Anorectal malformations with visible fistulas from a historical perspective

Michael D. Levin

Email: nivel70@hotmail.com;

Website: http://www.anorectalmalformations.com

The present study was undertaken to compare the concepts of anatomy and physiology of anorectal malformations with visible fistulas, methods, and results of treatment of two historical periods - before (1st) and after (2nd) the introduction of posterior sagittal anorectoplasty (PSARP) into widespread practice. For this purpose, we have selected online articles published in PubMed, starting with the classic study by Stephens [1]. First, he demonstrated the need to preserve the puborectalis muscle (PRM) during defect reconstruction because it plays an important role in stool retention. Second, he proposed the concept of a pubococcygeal (P-C) line, which runs from the lower part of the pubic bone to the distal coccygeal vertebra. He showed that this line corresponds to the location of the PRM, which is located between the rectum and the anal canal. If the blind end of the intestine is located above this line, these cases are considered a high type, and if below the P-C line these cases are a low type [1]. Since then, it was believed that if the gut is located caudally of the P-C line, it means the patient has an anal canal that needs to be preserved during surgery [2,3,4,5]. As the authors of the 1st period emphasize, they considered these types of defect as the anal canal ectopy, although they continued to use the term "fistula". In a low ARM, "the bowel traverses the pelvic floor with its crucial puborectalis sling but fails to migrate back to the normal anal site. Hence, a vestibular vulval or perineal opening (“ectopic anus” or fistula) exists. Like all ectopic openings, these tend to be stenotic" [2]. All pediatric surgeons during the 1st period believed that « in the ectopic anus and covered anus types the bowel traverses the levator before its

abnormal termination. Hence, in this latter, the puborectalis sling, the most important striated muscle involved in continence, retains a correct relationship to the bowel» [1-5].

Operations in 1st period.

In 1960-1982 for "ectopic anus (types: anterior perineal anus; vulvae anus; anovulvar anus; anovestibular fistula) the simple **cut-back** describes Denis Browne is all that is needed to make the imperfect anus large enough to work where it lies. It is, however, important to follow this with daily dilatations for 3 months until the wound is not only healed but supple again. In a newborn size 12 or 13 Hegar an adequate size and when the mother is continuing this at home her fifth finger will usually be of suitable size" [2,6]. Wilkinson confirms that it has been strongly recommended that the low fistula opening somewhere near the lower end of the vagina (vestibular ectopic anus) should be dealt with by a local operation. In the original so-called “cutback operation” one blade of scissors was placed in the fistula and the other across the perineum". "Because the fistula passed through the limbs of the puborectalis sling, if a sufficiently wide channel was made by dilatation, the child was continent" [3]. But he has not used this operation because "when the rectum filled with faeces and faecal impaction led to spurious diarrhoea" [3]. Therefore, with vestibular ectopia, he prefers **abdominoperineal operation**.

I want to analyze Wilkinson's decision to perform abdominoperineal operation in vestibular fistulas from the point of view of the current understanding of the g of the pathophysiology of ARM. The wide rectum and fecal impaction, what led to encopresis, is the result of a late operation. The operation could be postponed provided adequate dilatation of the ectopic anus. Due to the narrow ectopic opening, which could not provide normal emptying of the rectum, a large volume of feces was accumulating in the intestine, which led to the development of the megarectum. Large-diameter feces could not pass through the narrow ectopic

anus, they stretched the PRM and levator plates, which led to the so-called diarrhea (encopresis). Thus, the described complication is not a complication of cutback surgery.

Results of operations in 1st period.

“In the low cases (perineal procedure or dilatation), normal bowel control without the use of regular laxatives is regarded as a “good” result (46%), and “fair” (23%) if supervision was still required to treat constipation or stenosis. A “poor” result denoted incontinence or severe bowel stasis (0%)”. In 31% of cases, too little time passed after the operation to evaluate the results [5].

Dr. G. Willital of Erlangen has reviewed the follow-up records of 300 consecutive patients referred to may unit for anorectal anomalies. In the low abnormalities, normal continence was achieved in 93% of cases (ectopic 90% and covered anus 96%) [2].

Another article was "the long-term results of low anomalies are excellent, as expected, whether primarily treated by cutback or transplantation" [6].

de la Fuente et all described 61 cases of low anal atresia, 42 were females and 19 males. All of them were treated with a "cutback" technique with a follow-up longer than two years. Results are classified as good (90%), regular (8%), and poor (2%) [7].

