How to preserve the anal canal in patients with anorectal malformations

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Anorectal malformations (ARMs) include all forms of congenital anomalies of the anus and anal canal. Imperforate anus represents most ARMs, except for rare forms such as true cloaca, anal canal atresia (stenosis) and H-shaped fistula.

Part 1. Pathological anatomy and physiology of imperforate anus

1. Embryology is the key to understanding the pathological anatomy and physiology of imperforate anus

During the postcloacal period of embryonic development, a normal anal canal formed from two rudiments. The endodermal internal anal sphincter (IAS) migrates to meet with the exodermal primordium. They meet between the proximal two-thirds and distal one-third of the anal canal. Between them, the embryonic anal membrane forms, which is destroyed when the embryo is 13.5-135mm long. The dentate line forms at the site of the destroyed membrane. Thus, only a very rare true cloaca arises because of developmental arrest at the stage characteristic of birds and reptiles. In these cases, the rectum, uterus, and urinary bladder open into a cavity called the cloaca. The anal canal, vagina, and urethra are absent. The remaining ARMs arise during the postcloacal period. If the embryonic anal membrane does not degrade, membranous atresia of the anal canal results. If it does not degrade completely, anal stenosis occurs. Understanding that atresia and stenosis occur in the anal canal, and not in the rectum as previously believed, allows for excision of the membrane through the anal approach without disrupting the anal canal [1,2].

With an imperforate anus, the anal opening is absent, and fibrous tissue is sometimes covered (covered anus). The subcutaneous portion of the external anal sphincter (EAS) may be normal or hypoplastic. Clearly, all types of imperforate anus result from disruption of the development of the exodermal primordium of the EAS. The endodermal EAS, without encountering the exodermal primordium, reaches the subcutaneous tissue opposite the anal fossa. Further advancement of the EAS always occurs until it penetrates the outside or into a cavity. Except in cases of anal stenosis, the EAS is displaced anteriorly and superiorly strictly in the sagittal plane. However, outside the normal path, it creates a narrow, rigid channel. Thus, a narrow, rigid opening in the perineum, as well as in the urethra in boys and the vagina in girls, is ectopia

ani. Secondly, by the time the anus penetrates outward or into any cavity, in all cases there is already a normally formed anal canal, except for a few millimeters, depending on the thickness of the skin and subcutaneous tissue.

2. ARMs with visible ectopic anus and stenosis of the anus.

When the IAS penetrates the subcutaneous fat layer and the imperforate anus, the stenosis of the anus occurs (Figure 1a). Because the term "anal" refers to both the anus and the anal canal, these terms are often confused in the literature. To avoid confusion, congenital anal stenosis should be referred to as stenosis of the anus. A normal anal canal remains above the subcutaneous tissue, and a narrow fistula tract is caudally. Its length is equal to the thickness of the subcutaneous fat in which the subcutaneous portion of the EAS is located, and the thickness of the skin. In newborns, this thickness is 2 mm, and in infants under one year old, it is 4 mm **(Figure 1).**

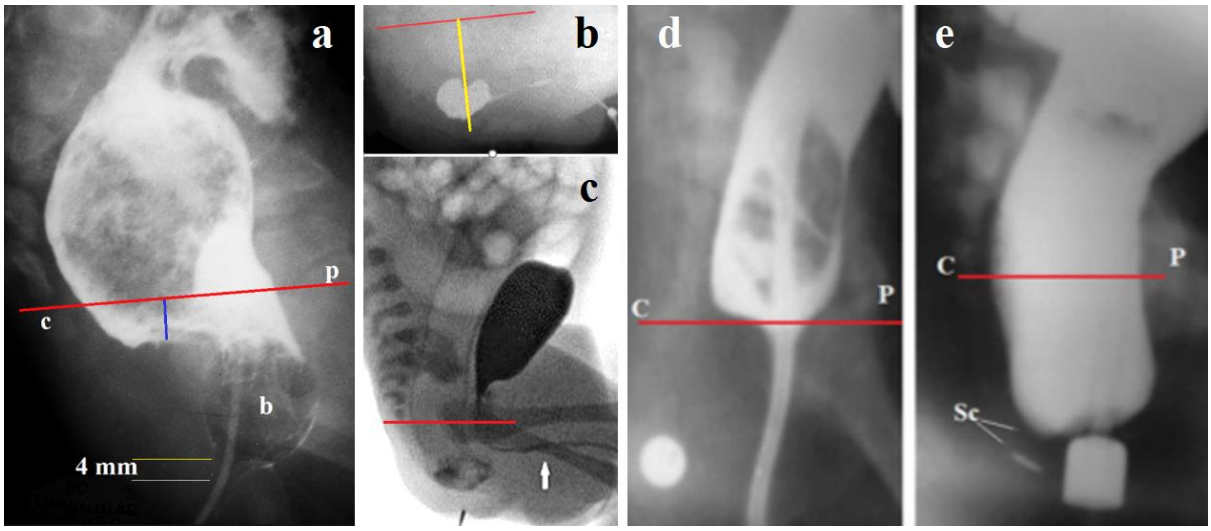

Figure 1. Radiographic studies of ARMs with visible fistulas (low-type).

