

What does the analysis of anorectal malformations diagnosed in adolescence indicate?

While most of the patients with anorectal malformations (ARMs) present in either neonatal period or early infancy, a small percentage presents in their adolescence. It is believed that the main reasons for the delay in presentation were misinformation, illiteracy, and poverty [1]. This mainly concerns females with visible fistulas, and the conclusion about the reason for the delayed diagnosis, although correct, is not complete. Some articles describe cases of successful treatment of these patients, but only the article by Rawat et al. provides a documented description of each of the 10 observations of females aged between 12 and 18 years (average 14.4 years) [1]. Among them were 5 girls with anovestibular fistula, 3 with anterior ectopic anus, 1 with perineal canal, and 1 with rectovestibular fistula. Four (40%) girls had no problem apart from an abnormal anal orifice and normal to mildly enlarged rectum on contrast study. Including 3 with anovestibular fistula and one with anterior ectopic anus. Six (60%) girls had constipation as a major additional symptom and the rectum was moderately to hugely dilated. Six cases were managed with posterior sagittal anorectoplasty (PSARP) while an anterior sagittal anorectoplasty (ASARP) was done in four. In a mean follow-up of 16 months (8-26), constipation and soiling were seen in 5 (50%) girls. The authors rated this outcome as satisfactory.

First, in three females with anovestibular fistula and one with anterior ectopic anus, fecal continence and defecation were normal. This indicates that the anteriorly displaced anus was elastic and of normal width and that the distal bowel had normal sensation and functioned as a normal anal canal. These compelling data contradict the contention of Levitt and Peña that the distal bowel lacks sensation and should not be preserved during correction [2].

Secondly, there were 6 vestibular fistulas in total. In addition to 3 cases with a normal opening, which were operated with ASARP, 2 cases had a narrow ectopic anus, which led to megarectum, and they were operated with PSARP. From these data it follows that the authors knew that in anovestibular ectopia there is an anal canal, but following unfounded recommendations, they destroyed it, since they resected the internal anal sphincter, tearing the rectum from the levator plates, and deprived of vascular nutrition and innervation, they brought it down to the perineum. In one case, a rectovestibular fistula was diagnosed, which was also operated with PSARP. Meanwhile, since 1953, based on the studies of Stephens, it has been known that in ARM with visible fistulas (perineal and vestibular), there is always a normally

functioning anal canal, which must be preserved [3]. The detection of gas above the pubococcygeal line only indicates that the anal canal is in a closed state (as normal) [4, 5].

Thirdly, after 16 months (8-26) after surgery, constipation and soiling were seen in 5 (50%) girls. The authors considered that this result is satisfactory, but this is contrary to the facts. (1) All four patients, who had no complaints before the operation, except for the ectopic anus, developed constipation. The time elapsed after the operation is too short to definitively determine the results, but it is known that constipation and fecal incontinence increase with time. Danielson et al. found that the ARM-patients had an inferior outcome ($P < 0.05$) for all incontinence parameters, 8 of 10 parameters for constipation, 2 of 6 for urogenital function and 7 of 13 quality of life parameters. Patients with rectobulbar and vestibular fistulas had the worst statistical outcome [6]. (2) None of the 10 patients had fecal incontinence before surgery. Four of the six patients had no constipation after PSARP, but the table does not show the follow-up results. It is obvious that destruction of the anal canal inevitably leads to fecal incontinence.

Fourthly, the cutback procedure, in contrast to the pull-through operation, entirely preserves the anal canal. As indicated by the outcomes of this procedure, intersecting the subcutaneous segment of the EAS never results in fecal incontinence. The objective of this operation is to incise the rigid ring to establish regular defecation. As noted by Nixon, "The simple cutback is all that is necessary to enlarge the imperfect anus adequately for functional use [7,8,9]. This procedure yielded positive functional outcomes in both male and female children with perineal and vestibular fistulas. For instance, Browne wrote, "Once it is understood that the vaginal opening is a true anus, complete with nerve and muscle sphincter mechanisms, albeit misplaced and often stenotic, treatment becomes more manageable and successful". If the opening is situated deep within the vaginal orifice, a backward transplantation can be conducted, preferably between ages 5 to 7 years" [10]. The theoretical rationale for modifying the cutback procedure may be useful to relocate the newly created anus to its normal location [4].