Currently, the Finnish group of pediatric surgeons continues to carry out the old tactics of treating low anomalies. "All males treated for low ARMs (rectoperineal fistula) with cutback anoplasty, incision of anocutaneous membrane, or dilatations had voluntary bowel movements; 98% of patients were socially continent ($p = NS$); 67% of patients and 64% of controls were totally continent ($p = NS$)". Constipation amongst patients (33 vs 3% in controls; $p < 0.0001$) declined significantly with age. Outcomes by bowel function scores were good at 85% and satisfactory in 15% [8].

The surgical tactics for ARM with a visible fistula was based on the following scientific facts:

A). The puborectalis muscle plays an important role in the retention of feces, which is confirmed by modern research [9,10].

B). If the intestine is located distal to the PRM (pubococcygeal line), then the child has an anal canal, including an internal anal sphincter [11,12,13].

C). Manometric and X-ray studies have shown that the function of this anal canal does not differ from the norm: there are a normal rectoanal inhibitory reflex, normal rectal sensitivity, and a normal defecation reflex [14,15].

D). On the other hand, the good results of treatment with procedures that preserve this anal canal confirm all the scientific premises. It should be understood that satisfactory (not good) results are mostly due to the fact that the operations were performed with an already developed megarectum with secondary damage to the function of the pelvic floor muscles (PRM and levator plates) (**Figure 1**).

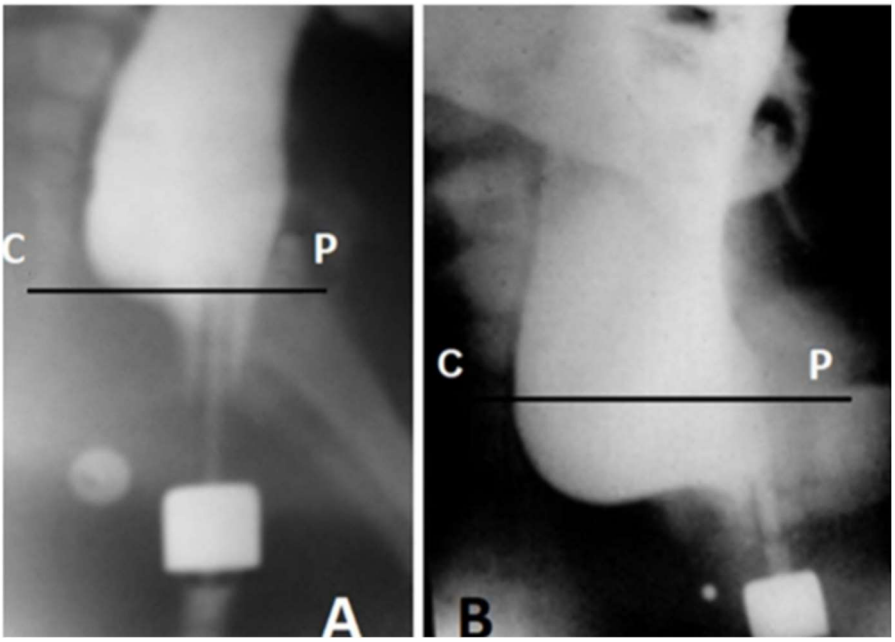

Figure 1. Lateral radiographs of the anorectum made in the same girl with vestibular fistula at a different age. P-C is the pubococcygeal line. The true diameter of the contrast marker strung on the tip of the enema is 1.6 cm. It is located near the fistula orifice. **(A).** At the age of 8 months. Permanent contraction of the anal canal was observed during the barium enema. Its length is 2.5 cm, which corresponds to the age norm. The width of the rectum is 3.4 cm, which is greater than the maximum normal limit (3 cm) (megarectum). Barium penetrates the anal canal behind the tip of the enema. This shows the weakness of the PRM, which does not pull the posterior wall forward. **(B).** At the age of 1.5 years, she had severe constipation and soiling. The width of the rectum is 5.5 cm, which significantly exceeds the maximum limit of the norm for this age (3.7 cm). A megarectum is combined with significant shortening of the anal canal. Its length is 1.9 cm (the minimal limit is 2.3 cm). Conclusion. ARM with vestibular ectopy, megarectum, and descending perineum syndrome.

At that time, controversy existed over the best management of the baby with a low anovaginal or an anovestibular fistula. Stephens and Smith believed that a "cutback" anoplasty with the creation of the "shotgun perineum" usually provides the patient with adequate bowel control and genital function. Other pediatric surgeons advocated a mobilization of the distal anorectum and its transplantation posteriorly with the creation of a perineal body – the perineal transplant anoplasty [16].

Operations in 2nd period.

