

Reaction to the article by Huijgen et al., «Long-term follow-up in patients with anorectal malformation: MRI findings in relation to bowel function».

Abstract

The present study analyzes the article by Huijgen et al., "Long-term follow-up in patients with anorectal malformation: MRI findings in relation to bowel function", which demonstrates the current state of pediatric colorectal surgery and the role of personality in a context where physiological science has ceased to be funded by the state. It presents evidence of the destruction of pediatric colorectal surgery by Dr. Peña and his followers. The article lacks an understanding of the physiology and pathology of the anorectal area in health and with ARM. It ignores the research of several generations of pediatric surgeons who have proven the existence of an anal canal and achieved good results by preserving it. There are no references to modern articles in which the authors reject PSARP due to the destruction of anal sphincters. The article lacks scientific methodology, resulting in incorrect and dangerous conclusions and proposals. In fact, everyone involved in the decision to publish it promotes PSARP, which causes lifelong suffering. Meanwhile, if all elements of the anal canal are preserved, these patients can be healthy.

Keywords: Anorectal malformations; posterior sagittal anorectoplasty; cutback; anal canal physiology; Alberto Peña; postoperative function; classification.

I read with great interest the recent article by Huijgen et al., "Long-term follow-up in patients with anorectal malformation: MRI findings in relation to bowel function" [1], as it reflects the current state of pediatric colorectal surgery. The authors provide anatomical data MRI-based long-term observations, which are little discussed among pediatric surgeons. Important methodological problems and interpretative issues merit scientific analysis.

First, the authors cite only articles that describe Peña's experience and completely ignore articles in which scientific data contradicts Peña's "experience." The article presents an unjustifiably truncated MRI image in patients after surgical correction of ARMs. Of the 55 patients in this study, 8 were excluded because they did not undergo surgery, as they had a very mild type of ARM. Thus, authors missed the opportunity to compare images before and after correction. Eighteen patients were excluded because ARM was associated with spinal pathology. Peña and Levitt declared posterior sagittal anorectoplasty (PSARP) to be an ideal

procedure, and poor results were attributed to spinal malformations. However, concomitant spinal pathology does not affect the function of continence and defecation [2]. In 4 patients, MRI data did not correspond to functional assessments or they underwent re-correction. All these exclusions have no scientific justification. They change the overall picture of postoperative results.

Second, the manuscript states that the Krickenbeck classification was followed. However, this classification does not distinguish between “simple” and “complex” malformations comparable to earlier Wingspread classification. In accordance with the Wingspread classification, the anatomical level (low or high type) correlated with different surgical strategies. Interestingly, in the discussion, the authors refer to “simple” cases as “low-type ARMs”. For low-types ARMs the authors of the article used not PSARP, but other surgery, which were coded as “VY-plasty.” No procedures under this name are described in the literature. It can be assumed that we are talking about a cutback procedure with perineoplasty. Thus, the authors of the article effectively used the Wingspread classification, changing only the names “low-type” to “simple” and “high-type” to “complex.” Why?

In 2005, Peña invited three of his associates and 23 unknown pediatric surgeons who had attended Peña's course, where he taught PSARP, to a conference in Krickenbeck. By this time, he had published 42 articles promoting his experience. The Krickenbeck conference recognized PSARP as the ideal procedure for all types of ARMs and modified the classification. It is no longer differentiated between high and low types, as PSARP is recommended in all cases. The conference decision made PSARP the method of choice. The protocol included the classification and a multifactorial method for assessing treatment outcomes. Thus, Peña was one of the first in modern medicine to convince conference participants that his proposal was ideal and, on this basis, all scientific studies that did not correspond to his "experience" were not allowed for publication. A declaration of acceptance of the Krickenbeck classification became the password for publication approval. In the article by Huijgen et al., nothing corresponds to the Krickenbeck classification. But PSARP is presented as the primary method of operation, and the cutback is encrypted beyond recognition. This, along with the password, was sufficient to obtain permission for publication. It's possible that the reviewer modified the article to a form that would allow them to give a positive review.

Thirdly, the statement that "Normally, the PRM consists of two parallel muscles that insert on the pubic bone and converge posteriorly to the rectum" is erroneous, as it is known that the PRM is a loop that encircles the posterior portion of the upper third of the anal canal and its two legs attaches to the pubis. Its contraction is essential for fecal retention. Therefore, before

the introduction of the posterior sagittal approach (PSARP), surgeons in high-types ARMs during surgery pulled the rectum through this loop. Contraction of the PRM causes a decrease in the anorectal angle and anterior displacement of the distal rectum and the anal canal. A large ARA indicates the absence of PRM function. These data are already proven and generally accepted axioms that require no additional references.

