

Dear colleagues,

In the Journal of Pediatric Radiology was published an article Kraus et al. "Augmented-pressure distal colostogram: the important diagnostic tool for planning definitive surgical repair of anorectal malformations in boys" [1]. The authors share their experience in applying high pressure during the production of colostography. It should be agreed that "accurate understanding of the anatomy of the anorectal malformation including an associated fistulous communication between the rectum and the urogenital tract is essential for optimal surgical management".

However, the article contains incorrect interpretations of the physiology of anorectum and contradictory of the statements.

1). The authors state that "... the first posterior sagittal exploration of the child with ARM was performed, allowing the first direct observation of the intrinsic anatomy of these defects. It was found that the "puborectalis muscle", traditionally considered an important landmark, actually was not the "rectal sphincter". The authors refer to the article Peña, where he came to this conclusion on the basis that he could not identify the puborectalis muscle (PRM) in the surgical wound. Based on this, they "believe that preservation of the "puborectalis" portion of the levator ani alone, as stressed by Stephens, should be amended to a consideration and preservation of the entire potential sphincteric musculature, which includes the striated muscle complex, levator ani, and external sphincter" [2].

First, operational findings cannot be more precise than histological and anatomical studies, which are the gold standard. It is known that PRM is difficult to differentiate during anatomical dissection [3]. Therefore, it is not surprising that Pena, like all surgeons before and after him, could not identify PRM during the surgery. However, there is no logical connection between what he did not see and his "discovery".

Secondly, this statement contradicts fundamental research prove that the PRM plays an important role in the fecal retention. Mittal et al. concluded: "...the proximal half of the anal canal is surrounded by the "U"-shaped PRM, and closure of the proximal half of the anal canal is related to contraction of the IAS and PRM. In fact, the PRM forms the inferior margin of the pelvic floor hiatus, and its contraction causes closure of not only the anal canal, but the vagina and urethra as well [4]. "Based on the physiologic studies, it appears that the puborectalis muscle is the 3rd constrictor or the sphincter of anal canal" [5].

Thirdly, unfortunately, this false assertion is a "theoretical" justification for the intersection of PRM during the posterior sagittal anorectopasty (PSARP).

Fourth, levator ani is not a sphincter. During contraction, it opens the anal canal to the width of the rectum [6,7]. But after PSARP, this muscle cannot perform its function, as it is cut off from the rectum, to which usually attaches. By the striated muscle complex, the authors mean the deep and superficial parts of the external anal sphincter (EAS), the circular fibers of which intersect simultaneously with the intersection of PRM (see Figure 1. j in article). At the final stage of the operation, suturing of the muscle mass is performed, but it is impossible to sew sphincters that do not differ from each other. By external sphincter, the authors mean the subcutaneous portion of EAS. After PSARP this thin striated muscle is contracted only with a volitional impulse no longer than one minute. It does not respond with reflex contraction in response to increased pressure in the rectum since as a result of denervation of the rectum, the reflex connection between the rectum and EAS is lost.

2) 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..."

First, it is known that there are no muscles around the rectum. The rectal function is to accumulate of the feces. The rectum is in the open state around the clock and contract only during defecation, when its strong peristaltic wave expels the feces through the open anal canal (Textbooks).

Secondly, in the description of Figure 3, the authors explain the closure of the "rectum" by the contraction of the "rectal sphincter" (**Figure 1**). However, only three sphincteric structures are described in the anatomy of the pelvic floor: an internal anal sphincter, a puborectalis muscle and an external anal sphincter. Rectal sphincter does not exist.

