Anorectal malformations

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Pathological physiology, by definition, is a damaged physiology of an organ or system. To determine the pathophysiology of anorectal malformations, it is necessary to know the normal physiology and anatomy of the anorectum.

I. **Anatomy of the anorectum.** The rectum is in the pelvis retroperitoneally. It starts from the third sacral vertebrae and ends at the level of the pubococcygeal line, where it borders on the anal canal. From a functional point of view, the rectum begins caudal to the rectosigmoid sphincter. While fecal retention, it performs a cumulative function, and during bowel movements its strong peristaltic wave, which starts from the rectosigmoid sphincter, expels the stool through the open anal canal. Table 1 shows the normal width of the rectum and the anal canal in patients of different ages after filling the colon with barium at least to the splenic angle [1,2].

Table 1. The normal size of the rectum and anal canal in different ages.

Ages	The width of the rectum (cm)	The length of the anal canal (cm)
5 days – 11 months	1.3 – 3.0 (2.24±0.09)	1.7 – 2.5 (2.21±0.15)
1 – 3 years	3.0 – 3.7 (3.21±0.11)	2.3 – 2.8 (2.55±0.10)
4 – 7 years	3.0 – 3.9 (3.43±0.14)	2.5– 3.6 (3.17±0.14)
8 – 10 years	3.2 – 4.1 (3.72±0.05)	2.6 – 3.7 (3.11±0.10)
11 – 15 years	3.6 – 4.6 (3.95±0.07)	3.1 – 3.9 (3.43±0.10)
23 – 64 years	3.5 – 4.8 (3.95±0.21)	3.4 – 4.2 (4.08±0.07)

The anal canal starts from the pubococcygeal line and continues to the anus. Its length is given in table 1. The anal canal consists of several elements, each of

which plays an important role in the fecal retention and defecation. In its center is the smooth muscle of the internal anal sphincter (IAS), which is a continuation of circular layer of the rectum. However, IAS are both morphologically and functionally different from the circular layer of the rectum. Alamovich et al. investigated the normal innervation of the internal anal sphincter. This study shows that the IAS itself has no autonomous innervation unlike the rest of the digestive tube [3]. During a bowel movement, it opens all along the length, completely and simultaneously. For 24 hours a day, except for a few seconds, during a bowel movement, IAS is in a closed position, preventing leaking of the stool and gas. The rectal basal pressure is low (2-5 cm H_2O), and anal canal pressure is high ≤ (42.43±8.9 mmHg).

The motor function of the colon and rectum obeys the law of the intestine (Bayleys and Starling's gut law), stating that a stimulus within the intestine (that is, the presence of food, feces, balloon) initiates a band of constriction on the proximal side and relaxation on the distal side and results in a peristaltic wave. This gut reaction is under control of the intramural nervous system and therefore can be caused in the intestine, isolated from the surrounding tissues [4]. The anorectal inhibitory reflex describes the relaxation of IAS and the contraction of EAS with PRM in response to stretching of the rectal wall. It depends both on the intramural nervous system (it is absent in Hirschsprung's disease) and on the neural connections of the rectum with striated sphincters. Therefore, after gut denervation, this reflex is absent. After a pull-through operation (posterior, anterior etc.) can be detected only relaxation of IAS. However, the basal pressure and depth of relaxation of the intestine are always significantly less than the norm. Due to the denervation of the intestine, the striated sphincters do not contract.

The loop of the puborectalis muscle (PRM), consisting of striated muscles, is also involved in the retention of feces. During its contraction, the posterior wall of the anal canal is attracted to the pubis, which contributes to the increase of the

anal pressure and is accompanied by a decrease in the anorectal angle. All three portions of the striated muscle of EAS (deep, superficial and subcutaneous) during contraction increase the tone of the anal canal [4, 6].

The two striated muscles of the levator plates (LP) are attached on the periphery fanlike to the bones of the pelvis, and the inner ends through the rectum to the longitudinal layer of the anal canal. Levator plates are contracted during bowel movements. They stretch of the anal canal, which leads to a wide its opening during the passage of the stool [7].

Nervous regulation of anorectum is very important. It includes sensitive elements in the rectum and in the anal canal, as well as 3 systems: (1) intramural nerve system of the rectum (myenteric ganglia, cells of Cajal), which coordinates the motility of the intestine; (2) sympathetic and parasympathetic innervation, which coordinate reflexes и (3) somatic innervation, originating from Onuf's nucleus, which is located in the sacral spinal cord, travel in the pudendal nerve, muscular branches and in the coccygeal plexus. Through them, cortical and spinal effects on the anorectum function occur (Figure 1) [5]. An example of the interaction of all neural networks can serve as an act of defecation. When the pressure in the rectum reaches the threshold level of defecation, this information is recorded by the sensitive elements of the rectum and transmitted through the networks. Through the intramural nervous system, the excitation of strong rectal motility and relaxation of IAS occurs. At the same time, there is a reflex relaxation of PRM and EAS, as well as a contraction of LP. Through somatic innervation supply striated muscles are under voluntary control, i.e. the final decision on defecation depends on whether it is possible in the present situation.

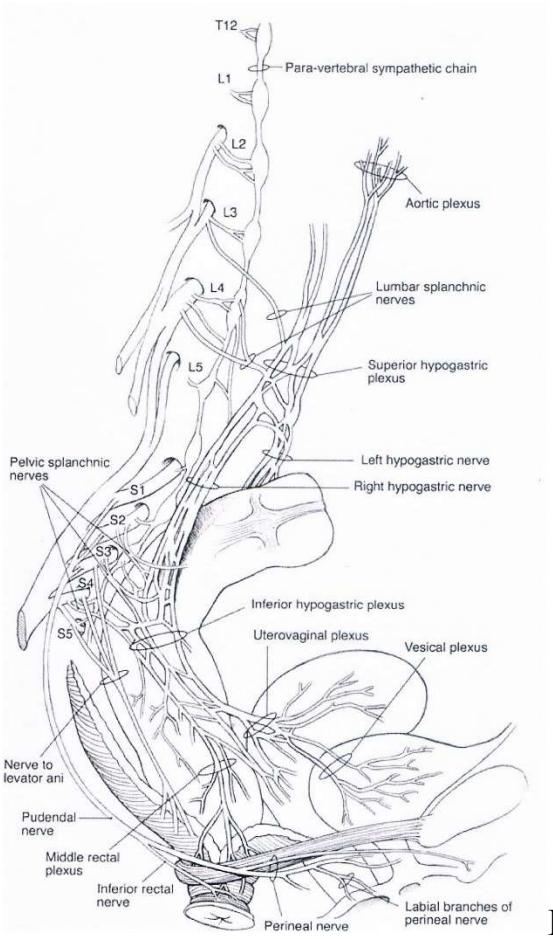

Figure 1. Scheme of the nervous supply anorectum from the article Bharucha [5].

It is obvious that damage (intersection) of the nervous supply will inevitably lead to dysfunction of the anorectum.

Ligamentous apparatus creates a stable framework for the normal function of all elements of the pelvic floor [8].

Normal anorectal physiology. There is not a single muscle that would provide a long overlap of the anal canal. Nevertheless, the anal canal is in continuous contraction around the clock. It is known that smooth muscle of IAS is capable of a longer contraction than striated muscle. Meanwhile, the contraction power of the PRM, EAS and LP is dramatically reduced after a few seconds (5-15). Coordination between the nervous systems allows the muscles to retain the feces with the necessary (optimal) force of contraction, depending on the amount and

consistency of the contents in the rectum, i.e. with minimal power of contraction with an empty rectum and stronger with a filled rectum, and with fluid contents.