During PSARP, the surgeon inevitably transects the loop of the PRM to reach the rectum. This results in the loop being transformed into two unconnected muscular branches, which, when contracting, do not pull the rectum forward, do not block its lumen and do not change the anorectal angle (ARA). That's why ARA looks large. However, this situation cannot be considered normal. The lateral rectal displacement at the PRM level indicates that the rectum is located outside the PRM ring. Thus, both transecting the PRM and passing the rectum outside the PRM ring leads to disruption of the PRM function. It is known that any non-functioning muscle atrophies and decreases in volume. Therefore, the authors' assumption that "intermediate/poor muscle development was more frequently observed in patients with complex ARMs, indicating an association between the severity of ARMs and muscle development", is unfounded. Moreover, as the authors claim, in PRM-preserving surgeries (ASARP and cutback), intermediate/poor muscle development was rarer. From this, it can be concluded that PRM atrophy is a consequence of traumatic surgery. The authors state that: - "Out of the 18 MRIs, 16 were performed preoperatively as part of the preoperative planning" [1]. If they were unable to identify the PRM loop in these 16 cases, the diagnostic accuracy of MRI is questionable. Furthermore, none of the authors' MRI findings were confirmed by other research methods. The authors assessed physiology but did not conduct functional studies. For example, the use of preoperative MRI in infants during periods of increased rectal pressure allows us to prove that the portion of the intestine located caudal to the PRM is a normally functioning anal canal (**Figure 1a, b**).

Fourthly, the following sentence contains two vague definitions: "However, in complex ARMs, the rectum may lie above the PRM, making it challenging to identify the correct plane between the often adherent sides of the PRM for rectal mobilization." It is known that Peña, without any evidence, began to assert that in ARMs the anal canal is absent. In PSARP, the internal anal sphincter (IAS) is being removed under the guise of a fistula or rectal pouch. The exsanguinated and denervated rectum was placed in place of the removed IAS. It follows that after PSARP, the rectum is located both above and below the PRM. Therefore, it is not surprising that the rectum is above the PRM. Neither the text nor the figures show any depiction

of the intestine (or lack thereof) below the PRM. Secondly, if the PRM sides are adjacent to each other, then the rectum is located outside the loop and therefore does not function.

Fifthly, the determination of rectal width is erroneous because: "All MRIs were conducted without prior bowel preparation...and measurements were not standardized for the timing of the last bowel movement." It is known that an empty rectum is closed and has no width. Its width increases with the influx of feces and gases. Therefore, the authors measured not the width of the rectum, but the volume of contents present in it at the time of the examination. To determine the width of the rectum, it is necessary to inject a contrast agent into it up to the splenic flexure. To determine the cause of megarectum, it is necessary to determine the width of the anal canal, including the anus, since megarectum is caused by a discrepancy between the width of stool formed in the wide rectum and the maximum width of the anal canal to which it can open. Only one of the eight figures accurately measured rectal width, which is due to fecal impaction (**Figure 1c**).