(a). A patient under 1 year old with chronic constipation and soiling. A Foley catheter is inserted through a normally located but narrow anus. After injecting barium into the rectum and introducing air into the balloon (b), the catheter was pulled up to the stop. It is stuck above the stenosis, 4 mm from the anal dimple. A large fecal stone is located in the rectum. A strong peristaltic wave attempts to push it out, but this only leads to stretching of the pelvic floor muscles (the puborectalis muscle (PRM) and levator plates). The anal canal, which should normally be located between the pubococcygeal line (red) and the anus, is shortened in this patient by the length of the blue line – descending perineum syndrome. Distension (weakness)

of the PRM manifests as fecal incontinence. As a result of late diagnosis of congenital stenosis of the anus, megacolon and the descending perineal syndrome developed.

(b-c). (b) Yang et al. described a rectopenile fistula [3]. Contrast medium, injected into the hole at the root of the scrotum, revealed a long and narrow ano-penoscrotal fistula. (c) The following day, when the gas volume reached the threshold volume, the anal canal opened (below the pubococcygeal line - red). Gas from the rectum approached the anal dimple. The contrast agent is in the anal canal, not the rectum.

(d-e). (d) Lateral radiograph of the anorectal area of a 9-month-old-female patient with vestibular fistula. The barium was injected into the rectum through a catheter passed from the ectopic orifice. A round thumbtack was glued to the anal dimple. The sphincters of the anal canal located caudal to the pubococcygeal line (red) have contracted around the catheter, preventing barium leakage. (e) The same patient at 1.5 years of age. Radiograph taken during an attempted defecation. A wide opening of the anal canal occurred. The distance from the wall of the IAS to the thumbtack in the anal dimple (Sc) is 4 mm (the true diameter of the marker near the ectopic anus is 1.6 cm). The subcutaneous part of the EAS is in the subcutaneous fat tissue in the space between the skin and the IAS. As can be seen in Figure 1 (c), its absence around the ectopic anus does not affect the function of barium (feces) retention.

These data are consistent with the Wingspread classification, in which ARMs with visible fistula were classified as low types and named anal stenosis, anocutaneous fistula, and anovestibular fistula. Stephens' research established that low types of ARMs are characterized by a normally functioning anal canal, and a cutback procedure was widely performed to preserve it [4]. After this procedure, fecal continence remained normal, and constipation, which was caused by late correction, decreased over time [5].

The name of the pathology is of great importance, as it must convey the pathoanatomical and physiological characteristics of the defect. For example, Caffey's textbook (1978) stated that pediatric surgeons came to the conclusion that an imperforate anus is an ectopia of the terminal part of the intestinal tube, but they, out of old habit, call it a fistula [6]. The Krickenbeck classification, which involved 26 pediatric surgeons, 22 of whom had no scientific publications, stated: "Therefore, the Pena classification does not distinguish between rectovestibular and anovestibular fistulas. When performing a PSARP procedure, however, this differentiation did not seem important." [7] As a result of this confusion, pediatric surgeons destroy the normally

functioning anal canal, calling the IAS a fistula or a rectal pouch. Peña believed the role of the PRM was exaggerated because he failed to identify it during PSARP [8]. There, he claimed to have been the first to discover the significant role of the external sphincter, having in mind the subcutaneous portion, the only part of the EAS that is not transected during PSARP. Because of their belief in Peña's "experience," pediatric surgeons are ignorant of the normal anatomy of the anorectum. For example, they refer to the mucocutaneous junction in the anal area as the dentate line. **Figure 2** shows a diagram of the anorectum's anatomy. The subcutaneous portion of the EAS is represented by muscle bundles, the total volume of which accounts for 1/10th of the EAS.