Skeletal muscles are capable of two types contraction: tonic and mechanical. Tonic prolonged contraction of the LP, PRM and EAS explained by postural reflex [9,10]. Each nervous axon has a connection to the muscle fibers scattered throughout the muscle. Therefore, even a small amount of the contracted muscle fibers results in a contraction of the whole muscle. The muscle tone is dependent on the number of fibers participating in the contraction, i.e. from the percentage of axons activating muscle contraction. The prolonged, tonic contraction is due to the continuous replacement of the axons activating the different groups of muscle fibers. At different time the different groups of the muscle fibers are contracted. During the contraction of one group, other groups of fibers restore ability to contract [11,12].

In mechanical contraction all fibers are involved, resulting in a significant shortening of the muscle. However, the duration of the muscle contraction is severely limited, typically less than one minute.

1. Fecal retention. At rest, the IAS and striated muscles of the pelvis floor are in a state of tonic contraction. They help to support the pelvic organs and participate in the continuous retention of feces. The pressure in an empty rectum is equal to the intra-abdominal pressure (IAP). During long-term fecal retention, while entering the rectum of a certain volume of feces, there is a periodic increase in rectal pressure above the IAP. This pressure we called the threshold pressure of the first order (TP-1). It causes a reflex relaxation of the IAS and contraction of the EAS and PRM (anorectal inhibitory reflex). The PRM pulls forward the upper part of the anal canal. In front, inside the PRM loop the anal pressure decrease as a result of the IAS relaxation. Between the rectum and anal canal there is narrow opening through which the gas and liquid feces can penetrate the upper part of the anal canal. In the mucosa at this level there are sensors that allow to

distinguish the liquid from gas. In the presence of gas is enough to strain the abdominal wall and diaphragm to increase rectal pressure and expel gas through the lower part of the closed anal canal. When the liquid feces penetrate the upper part of the anal canal the tone of the EAS increase, which leads to a contraction of the IAS and crowding out of the fluid from the anal canal into the rectum. During anorectal inhibitory reflex the formed stool remain in the rectum due to acute anorectal angle and the narrow hole between the rectum and anal canal. After a few seconds the rectum adapts to the new volume of the stool and relaxes. The rectal pressure drops up to IAP, resulting in the contraction of the IAS and relaxation of the PRM and EAS. After entering the rectum of another portion of feces this picture (anorectal inhibitory reflex) is repeated. This picture can be observed up to seven times per hour [13].

When the volume of stool in the rectum reaches a certain value, the rectal pressure rises from TP-1 to the threshold pressure of the second order (TP-2), in which a need for a bowel movement appears. If this need is does not coincide with the possibility of its implementation, the rectum continues to relax to a limited extent. At the same time, there is an increase in the tone of the recto-sigmoid sphincter, which prevents the further penetration of feces from the sigmoid colon into the rectum.

2. Defecation. When the need for a bowel movement coincides with the possibility of its implementation, a straining of the abdominal wall and diaphragm increases the abdominal and rectal pressure from TP-2 to the threshold pressure of the third order (TP-3). At the rectal pressure TP-3 the reflex defecation takes place: a strong peristaltic wave of the rectum expels stool through the open anal canal. Wide opening of the anal canal is due to relaxation of the IAS, PRM, and EAS, with a simultaneous contraction of the LAM. Any of pressure levels depend not only on the volume of feces, but also from the tone of the rectum. During the opening of the anal canal, its wall is stretched at the level of deep and superficial

portions of the EAS. The subcutaneous portion is relaxed, but it is not connected with LAM. Therefore, during the evacuation of soft feces, it forms a tape, the diameter of which depends on the viscosity of the feces.

II. Etiology of ARM

The etiology of anorectal malformations (ARM) is known only in general terms. It is known that both genetic and environment factors may contribute to the multifactorial etiology of ARM [14]. In recent years, have been found that rare CNVs (including 79 genes) and SNPs to genetically contribute to ARM. Out of disrupted 79 genes one such putative gene is DKK4. Down regulation of CDX-1 gene has also been implicated in isolated ARM patients. In syndromic ARM de novo microdeletion at 17q12 and a few others have been identified [15]. Consistently increased risks were observed for paternal smoking and maternal overweight, obesity and diabetes, but not for maternal smoking and alcohol consumption. But only few of these studies reported on the same risk factors [16].

III. Embryology of ARM.

The defective development of the cloaca results in anorectal malformations. However, the developmental and pathogenic mechanisms of ARMs are unclear [17]. According to Fritsch et al data of normal human development, both anorectal malformations occur during the postcloacal period and seem to depend on the interaction between epithelial and muscular development [18]. A study by Kruepunga et al shows that the anorectum, derived from three different regions: the caudal hindgut, the dorsal cloaca and the anal pit. The junction of the cloister (endoderm) and the anal pit (ectoderm) is the final stage of the creation of the anal canal [19].

In the literature there are two opinions about the time of occurrence of ARM. Some researchers, without convincing evidence, believe that ARMs are formed

as a result of disruption of the development of the cloacal membrane. At the same time, not even the presumptive stages of such pathological development are described. For example, "...the cloacal membrane is always too short in its dorsal part, thus, the dorsal cloaca is missing; and as a result, the hindgut remains attached to the sinus urogenitalis, forming the recto-urethral fistula" [20].

Other researchers believe that ARM occur during the postcloacal period. This view of fetal development, which leads to the emergence of ARM, has its supporters among clinicians.

It is known that the rectum and the upper part of the anal canal is formed from the endoderm, and the distal part of the anal canal is formed from the ectoderm. Normally, in the post-cloacal period, the endodermal internal anal sphincter migrates in the cranio-caudal direction to meet with the ectodermal portion [21]. With ARM, there is no hole in the anal fossa, which is convincing evidence that the ectodermal element of the anal canal has not been activated in the embryonic period. Not having met in its path ectodermal part of the anal canal, proximal part of the IAS continues to migrate with the deviation forward. It has a high proteolytic activity. Penetrating through the subcutaneous tissue and skin, it creates a perineal fistula. Penetrating through the wall of the vestibule, it forms a vestibular fistula.

IV. **Histology of ARM.** In a study by Holschneider et al. it was shown that in patients with ARM "Classical aganglionosis was found in 31% of the rectal pouch specimens, hypoganglionosis in 38%, neuronal intestinal dysplasia (NID) type B in 14%, and dysganglionosis in 10%". In the authors' opinion, "...the recommendation to use the distal rectal pouch and parts of the fistula in the reconstruction of anorectal malformations should be reconsidered" [22]. These histological results were confirmed by other researchers, who believed that the

histological structure of the rectal pouch should be consistent with the structure of the rectum [23,24]. Xiao et al., also concludes that the distal rectal pouch has distinct defects in the neuromusculature and need to be resected for better functional outcomes of the remaining gut [24]. First, like the previous authors, they compared the tissue specimens of the rectum, taken from 2 to 4 cm above the dentate line in the control group, with the tissue specimens, taken in the ARM group from 0.5 to 2.0 cm of the most distal part of the rectal pouch, i.e. compared the samples from the anal canal with the rectal samples. Secondly, they didn't consider that in the postnatal period there are changes in the terminal part of the intestine, associated with chronic constipation due to stenosis of the ectopic anus.

Alamovich et al. investigated the normal innervation of the internal anal sphincter. This study shows that the internal sphincter itself has no autonomous innervation unlike the rest of the digestive tube [3]. Thus, the research of Holschneider and other authors proves that what they called the "rectal pouch" is the anal canal. Lambrecht and Lierse in neonatal pigs with ARM found that the proximal region of the fistulae in ARM has most features of a normal anal opening: (1) it is surrounded by an internal sphincter, (2) the rectal pouch in the region of the internal sphincter as well as the fistulae are hypoganglionotic, (3) the proximal fistula region is lined by transitional epithelium, and (4) it contains anal glands. They, therefore, consider that the fistula should be designated as an ectopic anal canal. The most important result was the demonstration of a normal internal sphincter even in high and intermediate types of ARM [25]. Rintala et al have shown that in anorectal malformations the distal rectal pouch with the fistulous connection is an ectopic anal canal [26].