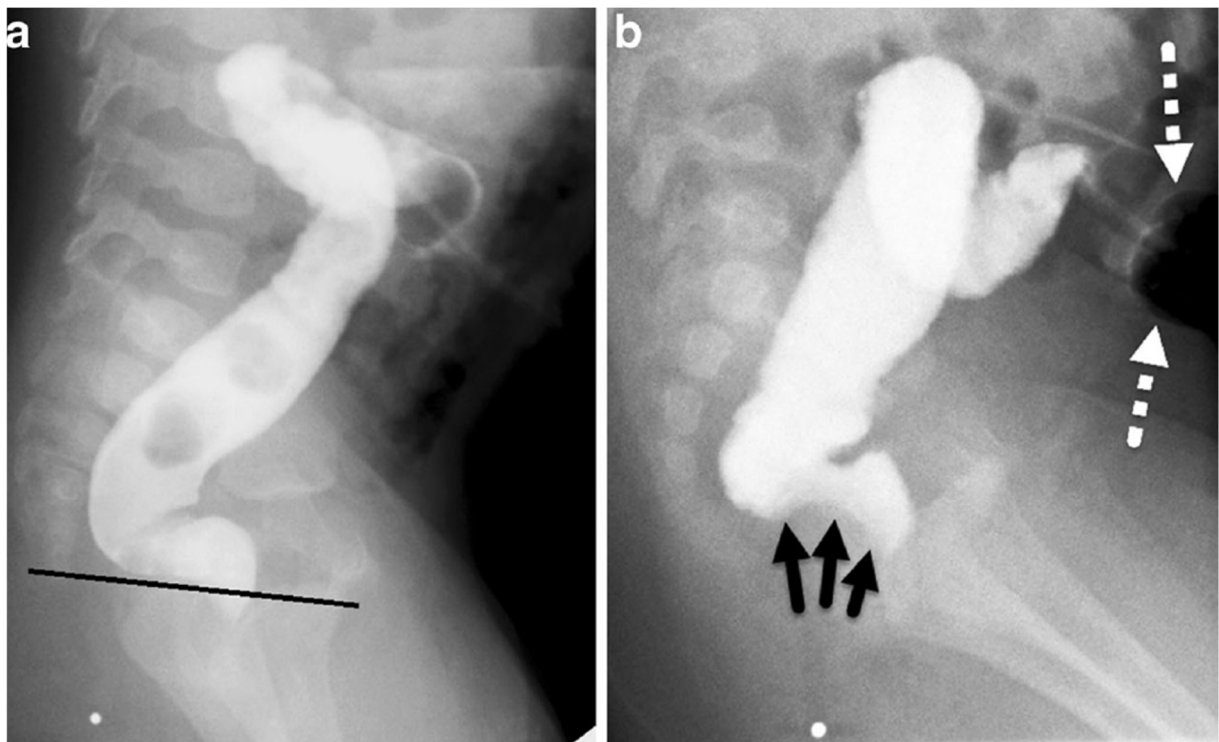


Figure 1. Figure 3 from the article under review. In the pictures, the distal part of the rectum is shifted forward. Displacement of the rectum forward with the formation of the lower horizontal branch occurs during ontogenesis under the influence of the PRM contraction. In the resting phase, the impression of puborectalis sling is visible on the posterior wall of caudal rectum and the anorectal angle is about 78° (A) and 90° (B). The concavity of the lower contour is a recognized symptom of PRM pressure [8]. The distance between the rectum and the marker on the perineum is equal to the normal length of the anal canal, which "...is closed because of inadequate pressure" [1].

Thus, the terminal part of the intestine, which is in a constant contraction, is surrounded by sphincters and opens in response to high pressure, corresponds to the existing ideas about the anal canal. The described parameters of this anal canal indicate the functioning of IAS, PRM, EAS and levator plates. This

channel, in contrast to the normal anal canal, is located outside the ring of the subcutaneous part of the EAS.

The authors of the article cannot call this part "an anal canal", since they without any evidence are claimed that in patients with ARM there is no anal canal [9]. In their articles, they call this intestinal part "a rectum", "rectal pouch" or "fistula". The problem is not the name. Calling the anal canal by "a fistula", they during surgery resect IAS [1,9].

3. The authors believe that «...the traditional old classification of the defects into “high”, “intermediate” and “low” has proved to be fictitious and misleading”. However, they repeatedly emphasize that the most important diagnostic conclusion of the augmented-pressure distal colostogram is “... reporting the level of the distal rectal pouch in relation to the last ossified sacral segment ...” It “...is important in planning the surgical approach. Perineal, bulbar-urethral and select prostatic-urethral fistulae are repaired primary by posterior sagittal approach only (because the rectal pouch is at or inferior the ossified sacral tip) “. «If the distal rectal pouch and fistula are above the ossified sacral tip ...likely necessitates an abdominal approach". Thus, the recommendations of these authors contradict their statement that the old classification is fictitious and misleading, since the main purpose of the augmented-pressure distal colostogram is the differentiation of the ARM into high (above the ossified sacral tip) and low types (inferior the ossified sacral tip).