In 1982, deVries and Peña published their experience in the use of pull-through operation through the posterior sagittal approach proposed by Jean Zulema Amussat in 1835. The first article deals with 34 patients who have been operated on between October 1980 and November 1981 [17]. Another article, published 2 months after the first one, deals with 54 patients operated on for the same period [18]. How could Dr. Peña operate on 20 patients with ARM in 2 months?

Pediatric surgeons saw a huge advantage in this method. After dissection of the posterior muscle group (levator plates and PRM), the rectum was easily detected. Probably many surgeons were surprised because PRM was known to play an important role in retaining feces and passage of the rectum through puborectalis

sling was a necessary element of the operation. Dr. Peña calmed their consciences by stating that PRM was not as important as previously thought. Have you seen Dr. Peña's research anywhere to support this claim? They are not and cannot be, **because it is not true**.

By separating the rectum from this approach, the surgeon removes the part of the intestine that is located between the rectum and the perineum. As it was proved by scientists (see above), it was an ectopic anal canal, while maintaining which good functional results were obtained. Dr. Peña calmed the conscience of pediatric surgeons by stating that there is no anal canal with ARM. Pena and Levitt began to assert that this is a fistula or rectal pouch. It supposedly cannot be saved during correction because there is no sensitivity, ganglion cells, etc. [19]. Have you seen studies with evidence of these claims? They do not exist and cannot be, **because it is not true**.

Why has PSARP become the most popular method for correcting all types of ARM? The reason for this is aggressive advertising.

1. From the beginning until now, Peña and Levitt and coauthors publish numerous articles which describe the remarkable results of PSARP. Of course, they say, some children have chronic constipation and fecal incontinence, but there can be no better results because they initially did not have an anal canal and in most of them the reason for the unsatisfactory results is due to malformations of the spine.

2. As can be seen from the above, the information provided by the authors in numerous articles is not related to science. The aggressiveness of advertising also lies in the fact that these authors control publications in the main pediatric journals. As a result of a special relationship with the editors of these journals, only articles are published that support Peña's false concepts.

3. Peña Course: Workshop for the Surgical Treatment of Colorectal Problems in Children. It acts like an advertisement that gives the impression of the superiority of PSARP.

Scientific papers can be read in other journals, but the authors of these articles can no longer count on publications in pediatric journals. That is why in the scientific community, despite scientific evidence and common sense, the cultivated opinion that PSARP is the ideal method of treatment of ARM, and poor results are due to congenital absence of the anal canal and violation of the nervous regulation.

Ultimately, the most important thing is the functional results of surgical treatment. As shown by researchers using during surgery ectopic anal canal, functional outcomes are close to normal (good at 85% and satisfactory in 15%) at the most rigorous assessment. No bad results reported [1,8].

In a systematic review, Springford et al presented the results of surgical treatment of ARM during the period of predominant use of PSARP. “Twelve studies including 455 patients with a history of anorectal malformation repair were included for analysis. The range of reported prevalence of long-term active problems was as follows: fecal incontinence, 16.7% to 76.7%; chronic constipation, 22.2% to 86.7%; urinary incontinence, 1.7% to 30.5%; ejaculatory dysfunction, 15.6% to 41.2%; and erectile dysfunction, 5.6% to 11.8%” [20].

The most indicative is the comparison of treatment results with perineal fistulas. "Among those with a perineal fistula, incontinence occurred in 42% of the females and in 10% of the males whereas constipation occurred in 62% of the females and 35% of the males". "Sacral malformations were associated with incontinence only in males with rectourethral fistulas [21].

The ARM with vestibular fistulas in girls and with urethral fistulas in boys is low-type anomalies. This means that these patients have a functioning anal canal.

Failure to admit this fact leads to the destruction of the anal canal and a more extensive dissection of the pelvic tissues than with perineal fistulas. This also leads to serious urological problems. Twenty-one (38%) of 55 patients suffered from mucosal prolapse, 18 (32%) patients had had megasigmoid/megacolon. Relevant stenosis of the neo-anus occurred in 13 (42 %) males and 4 (18 %) females, permanent neurogenic bladder dysfunction in 10 (32 %) males and 4 (18 %) females, recurrent urinary tract infections in 10 (32 %) males and 13 (59 %) females. Thirty-seven (70 %) patients had to be reoperated. Forty-one (75 %) patients needed means of aftercare to achieve social continence [22].

"Complete continence (voluntary bowel movement with no soiling) was depicted in only 40%of patients with perineal fistulas, 24% with vestibular, 17%with bulbar rectourethral fistulas, and 0% of patients with cloacal forms. These findings contrast the data provided by Pena and Levitt, who reported continence rates of 89% (perineal), 64% vestibular), 46% (bulbar, and 13-37% (cloacal)" [23]. Hashish et al showed that stooling patterns are perceived to worsen with age. This suggested that children with ARMs need long-term follow-up and counseling [24]. There was no statistically conclusive evidence that tethered cord by itself affect the urinary or fecal control in ARMs patients [25].