Conclusion. The anal canal is a unique structure that has no analogues in the body. It can be said with absolute certainty that in most cases the ARM there is a

functioning anal canal that needs to be preserved in order to obtain the best functional result.

V. Pathological anatomy and physiology of anorectal malformations

1. **Historical aspect.** In 1953, Stephens 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 high type of ARM, if at the level of this line it is an intermediate type, and low type if the intestine is located more caudally than this line [27]. Since then, it was believed that if the gut is located below the P-C line, it means the patient has an anal canal that needs to be preserved during surgery. "The results in low deformities operated by perineal rectoplasty showed "good" clinical scores, good preservation of the rectoanal reflex, and good electrical activity of the external sphincter [28]. Low type anomalies were treated by neonatal perineoplasty, anal transplants or cut-back. In patients with the vestibular ectopy "The sling of the puborectal muscle was well identified by causing contraction of the puborectal muscle with an electric stimulator". Patients with low type had good results in 100% [29].

In 1982, deVries and Peña published their experience in the use of pull-through operation through posterior sagittal approach proposed by Jean Zulema Amussat в 1835 году [30,31]. The first article deals with 34 patients have been operated between October 1980 and November 1981. The results of 12 cases after the colostomies closed are given in the tables: eight have had excellent results and in 4 patients, the results were unsatisfactory [30]. Another article, published 2 months after the first one, deals with 54 patients operated on for the same time period. All cases are summarized in the table (Table 2. A) [31]. An additional 20

cases of ARM appeared, that were operated during the same time period, which raised doubts about the reliability of the work. Therefore, we compare the data of this article with reliable statistical research. In the table 2.B. the statistical data from table 2. A. we compare with the first results of a European multi-center registry of patients with anorectal anomalies. This article provides a statistical analysis of 200 cases of patients with ARM registered in 14 European centers in 2007-2012 [32].

POSTERIOR SAGITTAL ANORECTOPLASTY

Table 1. Number of Patients and Type of Defect

Male	
Urethral fistula	25
High malf., no fistula	4
Vesical fistula	3
Low malformation (Perineal fistula,anal stenosis)	4
Complex malformation*	1
Secondary operation	2
	39
Female	
Vaginal fistula	1
High malf., no fistula	1
Low malf., no fistula	1
Rectal atresia and stenosis	2
Rectovestibular fistula	2
Complex malformation†	7
Secondary operation	1
	15

*Case #3, rectal atresia.
†Includes the cloacas, rectal atresia and rectal stenosis.

A

POSTERIOR SAGITTAL ANORECTOPLASTY

Table 1. Number of Patients and Type of Defect

Male		(%)	(%) [27]
Urethral fistula		25 (64)	(29)
High malf., no fistula		4 (10)	(15)
Vesical fistula		3 (8)	(6)
Low malformation (Perineal fistula,anal stenosis)		4 (10)	(43)
Complex malformation*		1 (2)	
Secondary operation		2	
		39 (72)	(49)
Female			
Vaginal fistula	(cloaca)	1 (7)	(8)
High malf., no fistula		1 (7)	(4)
Low malf., no fistula		1 (7)	
Rectal atresia and stenosis	(perineal fistula)	2 (13)	(41)
Rectovestibular fistula		2 (13)	(28)
Complex malformation†		7	
Secondary operation		1	
		15 (28)	(51)
	Low type	(15)	(63)
	High type	(67)	(31)

*Case #3, rectal atresia.
†Includes the cloacas, rectal atresia and rectal stenosis.

B

Table 2. Comparison of statistics from article of Peña and deVries [31] with the results of a European multi-center registry (red) [32].

Firstly, the number of patients operated on for one year at two centers (54) is disproportionately high compared with 203 patients at 14 centers for 6 years. Secondly, the ratio of patients by gender and different types and forms of ARA are known. They are the same in different countries and continents. These relationships are repeated in the European multi-center article and are fundamentally violated in the Peña and deVries article. The appearance of an

additional 20 cases and statistical contradictions cannot be explained from a scientific point of view.

The posterior sagittal anorectoplasty (PSARP) allowed surgeons easily to view the rectum but does not allow to see the anal canal. In all cases of ARM, regardless of the level (low or high), a PRM dissected and an "anal canal" is created from the rectum. The effect of the PSARP can be judged by comparing the results. After neonatal perineoplasty, anal transplants or cut-back in patients with a low type of ARM a good result was in 100% during 10-30-year follow-up [27]. Peña and deVries announced excellent results in 8 of 54 patients with short term follow-up (less than one year). In 4 patients were unsatisfactory results associated allegedly with severe sacral anomalies and nerve deficits. In the rest of the observations, the colostomy was not closed. It was a typical advertisement and the key word in this purely promotional article is “excellent”. It was a typical advertisement and the key word in this purely promotional article is “excellent”. As a result of massive propaganda, unfounded statements began to be perceived as true. I quote them from the article by Levitt and Peña [33]:

1. “…the most distal part of the rectum in these children is surrounded by a funnel-like voluntary muscle structure that keeps that part of the rectum collapsed and empty. The intraabdominal pressure must be high enough to overcome the tone of the muscles that surround the rectum …”

2. “Except for patients with rectal atresia, most patients with anorectal malformations are born without an anal canal; therefore, sensation does not exist or is rudimentary”.

3. “Patients with anorectal malformations have abnormal voluntary striated muscles with different degrees of hypodevelopment. Voluntary muscles can be used only when the patient has the sensation that it is necessary to use them. To appreciate that sensation, the patient needs information that can only be derived

from an intact anal sensory mechanism, a mechanism that many patients with anorectal malformations lack".

4. "It was learned through this procedure that the external sphincter is a functionally useful prominent structure. No puborectalis sling, as such, could be identified" [30].

It should be emphasized that the authors did not conduct any studies to prove these statements and did not cite references to the works with such evidences.

1. Scientific research.

A). X-ray examinations

In this paper, I cite only studies of patients with fistula on the perineum, vestibula, urethra and without the fistula, since the results of studies of other forms of ARM (in bladder, vagina, and cloaca) are not sufficiently straightforward. In patients with visible fistulas (perineal and vestibular), the distal intestine at rest is in a closed state. Its length in children of the first months of life without a serious megarectum is equal to the length of the normal anal canal. During a barium enema, penetration of the contrast agent into the upper part of the anal canal in front of the enema tip can be seen intermittently. At this time, the posterior wall of the anal canal at this level is pressed against the tip of the enema by the contracted PRM. It is the radiological equivalent of the anorectal inhibitory reflex. During a bowel movement, the anal canal opens. At this point, the caudal wall of the anal canal is approaching the anal fossa. The distance between the wall of the anal canal and anal dimple varies from 2 to 5 mm, depending on the age. It is equal to the thickness of the skin and subcutaneous tissue (**Figure 2**) [34].