4. Of the 13 radiographs with “the small radiodense BB markers”, only in 3 cases these markers are in the anal dimple projection (**Figure 2. A**). On 10 radiographs, a significant removal of the marker from the pubococcygeal line is determined (Figure 2. B). In 2 cases, the marker is outside the boundaries of the body (Figure 2.C).

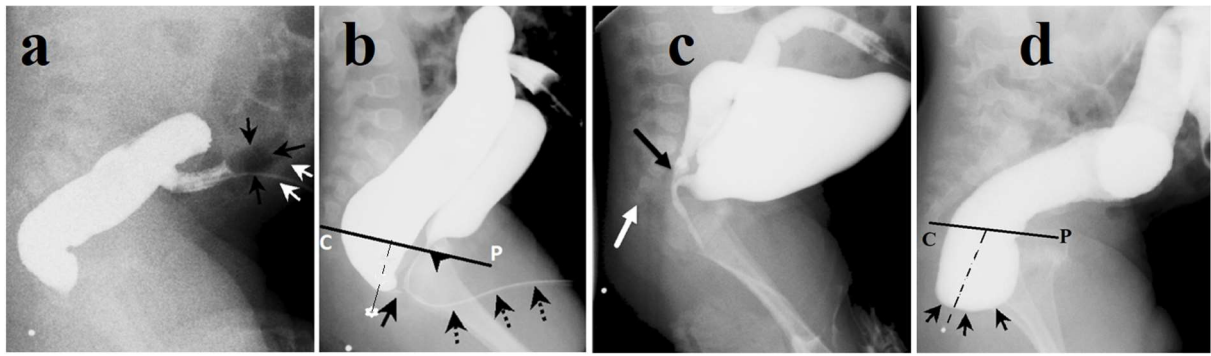


Figure 2. Figures from the peer-reviewed article. **(a).** The marker is located near the anal dimple. The distance between the rectum and the marker is a closed anal canal. **(b).** The marker is in the significant distance from the anal dimple. My asterisk is located near the anal dimple. The distance between the P-C line and marker is an open anal canal **(c).** The marker is located outside the body of patient. **(d).** In a 10½-month-old boy without fistula, the distance from the distal wall of the open anal to the marker is 24% of the total length of the anal canal. The average length of the anal canal at this age is 2.24 cm. Therefore, the distance from the marker to the wall of the anal canal is 5 mm. P-C: pubococcygeal line. The dashed line corresponds to the length of the anal canal.

Because of these errors, no matter how it happened, a false idea is created about the great remoteness of the distal contour of the open anal canal from the anal dimple. In fact, the distance between the open anal canal and the skin is from 2 mm to 5 mm, depending on the age and represents the thickness of the skin and subcutaneous tissue [10], as in Figures 2 b and d.

5. During the augmented-pressure distal colostogram, a very high pressure is created in the rectum, which the authors mistakenly call hydrostatic. When using a leur-lock syringe, an uncontrolled hydrodynamic pressure is created in the gut to mechanically overcome the contraction of the anal sphincters. There is a prolonged stretching of the anal canal, during which the researcher seeks to achieve reflux of the contrast medium into the urinary tract. Since this study is dangerous by rupture of the intestine, many researchers prefer to stop "on time".

Meanwhile, the opening of the anal canal can be achieved by provoking an act of defecation. For this, it is enough to fill the rectum with a volume corresponding to the capacity of the rectum and increase the pressure to the pressure of defecation

by the abdominal compressing. In newborns, an accurate determination of the level of ARM is easier and safer to determine 24 hours after birth, when the rectum is filled with a large volume of gas and meconium. The abdominal compression causes: an increase of the rectal pressure to the threshold level, the opening of the anal canal and the penetration of the gas into the distal gut, what outlines the distal contour of the anal canal. This reflex acting lasts a few seconds. For its registration it is necessary the fluoroscopic observation (**Figure 3**).