The ARM with perineal fistula was chosen by me to compare treatment outcomes. But a functioning anal canal is present in all females with a vestibular fistula (**Figure 2).**

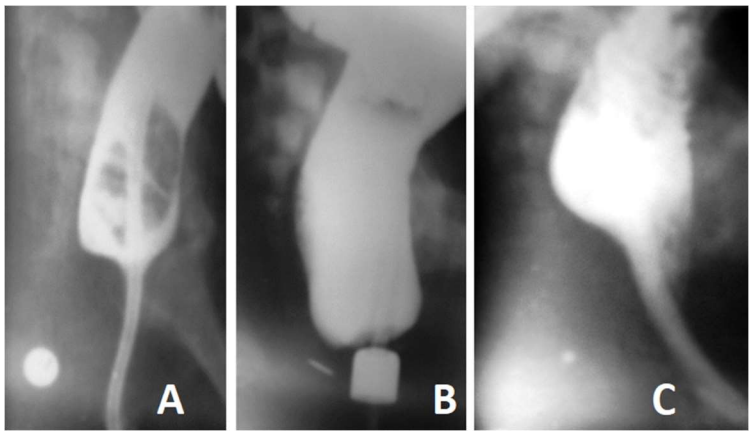

Figure 2. ARM with vestibular ectopy. (A). At rest, the anal canal is in constant contraction around the catheter, preventing barium from leaking. (B). During an attempt to defecate, a wide opening of the anal canal occurred because of the contraction of the levator plates. (C). Resulting in relaxation of the IAS the penetration of barium into the upper anal canal in front of the catheter occurred. At this time, the posterior wall is pressed against the catheter by the contracted PRM. Manometric tracking reveals a short-term pressure drop in the upper part of the anal canal (rectoanal inhibitory reflex) [14,15].

As shown in the article Kraus et al [26], in 90% of boys with urethral fistulas, the distal bowel is located below the last sacral vertebra, i.e., they also have a functioning anal canal (**Figure 3**).

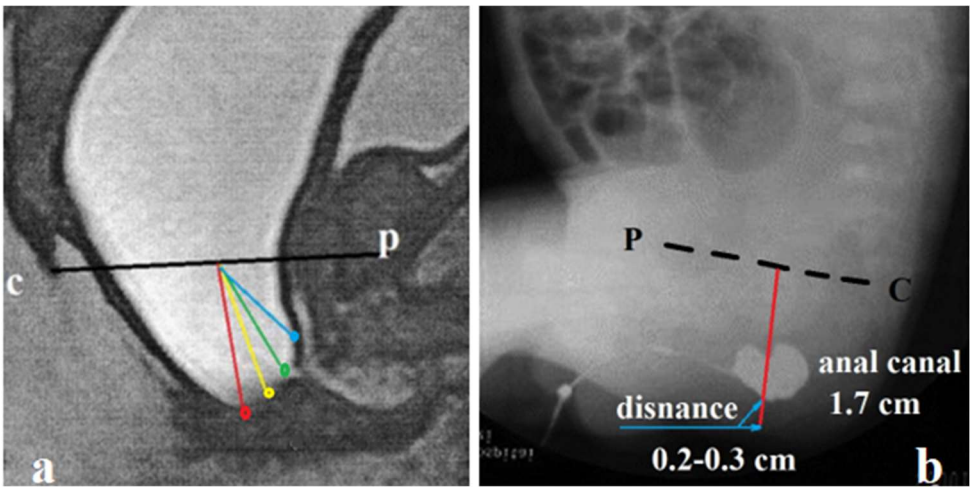

Figure 3. Low ARM (**a**). MRI with augmented-pressure distal colostogram in male with anourethral ectopy. The normal position of the anus is red, perineal ectopy is yellow, bulbar ectopy is green, and prostatic ectopy is blue. (b). Fistulography in newborn male through the fistulous opening on the sagittal raphe under the scrotum. The contrast agent depot is determined in the opened anal canal 2-3 mm from the anal dimple.

It is known that the anal canal is formed as a result of the fusion of the proximal endogenous and distal exogenous primordia. All low ARMs are characterized by the absence of canalization of the distal anal canal. The endogenous anlage moves caudally in the perineal tissues, forming the internal anal sphincter. Not meeting an exogenous channel on its way, it deviates anteriorly until it penetrates some cavity. Outside the perineal tissue, the canal acquires the property of a fibrous canal of various lengths - from 2 mm when penetrating through the skin and

subcutaneous tissue in perineal fistulas, to several centimeters in scrotal or penile fistulas. It is important to understand that the formation of an ectopic anus is the final stage in the formation of a defect. By this time, in all the low anomalies, a functioning anal canal had already formed.