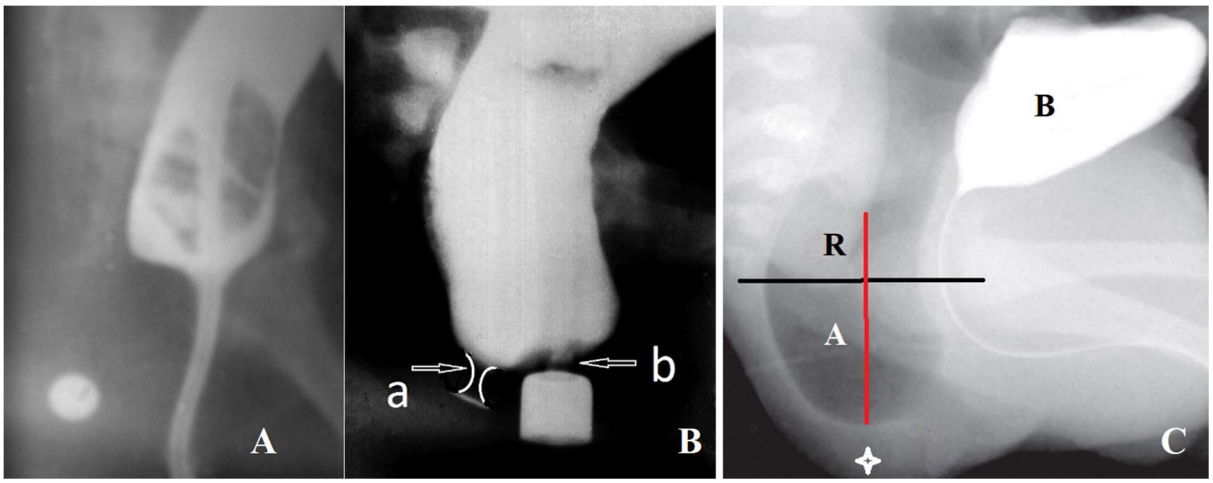

Figure 2. (**A-B)** The radiographs of the same girl with vestibular fistula performed at different ages. (**A)**. At the age of 3 months the rectum was filled with barium through the catheter, conducted through the fistula. A pushpin is located near anal dimple. The distal intestine, with a length equal to the length of the normal anal canal, constantly contracted around the catheter, preventing leakage of barium. **(B)** At the age of 9 months, during a barium enema the wide opening of the anal canal occurred. The distance from the pushpin to the distal wall of the open anal canal equals 4 mm (a). Barium does not penetrate outward, since the tip of the enema occluded the narrow and rigid ectopic anus (b). The true diameter of the marker on the enema tip is 1.6 cm. The width of the rectum is 4.3 cm at the maximum rate for children under 1 year is 3 cm. Conclusion: ano-vestibular ectopy, megarectum. The diastasis between the anal canal is (4 mm) the thickness of the skin and subcutaneous tissue. **(C)** Radiograph of a newborn boy with perineal fistula during a reflex opening of the anal canal. The distance from the blind end of the anal canal to the anal dimple is 2 mm. P-C line (black). Axis of anal canal (A) and rectum (R) (red line).

In patients without visible fistula, we see a similar X-ray picture. An increase in rectal pressure causes opening of the distal part of the intestine with the wall approaching the anal dimple (**Figure 3**).

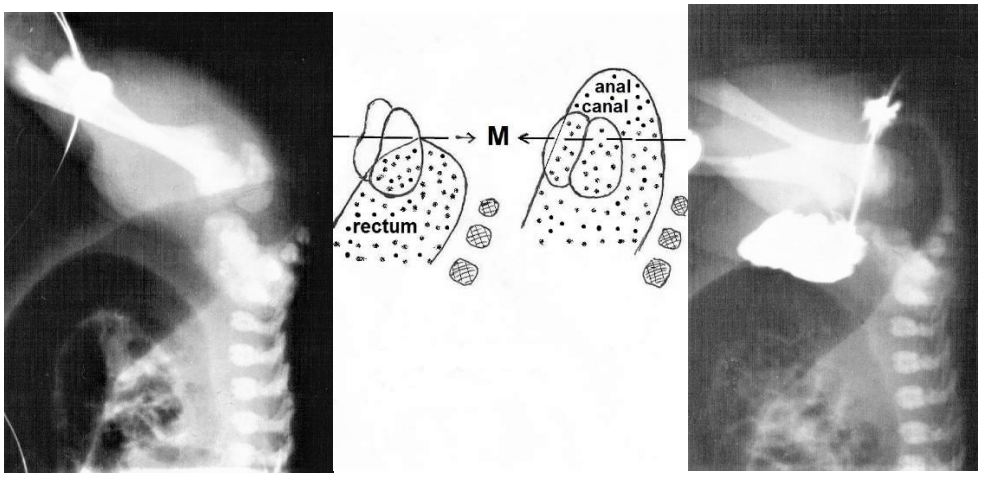

Figure 3. Examination of a newborn without visible fistula and a diagram to it. A. At rest. The distal intestinal wall is at the level of the "M" line. B. After attempting to inject a contrast substance into the rectum. The contrast agent was introduced into the soft tissues of the perineum. At this time, the anal canal opened, and the gas approached the injection site. Through the distal third of the sciatic bones, which have a pear-shaped form, a line "M" is drawn, which corresponds to the location of the P-C line.

Opening of the anal canal is seen at augmented-pressure distal colostogram. It is very important, firstly, that the anal fossa is located on the axis of the anal canal. Secondly, the ano-urethral fistula is in the distal part of the anal canal near the anus (**Figure 4**). Third, the fistula itself is a hole in the junction of the wall of the anal canal with the wall of the urethra and its length is equal to the thickness of these walls (**Figure 4. B**). In this case, it is appropriate to provide a description of augmented-pressure distal colostogram by Levitt and Peña : "…the most distal part of the rectum in these children is surrounded by a funnel-like voluntary muscle structure that keeps that part of the rectum collapsed and empty. The intraabdominal pressure must be high enough to overcome the tone of the muscles that surround the rectum …" [35]. An amendment should be made because there are no muscles around the rectum. They are around the anal canal, which is located caudal to the R-C line.

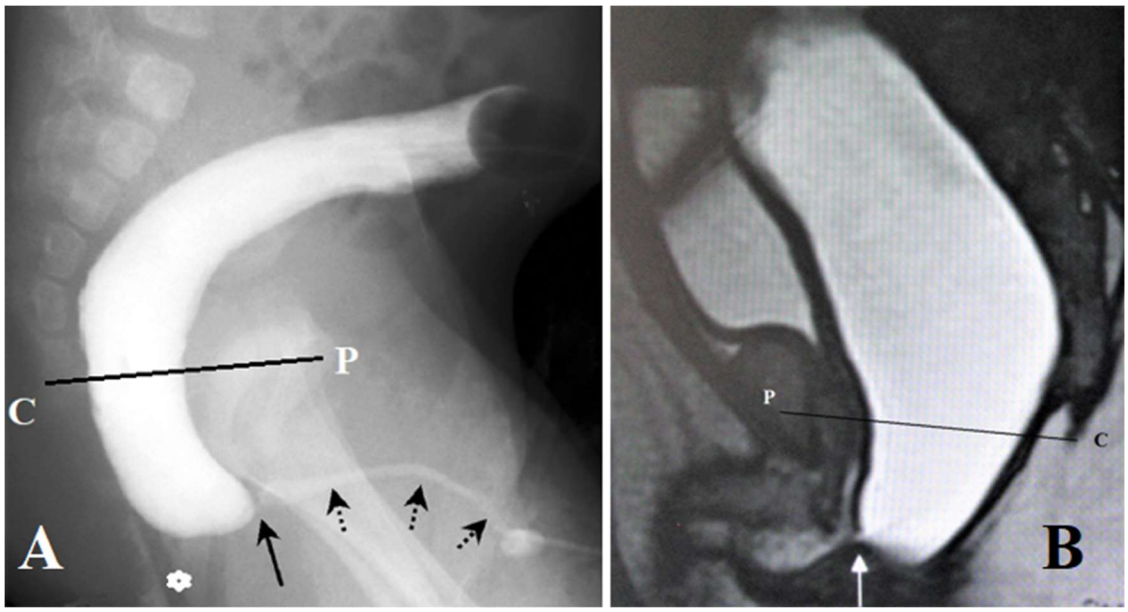

Figure 4. Examples of the X-ray (A) and MRI (B) augmented-pressure distal colostogram. In both cases during high pressure in the rectum, the contrast agent penetrated the wide anal canal. The fistula is in the distal part of the anal canal.

Thus, in patients with ARM, the distal intestine is in constant contraction. Its length is equal to the length of the anal canal. The anorectal inhibitory reflex arises in it. At the time of defecation or increase of rectal pressure to a certain level (compression of the abdomen) there is a wide its opening to the width of the rectum. This segment has all the characteristics of the anal channel. It differs from the norm by displacing the outlet opening forwards and varying degrees of its narrowing.

