 15 mm [15]. Obviously, the fistula is the opening between the urethra and the anal canal. The length of this anastomosis is equal to the thickness of the walls of the urethra and the IAS. And the fistula, which is removed by children's surgeons during the operation, it is closed IAS.

3) Operations that save only IAS or the anal canal are characterized by better functional results [5,7,16,17,18].

Table 1. Results - low anomalies (n=50; two patients died)

Operation	Score	(n)
Dilatation only	0	6
Cutback	0	29
	1	1
	9	1*
Anoplasty	0	11

Table 2. Results - intermediate anomalies (n=8; three patients died)

Operation	Score	(n)
Cutback	0	1
Transposition	0	1
	9	1*
Stephens	0	1
A-P pull through	5	1

*Associated neural tube defect

Table 3. Results - high anomalies (n=34; seven patients died)

Operation	(n)	Score (median) (range)
Stephens	12	4 (0-9)
A-P pull through	15	3 (0-9)

The results of the treatment of patients with ARM by Akroyda and Nur are given in 3 tables [16].

Long-term fecal continence was evaluated in 80 patients 6–18 years after the surgical treatment of ARMs. Patients with complete continence scored 0, and those with occasional problems with flatus or diarrhea scored 1 to 3. Patients with weekly flatus escape, incontinence of diarrhea or loss of control of solid motions scored between 2 and 6 and those with complete fecal incontinence scored 9.

The patients with low anorectal anomalies did very well with fecal continence achieved in 47 out of 48 surviving patients. Good results were obtained with anal cutback, anoplasty or dilatation only.

Please note that after the cutback procedure in which the anal canal is completely preserved, and after anoplasty, in which IAS is preserved, normal fecal continence is determined. Results after the pull-through operation are noticeably worse. Pull-through operation through the posterior sagittal approach is theoretically worse than other pull-through operations because it is accompanied by the intersection of PRM. Peña and Levitt, from the very beginning, stated that patients with ARM did not have an anal canal, so they removed the so-called fistula (rectal pouch) in all patients. As a result, almost all ARM cases have become high. The results of the operations are announced great, because in them from birth supposedly had no anal canal. Peña has described various degrees of postoperative constipation in 70% of patients with vestibular fistula and in 50% of patients with low-type ARM. When the malformation is

lower, the incidence of severe constipation is higher, leading to overflow pseudo-incontinence [19]. Is this mystification or fraud?

This article justifies the laparoscopic pull-through operation, which was performed in 330 patients with high-type (174 cases) and intermediate-type (156 cases) anorectal malformation [20]. As shown above, in 90% of boys with fistulas in the urethra, the distal contour of the intestine is below the pubococcygeal line, that is, in fact, they had a functioning anal canal. Obviously, the rectum cannot be in a contracted state like IAS. The striated fibers of PRM and EAS cannot be in a contracted state for more than 1 minute. Moreover, they do not contract at all, as the nerve connection with the distant IAS is interrupted. During bowel movements, LAM does not affect the width of the rectum. Therefore, the results of the operation cannot be good. Repeated operations were performed in 30 (9.1%) patients (rectal prolapse-25; anal stricture -3; anal retraction -2). 13 (3.9%) children had involuntary loss of feces. 217 patients (95.2%) had voluntary bowel movements. This means that as a result of the daily use of enemas and/or laxatives, they did not have an involuntary loss of feces. 202 patients (88.6%) were free from soiling or with grade 1 soiling, and 55 (16.6%) had chronic constipation. It should be borne in mind that with age, the degree of chronic constipation and fecal incontinence increase. In adults “A large proportion of the patients have persistent fecal incontinence, constipation and sexual problems that have a negative influence on their quality of life” [21].

These patients could be healthy if their anal canal was preserved [22,23].

Conclusion

Anatomical, functional studies of anorectum in patients with ARM indicate the presence of a functional anal canal in most children. This is confirmed by the results of operations preserving the elements of the anal canal. The aim of the authors of the reviewed article is to justify pull-through operations, which

irreversibly destroy the anal canal, which leads to the disability of patients. The authors deny the known scientific facts, cite articles in which there is no scientific evidence, neglect the fundamental knowledge of the anatomy and physiology of anorectum. I believe this article is not scientific. The results of the "coup" in pediatric colorectal surgery performed by Peña and his followers need to be given a scientific assessment.

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