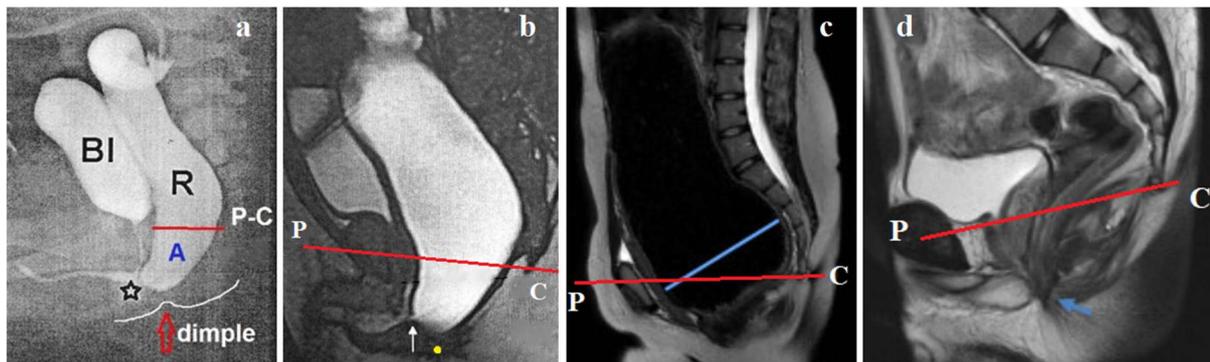


Figure 1. (a) Augmented pressure colostogram in a patient with an ectopy of the anus into the bulbar urethra (asterisk). The anal canal (A) is located caudal to the pubococcygeal line (p-c). At low rectal pressure, the anal canal is closed. Under high rectal pressure (R), it opens as during defecation. The distance from the distal wall of the anal canal to the anal fossa is 2 mm. **(b)** MRI augmented pressure colostogram with an ectopic anus into the bulbar urethra (arrow). High pressure is applied to open the anal canal, which has been close by contracted by the EAS, IAS, and PRM. **(c)** MRI from the article by Huijgen et al., with the caption "sagittal image measuring rectal diameter (blue line, 104 mm)" [1]. I have drawn a red pubococcygeal line (p-c), below which the anal canal should be located. All the anatomical structures responsible for fecal continence are damaged. The cause of megacolon is stenosis of the neoanus. The surgery destroyed the anal canal. No questionnaire can state otherwise. Only resection of the megacolon and encirclement of the distal part of the intestine by the muscle Gracilis can save the situation. **(d)** With the caption "Sagittal image of an anteriorly positioned anus within the external

sphincter (blue arrow). I have drawn a red pubococcygeal line (p-c). The authors refer to the subcutaneous portion of the external anal sphincter as the external sphincter. Why do they not describe other important structures (the deep and superficial portions of the external anal sphincter, PRM, including the unusual masses behind the bowel (white arrow)?

Sixthly, before the introduction of PSARP, functional outcome was assessed using the following formula: ratings were deemed “good” when normal fecal retention and absence of constipation were achieved, “fair” when patients required laxatives or enemas, and “poor” when fecal incontinence and/or uncontrollable constipation occurred. Since these characteristics are objective, i.e., they have clear boundaries and do not require special knowledge to them determine, I applied them to compare functional outcomes in boys with perineal fistulas treated with the cutback procedure and PSARP [2]. After the cutback procedure, good results were observed in 90% of those operated on. After PSARP, all patients had poor results (**Table 1**).

Table 1. Comparison of functional results after cutback and PSARP in boys with perineal fistula

Authors	Good (%)	Fair (%)	Poor (%)
1. Nixon [7]	98	0	2
2. Ackroyd et al. [8]	85	15	0
3. Kyrklund et al. [9]	90	8	2
4. de la Fuente [10]	90	?	?
A) Schmiedeke et al [11]			≈ 60
B) Lombardi et al. [12]			≈ 61.4
C) Stenström et al. [13]			≈ 100
D) Abo-Halawa et al. [14]			?

(1-4) – After the cutback operation; (A-D) – after PSARP.

After 23 years (1982-2005) of massive PSARP propaganda, in which Peña claimed excellent results based on his experience (not scientific research), it turned out that this was not at all the case. Therefore, at the Krickenbeck conference, a multifactorial assessment was adopted, in which a good outcome was defined as "defecatory control." In other words, success was considered the absence of involuntary bowel movements. The Rintala questionnaire is one such attempt. Questionnaires are not scientific instruments because they are completed by children, whose responses are influenced by doctors' assertions that they did not have an anal canal, that the result should be considered good if stool does not fall out spontaneously, and that improvement will occur over time. Responses are influenced by the hope that time will

improve and the desire to thank the doctors for their attention. Acceptable results cannot be achieved after destroying all structures of the anal canal.

Seventhly, the authors demonstrated that many patients after pull-through surgery have impaired PRM function, stenosis often occurs around the rectum retracted into the perineum, and sometimes a portion of the IAS remains unremoved, which they term a fistula. First, they assume a false belief that with ARMs lack an anal canal and therefore have no alternative to pull-through surgery (PSARP, ASARP, and their modifications). Second, they fail to correlate their findings with the pathophysiology of ARMs after surgery. For example, a large anorectal angle is considered not because of damage to PRM, but as an independent characteristic. Therefore, they recommend creating a more acute ARA. Since the PRM is dysfunctional, fibrous tissue develops around the rectum, limiting bowel opening during defecation; megarectum almost always develops, and the levator plates, torn from the rectum, do not create a wide channel during defecation, then creating an acute ARA will further complicate defecation.

Eighth, since 1982, Peña, without any evidence, began to assert that there is no anal canal in ARMs, which was the basis for PSARP. Meanwhile, before 1982, it was known that low-types ARMs have an anal canal and as stated above, preserving the anal canal led to better functional results. In the article by Kraus et al., together with Peña, the following is stated: - "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..." [3]. Peña describes the normal function of the anal canal in patients with "high types" of ARMs (urethral fistulas) but calls it the rectum. The article presents radiographs with a contracted anal canal, with normal function of the PRM. The anal canal opens during high pressure in the rectum. It differs from the norm only by the displacement (ectopy) of the anus into the urethra. Finally, the Arm-Net Consortium stated: "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]. Recently, there has been a trend to perform less traumatic surgeries instead of PSARP [5,6]. Why did the authors cite only Peña's experience and completely ignore scientific data? Obviously, they, following their oath (passport), could not argue against Peña's experience.