Knowing the width of the rectum and the length of the anal canal (see table 1), it is possible to approximately calculate the volume of the cylinder (open anal canal) that appears in the tissues of the perineum during compression of the abdomen. Its volume increases from 1.5 cm^3 in newborns, to 40 cm^3 in children 15 years of age. Since there are no voids in the tissues of the perineum that this cylinder having the diameter of the rectum could fill, it can be concluded that this is the result of the levator plates contraction. Therefore, these cases are an ectopy of the anal canal.

B). **Manometric research**

In ARM with visible fistulas, a manometric study was unavailable due to the narrow and rigid ring of the ectopic anus, through which it was impossible to pass a rectal balloon. We performed a manometric study without a rectal balloon. To create a high pressure in the rectum, we sharply injected 50 cm 3 of air into the rectum. Anorectal inhibitory reflex was found in all patients in whom it was possible to carry out a measuring device into the rectum. Basal pressure in the anal canal was within the normal range [34]. Ruttenstock et al produced preoperative rectal manometry of rectoperineal or rectovestibular fistula. It showed the presence of functional anal structures within the fistula in all patients. Complete transposition of the fistula was achieved in all patients. All patients had voluntary bowel movements, with no incontinence or soiling. They speculated that fistula-preserving surgery in patients with anorectal malformations is associated with improved bowel function outcome [36].

C). **The overall picture is as follows:**

In the early embryonic period, the internal anal sphincter penetrates the tissue of the perineum and moves in the caudal direction to the subcutaneous tissue. By this time, all the muscle structures around it (PRM, Levators Ani, as well as the deep and superficial parts of the external anal sphincter) were formed normally. Not meeting the ectodermal part of the anal canal on its way, the internal sphincter turns forward and continues to create a canal until it penetrates some cavity. The penetration site is usually a narrow and rigid ring. In all cases, the anal canal is located outside the subcutaneous portion of the external sphincter. The caudal wall of the anal canal during its opening, regardless of the location of ectopy, is located opposite the anal dimple. The distance between the intestinal wall and the skin varies from 2 to 5 mm depending on the thickness of the subcutaneous tissue.

VI. **Diagnostics of ARM.**

1. **Diagnostic method in visible fistula.**

The purpose of diagnosis, first, to estimate the width of the rectum. Secondly, to determine the condition of the pelvic floor muscles. Third, to determine the distance between the wall of the anal canal and the anal dimple. Hydrostatic barium enema [37] and comparison of the obtained data with the standards (see Table 1) allow us to answer the first two questions (**Figure 5, A-B**).

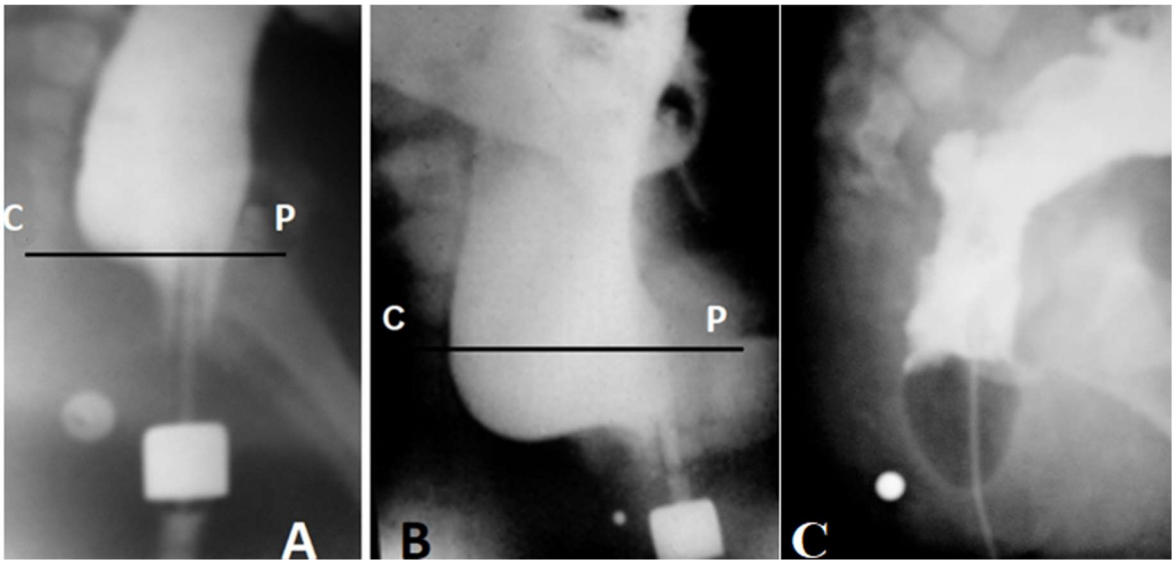

Figure 5. Lateral radiographs of the anorectum made in the same girl with vestibular fistula at a different age. P-C is the pubococcygeal line. The true diameter of the contrast marker, strung on the tip of the enema is 1.6 cm. It is located near the fistula orifice. **(A).** At the age of 8 months. Permanent contraction of the ectopic anal canal was observed during the barium enema. Its length is 2.5 cm, which corresponds to the anal canal 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 is not pulls of the posterior wall forward. **(B).** At the age of 1.5 years she had a 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 (minimal limit is 2.3 cm). Conclusion. ARM with vestibular ectopy, megarectum and descending perineum syndrome. (**C**). After emptying the gut, the Foley catheter was inserted into the rectum. After inflating the balloon, the catheter is retracted down

until it stops. It lingered over the narrow and rigid ring of the ectopic anus. The distance from the wall of the anal canal to the marker near the anal dimple is 2 mm.

In newborns with ARM, the rectum is of normal size. It expands as a result of chronic constipation caused by a stool retention due to stenosis of an ectopic anus (secondary megacolon or organic chronic constipation). In severe cases, wide fecal masses that cannot pass through the anal canal, under the influence of a strong peristaltic wave of the rectum, stretch the muscles of the pelvic floor. As a result of the weakness of the PRM and the levator plates, the upper part of the anal canal does not function to hold the feces, and becomes, as it were, a part of the rectum (descending perineum syndrome) [1]. In such cases, even an ideal operation cannot bring success, since chronic constipation and fecal incontinence or encopresis will result from weakness of the pelvic floor muscles and megarectum. Therefore, correction of the ARM with visible fistulas, should be performed before the appearance of the hard feces. If the child's condition forces to do a delayed operation, it is necessary to cut the ectopic anus in order to create a normal bowel movement.

2). **Diagnostic method in ARM without visible fistula in newborns.**

The study should be carried out in the interval of 24 -30 hours, when gas and meconium accumulate in the rectum in a volume that allows you to cause a defecation reflex during the abdominal compression. After 30 hours, theoretically there is a danger of bowel perforation and / or vomiting with aspiration hazard, despite the catheterization of the stomach. The study is carried out in a horizontal position of the child on the side under fluoroscopy. If during the abdominal compression between the palms of the radiologist gas penetrates the tissue of the perineum and approaches the marker near the anal dimple, then the patient has a functioning anal canal (**Figure 6**).