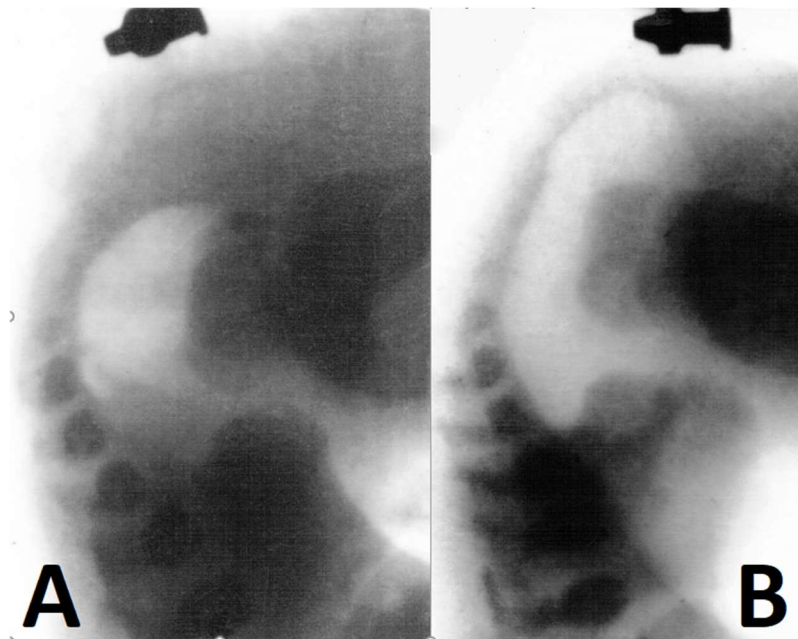


Figure 3 Lateral radiographs of a newborn boy with ARM, made in a horizontal position. (A). At rest. (B) . During abdominal compression, as a result of the reflex opening of the anal canal, the distal anal wall approached the anal dimple. The distance between the marker in the anal dimple and the caudal wall of the anal canal is 2 mm. Conclusion: low type of ARM.

The use of this method allows us to state: a) this diagnostic method is completely safe, c) the opening of the anal canal indicates a low type of ARM: c) 90% of boys with urethral fistulas have a normally functioning anal canal.

6. From this it follows that in 90% males with ARM and a fistula in the urinary system have the functional anal canal and therefore they belong to a low type of ARM.

During PSARP, (a) the contracted IAS is removed as fistula, and in its place the rectum moves, which cannot be in the contracted state; (b) PRM intersects ; (c) all 3 parts of EAS are injured; (d) the rectum is separated from the levator plates; (e) the neural connections of the rectum with the muscles of the pelvic floor intersect (denervation). Thus, instead of a functioning anal canal, a fistula on the perineum is created. Even though in almost all articles the authors report good results after PSARP, long-term results studies show that among adults operated on for ARM, there are no healthy patients: 75% have constipation and 25% have fecal incontinence. Among the interviewed 123 patients operated in Germany, to varying degrees fecal incontinence was in 74% of cases [11]. Levitt and Peña claim PSARP is the perfect operation. The results of ARA treatment could not be better, since these patients have no anal canal [9]. Meanwhile, it was shown that the use of a fistula (read IAS) in the reconstruction of the ARM leads to a noticeable improvement in functional results [12-18]. I offer a scientifically sound alternative to pull-through operation [10,19].

Conclusion

The entire PSARP concept is based on false beliefs about the pathological anatomy and physiologists of the ARM. Therefore, it turned out to be impossible to evaluate the diagnosis of ARM with fistulas in the urinary system in boys in isolation from the whole complex of knowledge about ARM, including fundamental knowledge, diagnosis and treatment. Peña et al, actually created an alternative pseudoscience, not only regarding the anatomy and physiology of anorectum, even the anatomical terms do not coincide with modern scientific knowledge. I believe that pediatric surgeons who are involved in treatment of ARM should to know the pathological anatomy and physiology of the ARM, for to select evidence-based and effective methods for diagnosing and treating ARM.

Yours faithfully

M.D. Levin, MD, PhD, DSc. Radiologist,

Department of Pediatric Radiology of the 1-st State Hospital, Minsk, Belarus.

Dorot-Netanya Geriatric Medical Center, Israel.