An analysis of ARM cases diagnosed in adolescence has shown that one of the reasons for the late presentation of patients for medical care was the absence of complaints. This was due to the wide and elastic ectopic anus and the presence of a normally functioning anal canal. Therefore, they did not have constipation and fecal incontinence. In other studies, the presence of a normally functioning anal canal was proven by radiographic [4,5,11] and manometric [5,12] studies (**Figure 1**).

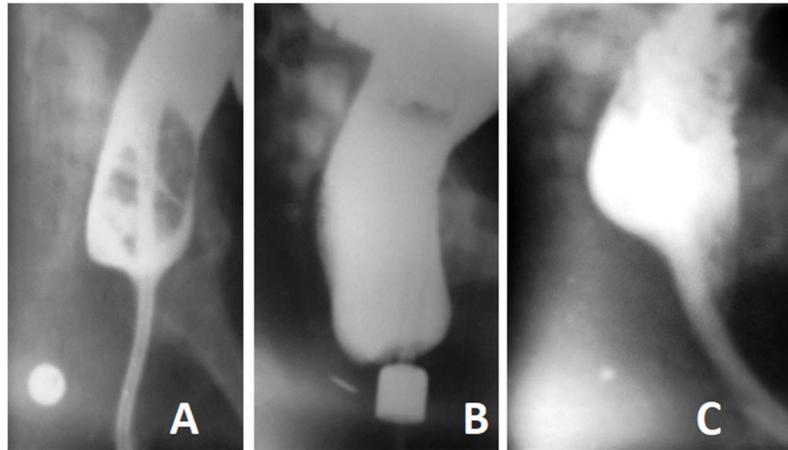


Figure 1. Lateral radiographs of the anorectum in patients with ARMs with visible fistulas. **(a-b).** Images of the same child taken at various times. **(a).** A catheter was inserted through the vestibular fistula into the rectum to enter contrast substance into the intestine. At rest, the anal canal is contracted around the catheter. **(b).** During a follow-up examination, an attempt to defecate resulted in a wide opening of the anal canal. The distance between the marker in the anal dimple and the wall of the open anal canal is 4 mm. It is equal to the thickness of the skin and subcutaneous tissue. **(c).** During the administration of barium through the endotracheal tube the relaxation of the IAS occurred, allowing barium to enter the upper part of the anal canal, in front of the rectal tube. This was accompanied by a drop in rectal pressure, which was measured by a balloon connected to a pressure gauge [13]. At this moment, the posterior wall of the anal canal is pressed against the rectal tube by the contraction of the PRM, which together with external anal sphincter closes the anal canal.

As shown above, preservation of the anal canal leads to significantly better results than pull-through surgery, regardless of the approach. This is evidenced by the statement of the European 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” [14].

Currently, all surgical methods of treating ARM are based on Peña's unfounded assertion that there is no anal canal in ARM, which results in the destruction of the normally functioning anal canal with severe complications for the rest of life (constipation, fecal incontinence, urological and sexual problems). The only scientific justification for the removal of the so-called "rectal sac or fistula" was the absence of the intermuscular nerve plexus in its wall [15]. However, this fact is another confirmation that the so-called fistula is an anal canal.

Since in the anal canal, unlike other parts of the digestive system, there are no or only isolated nerve ganglia [16,17,18,19].

Peña trained a generation of pediatric surgeons who disdain scientific evidence. They still cite Peña's experience, which consisted of numerous articles where he broadcast his experience. The result of the destruction of pediatric colorectal surgery is evident in the recently published article by Munyali et al [20]. The article consists of two parts. The authors' description of the cases does not contain characteristics of the stool, displaced anus, and rectum. There are no numbers. After sagittal anorectoplasty according to Pena and dilatations using Hehar candles fecal continence, assessed according to the Hassink criteria, was normal [20]. The article has no scientific or practical content. It was published to demonstrate successful healing after PSARP. However, firstly, there are no Hassink criteria for determining the quality of fecal continence in literature. Secondly, this conclusion is fundamentally false, especially since such patients often experience constipation, which is not mentioned at all. The second part (Discussion) was written by an anonymous reviewer, who described the current dead-end state of colorectal surgery.