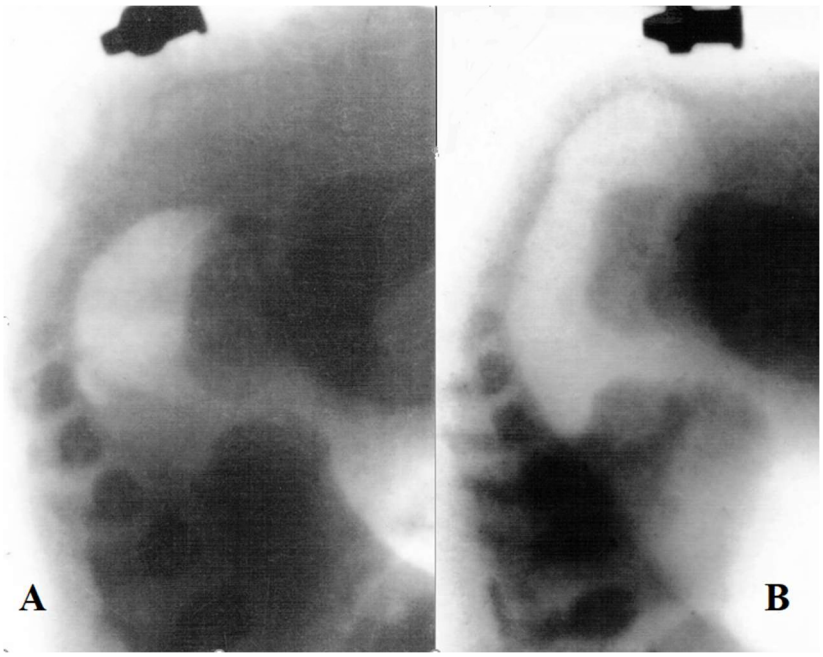

Figure 6. Examination of a patient with ARM without visible fistula. (A) Before compression of the abdomen. (B) During the compression of the abdomen, the anal canal opened, and its caudal wall approached the marker near the anal dimple. The marker is located on the axis of the anal canal 2 mm from the distal wall of the anal canal.

If the gas is at the level of the P-C line, but the true width of the rectum is less than 1.0 cm, there are 2 possible options: either there is a gas passed through an invisible fistula or volume the gas and meconium are too small to cause a defecation reflex. In any case, the study should be repeated after 4-6 hours. The narrow rectum indicates the absence of perforation danger. If the gut after an additional 6 hours has not expanded, this indicates the presence of a fistula. If the width of the intestine is more than 1.3 cm and the anal canal does not open, it means that there is a high type of ARM.

3). **Diagnostic method in ARM without visible fistula in patients with colostomy.**

Balloon-tipped catheter is inserted into the stoma far enough so that the balloon is into the proximal lumen of the ostomy of the mucous fistula. The balloon is then carefully inflated with 1–2 mL of air, just enough to occlude the stoma but not over-distend the bowel lumen. A 60-mL catheter-tipped syringe, without a

piston, is filled with 50 ml isosmotic watersoluble contrast agent. It is connected to the catheter and rises above the baby so that the tip of the syringe was 40 cm from the anus. Then need to open the faucet and wait until the level of the contrast agent in the syringe stops falling. This means that the rectum is filled with a contrast agent under low hydrostatic pressure, at which bowel perforation is impossible. Then under the control of fluoroscopy the abdominal compression is produced. In the presence of a low type of ARM, the contrast agent will approach the marker in the anal dimple.

VI. **Treatment of anorectal malformations**

1. **Analysis of posterior sagittal anorectoplasty.**

In the case of high-type ARM, where there is no anal canal, there is no other option except as pull-through operation. The choice should be made between methods that attempt to preserve PRM (anterior sagittal approach or laparoscopic pull-through operation). In cases where the presence of the anal canal is proven, the need to preserve it does not raise doubts. Saving IAS is accompanied by improved long-term functional outcomes [27,36,38,39,40,41,42]. The results of treatment after operations preserving IAS are significantly better than after the PSARP. But only children's surgeons from Finland continue to publish their results at the present time. Long-term functional outcomes by bowel function score in males treated for low ARMs (perineal fistula) were good in 85%, satisfactory in 15% and poor in 0%. Constipation was in 33% patients [39]. The cutback anoplasty is the most physiological operation, because all elements of the anal canal are preserved, except for the subcutaneous portion of EAS, which performs an emergency enhancement of stool retention during rectal pressure rise (during coughing, lifting from the spot, etc.) The results of this operation could be close to normal if it were performed before the development of irreversible changes in the rectum (megarectum) and in the pelvic floor muscles (descending

perineum syndrome) because of the stenosis of the ectopic anus. Based on the foregoing, it is important to emphasize, firstly, that for the optimal choice of the method of surgery it is necessary to establish the type of ARM (low or high). Secondly, the concept of "an intermediate level» makes no sense, i.e. either there is an anal canal, or it is not. Third, the ARM classifications, where all the urethral, vestibular and vaginal fistulas are considered as high types are erroneous.

Except for cut-back anoplasty, all other methods of surgery for the ARM to some extent damage the anal canal. Most pediatric surgeons perform the operation according to the original PSARP and the latest recommendations, with resection of the "fistula". In fact, they remove IAS, and in its place, they lowered the devascularized and denervated rectum. (**Figure 7**). In accordance with Peña's explanation, this procedure is unavoidable: "Putting traction on the rectum makes it possible to identify bands and vessels of its wall. Since these impede its being pulled through, they must be cauterized and cut" [30]. Obvious that the space available for accommodating an ectatic rectum is extremely small. Therefore, the author recommends tailoring the rectum, as a result of which the circular rectal muscle fibers are crossed (**Figure 7.b**).

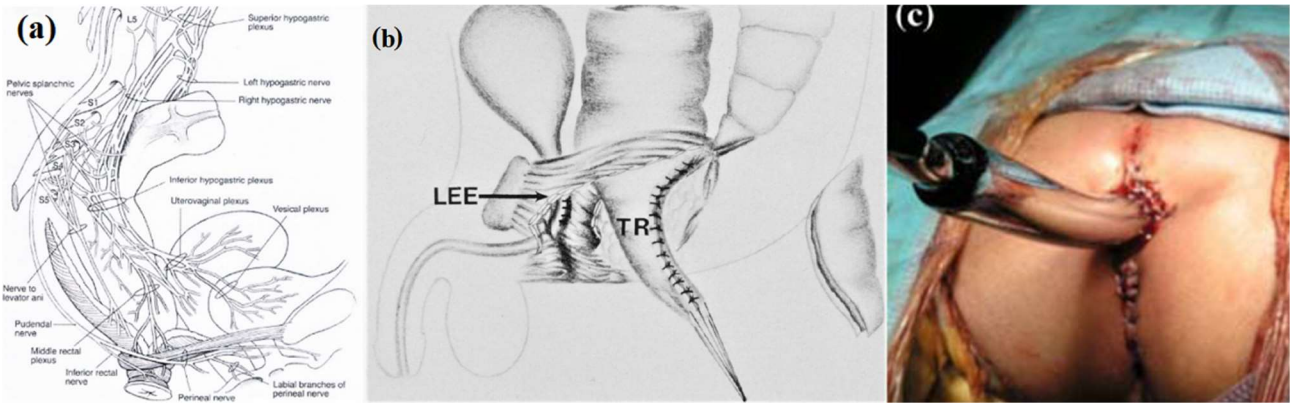

Figure 7. Stages of the pull-through procedure. **(a)** Scheme of the nervous supply anorectum from the article Bharucha [5]. **(b)** Tapering of the rectum [30]. **(c)** The final stage of the operation.

Figure 7 demonstrates that the segments of the intestine, both outside and in the tissues of the perineum (neoanus), is not only deprived of blood supply, but completely denervated. The authors' assumptions that the poor results of the operation are allegedly related to the pathology of the spine are unfounded.

Therefore, it is not surprising that, during an ultrasound study of anorectum after PSARP in 11/17 of the examined females the rectum did not reach the skin and a subsequent pocket under the skin and superficial EAS was found. Fragmentation of neo-IAS was noted in most of the females. In 11 females diastases in both the deep and superficial component of the EAS were identified. Most of the patients had a gap of 5 mm from the rectum to the skin anteriorly where also a lack of tissue under the superficial EAS and the skin was found [43]. The stitches on the background of devascularization and denervation were the cause of wound dehiscence in 17 (43%) without colostomy [44], which precedes the fibrosis of the tissue and the formation of stenosis.

In all pull through procedures, the levator plates are disconnected from the rectum. Therefore, after the operation, they do not participate in the opening of the anal canal, which leads to great resistance during an attempt to defecate and to the chronic constipation.