As shown by Stephens [1], a significant number of patients with vaginal fistula, including patients with the so-called persistent cloaca, also have an anal canal, because the ectopic anus is located below the P-C line (Figure 4).

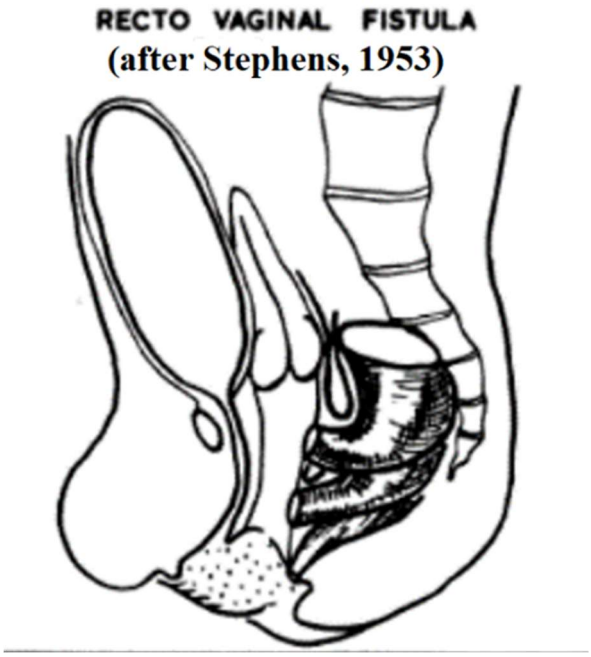

Figure 4. The scheme of the ARMs with vaginal fistulas [1].

Comparing the results of the treatment of perineal fistulas of the two periods, we can confidently assert that (1) the results of PSARP are significantly worse than those that preserve the anal canal, (2) surgery that destroys the anal canal, but not spinal abnormalities, is responsible for the poor function of the anorectum, (3) Painful lengthy bowel management programs are a temporary solution. They cannot normalize anorectal function. And resection of the dilated rectosigmoid is also a temporary victory. The reason for the patient's suffering after PSARP is the absence of the anal canal which is irreplaceable. Therefore, there is no need to torment the child and his family. It is necessary to cleanse the colon with minimal

means, realizing that this is a problem to all life. For example, the bowel management program of Märzheuser et al. includes the following. "These patients received oral polyethyleneglycol to evacuate stool impaction. Then anorectal irrigation was initiated and repeated every 24 or 48 hours" [27].

Children and adolescents with low ARM do not differ regarding their QoL, even though they have impaired bowel function and worse emotional functioning compared to the healthy control group [28]. Numerous articles with similar patient responses reflect hope that fades with age. Secondly, from the very birth, the parents were told that the child was born without an anal canal with spinal defects. But surgeons will do their best to prevent feces from fell out of the rectum. Therefore, children who have undergone repeated surgeries (70%), difficult experiments with high doses of Senna, with complications of a senseless antegrade enema, and who continue to maintain their intestines with frequent flushing throughout their lives, are happy that they are socially active. And if they find out that they were born with a functioning anal canal and their entire difficult childhood was meaninglessly disfigured, how will they feel, and what should they do?

The results of pull-through operations (posterior sagittal anorectoplasty, anterior sugittal anorectoplasty, and laparoscopic-assisted anorectal pull-through) differ little from each other. Anorectum is a very complex functional structure. Fortunately, most patients with ARM have a functioning anal canal that can provide acceptable functions fecal retention, and defecation. Extirpation of IAS or even simple isolation of it from the surrounding tissues is an irreparable loss of the function of retaining feces. The rectum cannot replace IAS, because these are completely different functional structures. And denervated and devascularized rectum - even more so. Normal retention of feces cannot be expected after damage to the PRM. After separation of the rectum from the levator plateaus, there can be no reflex defecation. The intersection of invisible

nerve fibers breaks the reflex connections of the pelvic organs, as a result of which the reflexes die forever. The surgery of the patients with ARM is not handicraft. In order to achieve good functional results, it is necessary to preserve all the elements of the anal canal.

First of all, you need to make sure that the patient has an anal canal. During X-ray control, you need to create high pressure in the rectum. If the gas or contrast agent approaches the perineal skin, the anal canal is functioning normally [30,31]. The choice of the method of surgery preserving the anal canal depends on the presence or absence of a visible fistula [32].

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