Conclusion. The Peña heirs have never debated the existence of the anal canal. They hold pediatric surgeons and patient's hostage because they fear responsibility for the destruction of the anal canal. Therefore, they control the publication of articles and do not allow dissent. Sending questionnaires to former patients in which patients and their parents claim that they are happy with life despite fecal incontinence, severe constipation, sexual problems is a dead end. The work of the American Pediatric Surgical Association, who report in an article on almost gender equality among pediatric surgeons, is a dead end. I call on pediatric surgeons to verify the accuracy of my evidence and publish it to return science to pediatric colorectal surgery.

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

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

nivel70@hotmail.com

<http://www.anorectalmalformations.com>

References

1. Rawat J, Singh S, Pant N. Anorectal Malformations in Adolescent Females: A Retrospective Study. *J Indian Assoc Pediatr Surg.* 2018 Apr-Jun;23(2):57-60. doi: 10.4103/jiaps.JIAPS_125_17.
2. Levitt MA, Peña A. Anorectal malformations. *Orphanet J Rare Dis.* 2007 Jul 26;2:33. doi: 10.1186/1750-1172-2-33. Erratum in: *Orphanet J Rare Dis.* 2012;7:98.
3. Stephens FD. Imperforate rectum. A new surgical technique. *Med J Australia.* 1953;1:202.
4. Levin MD. Anorectal Malformations with Visible Fistulas: Theoretical Substantiation of a New Version of the Cutback Procedure. *J Pediatr Perinatol Child Health* 2024. Volume 8 • Issue 4. 210-216. DOI:10.26502/jppch.74050203
5. Levin MD. Embryological Development of Anorectal Malformations: A Hypothesis. *Qeios*, CC-BY 4.0. Preprint · Article, October 22, 2024. <https://doi.org/10.32388/HIMVOF>
6. Danielson J, Karlbom U, Graf W, Wester T. Outcome in adults with anorectal malformations in relation to modern classification - Which patients do we need to follow beyond childhood? *J Pediatr Surg.* 2017 Mar;52(3):463-468. doi: 10.1016/j.jpedsurg.2016.10.051.
7. Nixon HH. Anorectal anomalies: with an international proposed classification. *Postgrad Med J.* 1972 Aug;48(562):465-70. doi: 10.1136/pgmj.48.562.465. PMID: 5078222; PMCID: PMC2495259.
8. Kyrklund K, 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. PMID: 25435141.
9. Wilkinson AW. Congenital anomalies of the anus and rectum. *Arch Dis Child.* 1972 Dec;47(256):960-9. doi: 10.1136/ad.47.256.960. PMID: 4647050; PMCID: PMC1648405.
10. BROWNE D. Congenital deformities of the anus and the rectum. *Arch Dis Child.* 1955 Feb;30(149):42-5. doi: 10.1136/ad.30.149.42.
11. Levin MD. Pathological physiology of the anorectal malformations without visible fistula. A short review. *Pelvipерineology* 2023;42(2):74-79. DOI: 10.34057/PPj.2022.41.02.2021-9-1.
12. Ruttenstock EM, Zani A, Huber-Zeyringer A, Höllwarth ME. Pre- and 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. PMID: 23478618.
13. Levin MD. The pathological physiology of the anorectal defects, from the new concept to the new treatment. *Eksp Klin Gastroenterol*. 2013; (11):38-48.
 14. Amerstorfer EE, Schmiedeke E, Samuk I, Sloots CEJ, van Rooij IALM, Jenetzky E, Midrio P, 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.
 15. 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.
 16. Duhamel B. Physio-pathology of the internal anal sphincter. *Arch Dis Child*. 1969 Jun;44(235):377-81. 13.
 17. Howard ER, Nixon HH. Internal anal sphincter. Observations on development and mechanism of inhibitory responses in premature infants and children with 10 Hirschprung's disease. *Arch Dis Child*. 1968 Oct;43(231):569-78. doi: 10.1136/adc.43.231.569.
 18. 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.
 19. 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.
 20. Munyali A, Buhendwa C, Ganywamulume B, et al. Adolescent anorectal malformations: Case series about 3 cases. *Int J Surg Case Rep*. 2025 Feb;127:110913. doi: 10.1016/j.ijscr.2025.110913.