In the description of the operation it is indicated that "... a median sagittal incision that runs from the sacrum to the anal dimple, cutting through all muscle structures behind the rectum" [30]. In the discussion of this article, all known surgeons objected to the intersection of puborectalis muscle. So, for example, D. Johnson (Salt Lake City, Utah) put it: "Many of us were dismayed at first showing by Dr. Peña's suggestion that we cut through the entire sphincteric mechanism. One can legitimately question whether the obvious benefits from the vastly improved surgical exposure are counterbalanced by functional loss from incisional damage."[30]. Here are the explanations of Dr. Peña: "First, the anatomy of the sphincteric mechanism differs considerably from that described based on cadaver

dissections. Second, the disadvantage of cutting the sphincter is more than offset by increased precision in sphincter identification, bowel position and sphincter reconstruction. Third, rectal segment tapering of the blind ectatic pouch is essential to prevent damage by overstretching of the sphincteric mechanism". Dr. Peña's answers could not convince the well-known pediatric surgeons that he was right, because its answers were outside the scope of scientific discussion.

Concerning the 1st answer. "Drs. Peña and deVries have been given anatomical dissections in live patients". Dr. Peña's discovery was that "... we've been unable to identify it (PRM)," and adds: "... that probably nobody has actually seen it by means of the conventional incisions." However, six pediatric surgeons participated in this discussion, and each of them saw and saved PRM during surgery. Attempting to stitch the edges of the PRM at the last stage of the operation is not real, since it is impossible to stitch what is not visible.

Concerning the 2nd answer. Dr. Peña does not deny damage to PRM but claims that "cutting the sphincter is more than offset by increased precision in sphincter identification...". What sphincter is he talking about? The internal anal sphincter and puborectalis muscle are not identified. In general, what can compensate for damage to PRM, which plays an important role in fecal retention?

Conclusion: In most cases, the ARM has a functioning anal canal with an ectopic outlet in the perineum, vestibule, urethra or vagina, with a diastasis between the anal canal and the anal fossa from 2 to 5 mm. Pediatric surgeons perform: excision of the internal anal sphincter, intersection of the puborectalis muscle, damage to the external anal sphincter, disconnecting of the levator plates from the rectum, devascularization and denervation of the rectum, and the destruction of the reflex connections between the intestine and sphincters. In fact, instead of a functioning anal canal, a long artificial fistula is created in the perineum, in which, depending on the degree of stenosis, either constipation or fecal incontinence prevails.

2. **The purpose of the surgery** is to preserve all elements of the anal canal in low types of ARM. For this purpose, must create a channel between the distal wall of the open anal canal and the skin in the anal dimple. This channel in the skin and subcutaneous tissue has a length of 2 to 5 mm, depending on the age of the patient. In patients without visible fistula, this procedure is enough to create a functioning anal canal. In patients with visible fistulas, it is necessary to close the ectopic anus, after reaching the normal size of the newly created anus.

Justification why it is not necessary to search for the place of the fistula and cut it off in cases of urethral fistula. First, the diameter of the urethra in a newborn is 2 mm. The fistula occupies only a small portion of its posterior wall. Sometimes the fistula is not functioning because of the strong narrowing or it obliterated and presented in the form of cord [45]. (I have never seen in newborns the disappearance of gas from the rectum during compression of the abdomen). Secondly, the fistula is in the distal part of the anal canal. Since the anal canal is permanently contracted, the fistula is closed by a tonic contraction of all sphincters of the anal canal. During defecation, a reflex contraction of the urethra occurs, which prevents the transfer of feces into the urethra [46]. Third, if a functioning fistula is found, it can be closed through the anus (see figure 4).

Justification why should not the internal anal to sphincter separated from the surrounding tissue and to sew it with the skin. The internal anal sphincter has a high proteolytic and creative activity. In the early embryonic period, it migrates to the cranio-caudal direction, creating a channel up to the subcutaneous tissue. This creative activity is preserved after birth, which explains the difficulty of treating of the anal fistulas. The diastasis between the wall of the anal canal and the skin in a newborn with ARM is 2 mm. Within 7-10 days, healing takes place without scarring and stenosis [47].

3. **Perineal perforation procedure (PPP) for newborns without visible fistula.**

On the X-Ray table under general anesthesia the cross-section of skin is done above contraction of the subcutaneous portion of external anal sphincter (EAS). After stretching of its fibers, the child is transferred to a lateral position. During fluoroscopy, when the anal canal is opened during abdominal compression, the needle is inserted from an incision in the skin into the rectum through the open anal canal (Figure 8, A-B). Only that step is performed under fluoroscopic control. Before proceeding it is necessary to make sure that the needle is in the rectal lumen. This is evidenced by the sound of the outgoing gas or the appearance of meconium. If there is no certainty that the needle is in the rectum, appropriate to introduce a contrast agent into the intestine through the needle. Then, a conductor with soft floating end is inserted through the needle into the rectum (Figure 8, C). The needle is removed and the tight conical bougie with a maximum diameter of 0.8 cm is introduced into the gut along the conductor (Figure 8, D). After that, the tracheostomy tube with diameter 0.8 cm introduced into the rectum and the conductor is removed (Figure 8, E). Four cm^3 of air is introduced into the balloon of this tube that allows to fix the tube for 7 days. After 7 days, the tube is removed.

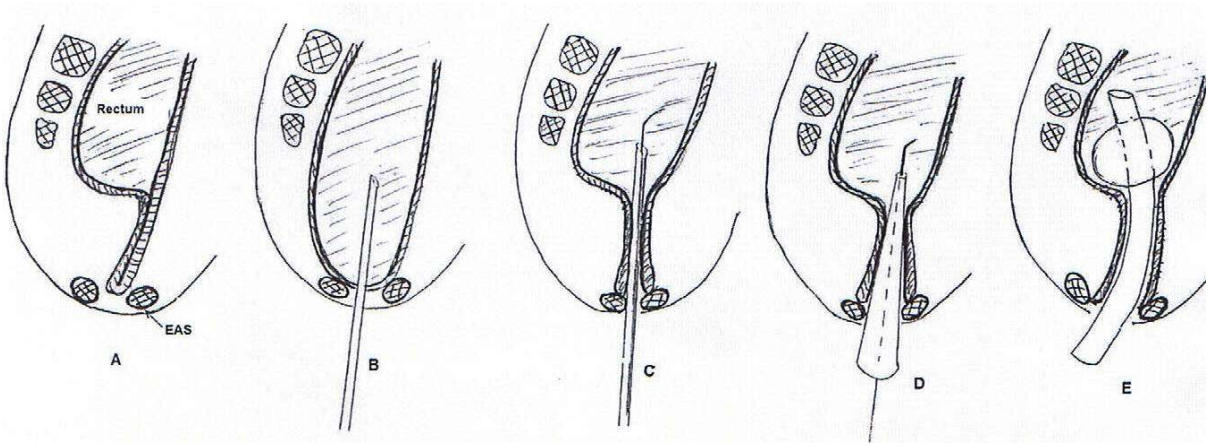

Figure 8. Scheme of the "perforation perineum procedure" (PPP) to treat low imperforate anus without visible fistula. Lateral view.

A. The anal canal is closed. **B.** Anal canal is opened due to abdominal compression. Under X-ray control a needle is introduced into the anal canal. **C.** Conductor for

vascular catheterization is introduced into the rectum through the needle. **D.** The tapered dilator introduces on a conductor for expansion of the newly created channel. **E.** After removal of the dilator, the tracheostomy tube introduced along the conductor. Its balloon is inflated in the rectum.

The tube is removed through a week after the procedure. During this time, the emptying of the rectum there is through the tracheostomy tube. The internal anal sphincter is not separating and is not sewn to the skin.