Thus, if we consider only scientific facts, and not the mythical "experience" of A. Peña, it becomes obvious that all ARMs have a normally functioning anal canal, which must be

preserved. All pull-through surgeries remove the internal anal sphincter, which is responsible for 50% of fecal continence. The denervated and avascular rectum which serves as a storage for feces but not a continent is lowered on the place, on the place where IAS was. The rectum is separated from the levator plates, which normally open the anal canal to reduce resistance to fecal passage. Due to the separation of the levator plates from the rectum, their function is lost. In PSARP, the PRM, which normally plays a major role in fecal continence, is transected. The deep and superficial portions of the external anal sphincter are transected. Only the subcutaneous portion of the external sphincter, which plays no significant role in fecal continence, not intersect. However, it is damaged because of muscle overstretching and denervation. It responds to skin stimulation but does not participate in the retention reflex (rectoanal inhibitory reflex). Invisible nerve endings of the pelvic organs transected, resulting in the loss of reflex connections between the pelvic organs including the urinary system. Consequently, a perineal fistula occurs after surgery instead of an anal canal. Damage to the urinary system often leads to renal failure. This is especially true for the so-called persistent cloaca, which differs from the true cloaca. In fact, this is an internal anal sphincter ectopy within the vagina with a sharp narrowing of the vagina caudal to the ectopic site. Without understanding the pathophysiology of ARMs and the destructive effects of pull-through surgery, MRI is of no use.

he authors of the peer-reviewed article cite review articles by Peña et al., which lack any evidence and most of the statements contradict known scientific facts. To understand Peña's experience, one must refer to his first article, in which he claimed good results with PSARP [15]. This article, published 2 months after the first one [16]. It additionally included 20 new patients whom Peña operated on in Mexico City (1982), while he had been working in the United States since 1972. To claim good results, a scientist must compare the results of the proposed operation with the gold standard known at that time. Firstly, there were no comparisons. Secondly, at that time, with high-types ARMs, the surgeons pulled the rectum through a loop of the PRM. But Peña claimed that he could not identify the PRM [15], therefore, he did not use the most effective operation. Thirdly, at that time, he did not have any available publications at all. Fourth, the diagnoses of the patients he operated on were so inconsistent with the frequency patterns of different forms of ARMs that it suggests false information. Fifth, Peña began using PSARP for low-type ARMs, claiming that the anal canal is absent in all ARMs, which contradicts scientific evidence.

In addition to PSARP, A. Peña proposed other operations that lacked scientific justification and were not compared with other methods. For example, he proposed performing total urogenital mobilization for the so-called persistent cloaca, without studying the pathological physiology of the urinary system. During the presentation of the article, when W.H. Hendren asked whether he thought this operation would damage the external urethral sphincter, Peña replied: "...my experience in the management of cloaca is that the girls suffer from urinary incontinence don't suffer from the lack of urinary sphincter, but rather because of the lack of contractility of the bladder" [17]. As an analysis of the literature shows, the most severe complications are observed after the operations proposed by Peña for cloaca, which have not been described previously and do not occur unless the urinary tract is operated on [18]. To reduce the number of laxatives used in severe functional constipation, Levitt, Peña, et al. proposed a Swenson-like approach for Hirschsprung disease. After removing 2/3 of the internal anal sphincter, the number of laxatives significantly decreased [19], but as expected, they developed fecal incontinence [20]. The above operations have not been scientifically substantiated. The above operations were not scientifically substantiated. What Peña and his followers call the Peña experience were, in fact, unjustified experiments.

Conclusion

This peer-reviewed article presents evidence of the destruction of pediatric colorectal surgery by Dr. Peña and his followers. Not only the authors, but also at least two reviewers and the editors of the Journal of Pediatric Surgery participated in the decision to publish an article that lacks an understanding of the physiology and pathology of the anorectal area in health and with ARM. It ignores the research of several generations of pediatric surgeons who have proven the existence of an anal canal and achieved good results by preserving it. The article lacks scientific methodology, resulting in incorrect and dangerous conclusions and proposals. In fact, everyone involved in the decision to publish it promotes PSARP, which causes lifelong suffering for patients. Meanwhile, if all elements of the anal canal are preserved, these patients can be healthy.

Why do pediatric surgeons continue to follow Peña's example, ignoring scientific evidence, and continue to perform PSARP despite the poor results? Peña Levitt and his associates have attracted young pediatric surgeons who blindly believe Peña's false claims. Thanks to him, colorectal departments were created, where his supporters' received jobs and scientific specialist status. If they switch to surgeries that trainees can perform (cutback), they will lose

everything they have achieved. Children with ARMs have become hostages of Peña's "experience."

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