Amnon VeTamar, 1/2, Netanya, 42202, Israel.

nivel70@hotmail.com; michael.levin@dorot.health.gov.il

<https://orcid.org/0000-0001-7830-1944>

<https://www.anorectalmalformations.com>

Scopus [Author ID: 7402571390](#)

References

1. 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 Feb;48(2):258-269.
2. Peña A, Devries PA. Posterior sagittal anorectoplasty: important technical considerations and new applications. *J Pediatr Surg*. 1982 Dec;17(6):796-811.
3. Bharucha AE. Pelvic floor: anatomy and function. *Neurogastroenterol Motil*. 2006 Jul;18(7):507-19. Review.
4. Mittal RK, Bhargava V, Sheean G, et al. Purse-string morphology of external anal sphincter revealed by novel imaging techniques. *J Physiol Gastrointest Liver Physiol*. 2014 Mar;306(6):G505-14
5. Raizada V¹, Mittal RK. Pelvic floor anatomy and applied physiology. *Gastroenterol Clin North Am*. 2008 Sep;37(3):493-509, vii. doi: 10.1016/j.gtc.2008.06.003.
6. Bush M¹, Petros P, Swash M, et al. Defecation 2: Internal anorectal resistance is a critical factor in defecatory disorders. *Tech 16 Coloproctol*. 2012 Dec;16(6):445-50. doi: 10.1007/s10151-012-0860-3. Epub 2012 Jul 24.
7. Levin MD. Anatomy and physiology of anorectum. The hypothesis of fecal retention and defecation. https://docs.wixstatic.com/ugd/4d1c1d_05ebf5341ad14290938136ef13e1bcdd.pdf
8. Kim AY. How to interpret a functional or motility test - defecography. *J Neurogastroenterol Motil*. 2011 Oct;17(4):416-20. doi: 10.5056/jnm.2011.17.4.416.
9. Levitt MA¹, Peña A. Anorectal malformations. *Orphanet J Rare Dis*. 2007 Jul 26;2:33.

10. Levin MD. Anorectal malformations.
https://docs.wixstatic.com/ugd/4d1c1d_0961be5f16f34858bd226cc847191b83.pdf
11. Grasshoff-Derr S¹, Backhaus K, Hubert D, Meyer T.
A successful treatment strategy in infants and adolescents with anorectal malformation and incontinence with combined hydrocolonic ultrasound and bowel management. *Pediatr Surg Int*. 2011 Oct;27(10):1099-103. doi: 10.1007/s00383-011-2950-0.
12. Stephens FD. Imperforate rectum. A new surgical technique. *Med J Australia*. 1953;1:202.
13. Ruttenstock EM¹, Zani A, Huber-Zeyringer A, Höllwarth ME. Preand postoperative rectal manometric assessment of patients with anorectal malformations: should we preserve the fistula? *Dis Colon Rectum*. 2013 Apr;56(4):499-504. doi: 10.1097/DCR.0b013e31826e4a38.
14. Sangkhathat S¹, Patrapinyokul S, Osatakul N. Crucial role of rectoanal inhibitory reflex in emptying function after anoplasty in infants with anorectal malformations. *Asian J Surg*. 2004 Apr;27(2):125-9.
15. Kyrklund K¹, Pakarinen MP, Taskinen S, Rintala RJ. Bowel function and lower urinary tract symptoms in males with low anorectal malformations: an update of controlled, long-term outcomes. *Int J Colorectal Dis*. 2015 Feb;30(2):221-8. doi: 10.1007/s00384-014-2074-9. Epub 2014 Dec 2.
16. Kyrklund K¹, Pakarinen MP², Rintala RJ². Manometric findings in relation to functional outcomes in different types of anorectal malformations. *J Pediatr Surg*. 2017 Apr;52(4):563-568. doi: 10.1016/j.jpedsurg.2016.08.025. Epub 2016 Sep 2.
17. Smith EI, Tunell WP, Williams GR. A clinical evaluation of the surgical treatment of anorectal malformations (imperforate anus). *Ann Surg*. 1978 Jun;187(6):583-92.
18. Nainan KM, Mitra SK, Pathak IC. Perineal anal transplant in anorectal malformation in female patients. *Surgery*. 1975 May;77(5):694-702.
19. Levin MD. [The pathological physiology of the anorectal defects, from the new concept to the new treatment]. *Eksp Klin Gastroenterol*. 2013;(11):38-48. [Article in Russian]