3, A). **PPP for patients without visible fistula with colostomy.**

The technique of surgery can be the same as described above. The opening of the anal canal is created by the hydrostatic pressure of the contrast medium (see diagnostics) or after the introduction of air.

4. **The proposed methods for the treatment of patients with visible fistulas.**

Considering that all patients with visible fistulas have a functioning anal canal, the blind end of which is located near the anal dimple, I suggest perforation of the perineum with subsequent suturing of the ectopic anus using a colostomy[34]. The operation, including colostomy, should be done at the age of over a year. Firstly, because in pediatric surgery, there is an axiom that the older the baby, the better the wound healing and less inflammatory complications are observed. In addition, the accuracy of determining the anatomical structures increases and, consequently, the efficiency of reconstruction of the defect increases. Secondly, in newborns, the anal dimple is located close to the ectopic anus. Therefore, there is a danger during the bougienage of the newly created anus to break the jumper between the holes. To prevent the development of irreversible changes in the rectum (megarectum) and in the pelvic floor muscles (descending perineum syndrome), it is necessary to cut the stenotic ring of the ectopic anus, and thus avoid chronic constipation.

Recently, colorectal surgeons began to use PSARP in newborns. For example, Liu et al state: "The 1-stage PSARP procedure in the neonate not only achieves the same long-term outcome as the conventional PSARP procedure but also involves fewer short-term complications" [48]. However, randomized controlled trial showed that "Females with VF (vestibular fistula) or PF (perineal fistula) undergoing primary definitive procedure have a significantly higher incidence of wound dehiscence (P = 0.04), immediate (P = 0.001) and early postoperative complications (P = 0.01) [49]. Lauriti et al also found that, in females with rectovestibular fistulas, the PSARP performed without protective colostomy increases the risk of postoperative complications. And, this one-stage approach not associated with reduced fecal continence [50]. In none of the numerous articles, I found a discussion of the reasons why functional results after single-stage repair in neonates are the same as after 3-stage treatment. The comparison groups were similar in all parameters. The only explanation is the fact that nothing can be worse than the destruction of the anal canal.

I suggest 2 methods of perforation of the perineum. One using a Foley catheter (Figure 9) and the other is compression anastomosis using a simple mechanical device (Figure 10). It has been proven that compression anastomotic healing is associated with less foreign body reaction, scarring, and inflammation as compared with stapled anastomosis [51].

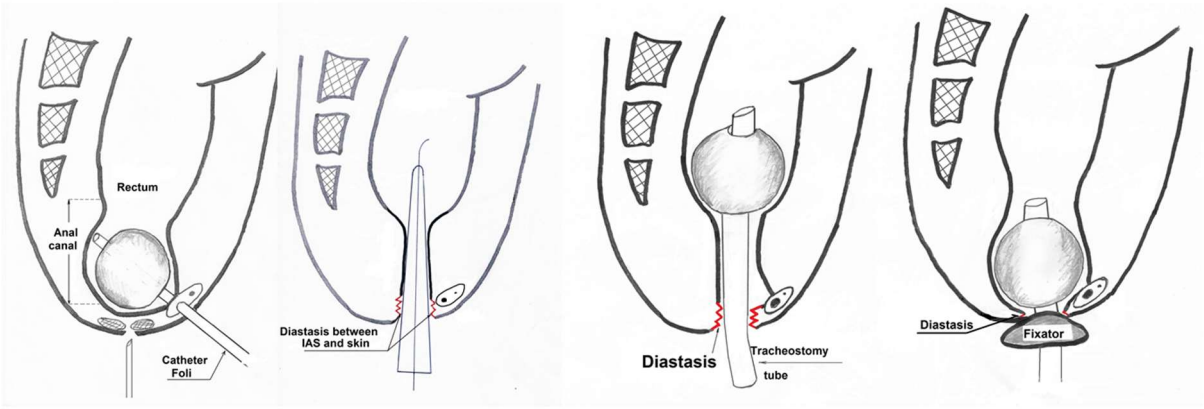

Figure 9. Perineal perforation scheme in patients with visible fistula using Foley catheter. (**A).** A Foley catheter is introduced into the rectum through the ectopic anus. From 5 to 10 cm^3 of air is introduced into its cuff. After that, the catheter is pulled up to a stop. The inflated balloon is held in place over the rigid and narrow fistula. At this point, the wall of the internal anal sphincter is approaching the anal dimple (see Figure 5, C). The needle is inserted into the rectum from the skin incision inside the ring of the EAS. It pierces the balloon what allows the catheter to be removed from the rectum. **(B).** A guide is introduced into the rectum through this needle. Along this guide a conical dilator with a maximum diameter of 8 mm to 12 mm (depending on age) is screwed into the rectum. **(C).** After removing this dilator, a tracheostomy tube is introduced into the rectum through the guide, and 10-15 ml of air is injected in its cuff. At this point diastasis between the perforated wall of the anal canal and perineal skin is from 2 to 5 mm. **(D).** The inflated cuff is descended until it stops and is then fixed in place. Thus, the perforated wall of the anal canal approaches the skin. Diastasis dramatically decreases, which promotes rapid healing without significant scarring.

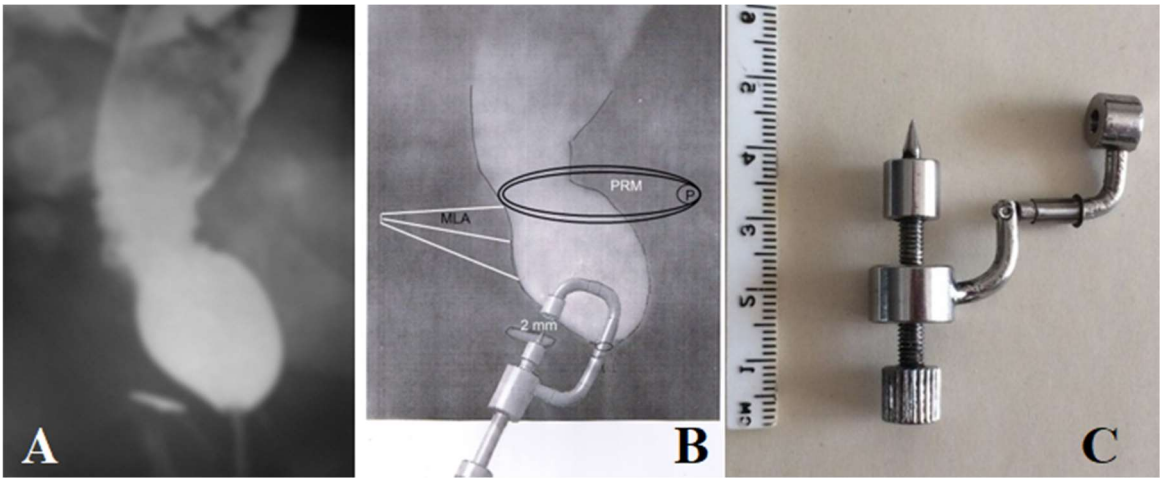

Figure 10. Perineal perforation scheme in patients with visible fistula using compression anastomosis. **(A).** Lateral radiograph of a female with a vestibular fistula. After a barium enema, a Foley catheter is inserted into the rectum. A liquid

contrast agent is inserted into its balloon, and the balloon is pulled up to the stop. The distance from the balloon to the button near the anal fossa is 2 mm. **(B).** The upper cylinder of the device with a diameter of 1 cm is inserted into the anal canal through the ectopic anus. The lower cylinder of the same diameter is inserted into the wound inside the fibers of the subcutaneous part of the external anal sphincter. During screwing up, the cylinders meet and gradually squash soft tissue. (C). Photo of the device for compression anastomosis.

In those newborns in whom I did the perforation perineum procedure (PPP) without stitching the internal anal sphincter with the skin, healing took place without complications and without the formation of scars. Their anorectum function did not differ from the norm [34]. Therefore, I am sure that PPP, in children with low type of ARM, can ensure a normal future for these patients.

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