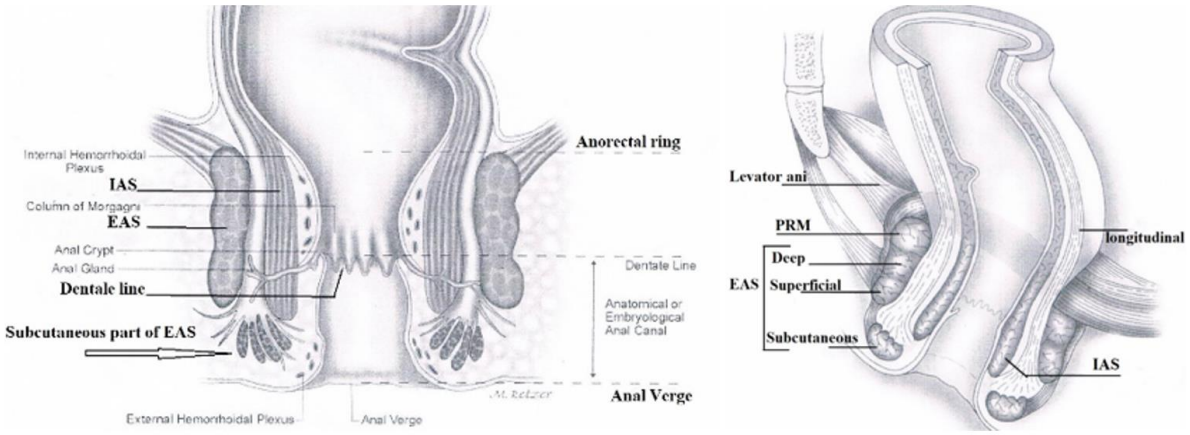

Figure 2. Diagram of the anorectum from the ASCRS Textbook of Colon and Rectal Surgery [9]. The subcutaneous portion of the EAS is the 10th part of the EAS, which does not play a role in fecal continence. Therefore, complete transection of this portion of the EAS during a cutback procedure does not result in fecal incontinence [10].

3. ARMs without visible ectopic anus

The Arm-Net Consortium made an important statement: “According to current knowledge, the ‘fistula’ in ARM represents an ectopic anal canal and should be preserved as far as possible to improve the chance of fecal continence” [11]. This means that if, with an imperforate anus, the fistula has not penetrated the outside, it continues to migrate forward and upward until it penetrates the urethra or vagina. In any case, by the time an ectopic anus develops, a normally formed anal canal already exists. The ectopic opening can be located at any distance from the anal dimple, but the wall of the anal canal is always at the same distance from the anal dimple – from 2 mm in newborns to 4 mm in infants. From this perspective, all cases of imperforate anus are low-type.

The idea that high types of ARMs are possible was based on a lack of knowledge about the physiology of the anal canal. The anal canal is normally contracted and opens only when rectal pressure increases to a certain level. Therefore, a contracted anal canal was mistaken for a fistula tract during radiographic examination and surgery. The absence of the intermuscular nerve plexus is characteristic of normal IAS. This has been confirmed by histological studies of IAS in children without ARM. Secondly, all pediatric surgeons know that to diagnose Hirschsprung disease, samples must be taken above the anal canal for histological examination. Therefore, the absence of nerve ganglia is normal and does not indicate a fistula or rectal pouch. Meanwhile, histological studies in patients and piglets with ARM indicate the presence of a normal anal canal. Manometric examination revealed a positive rectoanal inhibitory reflex. Radiographic studies showed wide opening of the anal canal during increasing rectal pressure in all ARMs, including those without visible fistulas, which were considered high types **(Figure 3a)** [10,12].

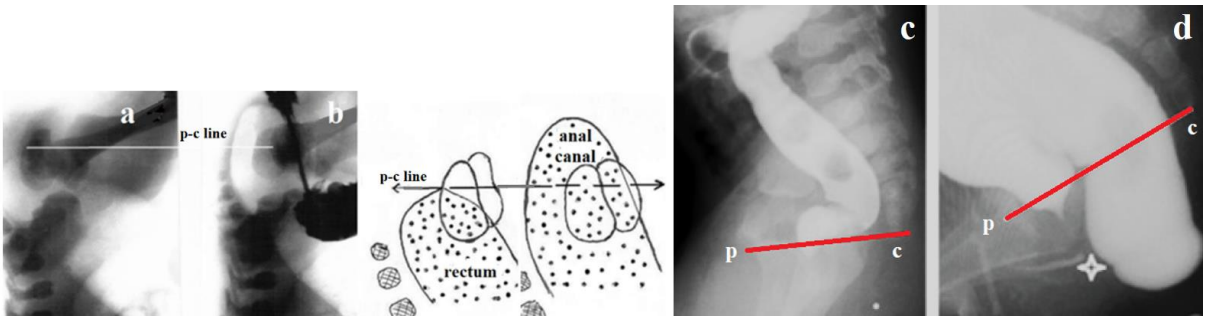

Figure 3. Radiographic studies of patients with urethral ectopy of the anus. **(a).** In a newborn without a visible fistula, the distal intestinal contour, filled with gas, is located at the level of the pubococcygeal line, i.e., on the border between the anal canal and the rectum. **(b).** During an unsuccessful attempt to inject contrast agent into the rectum, the anal canal opened and gas approached the perineal skin. (c-d). Radiographic studies of boys with "rectourethral fistula" performed using high rectal pressure from the article by Kraus et al [13]. **(c).** The authors state that the rectal sphincter is closed and not distended because of inadequate pressure. **(d).** With adequate pressure, the contrast agent filled the wide rectum, which contacts the urethra [13]. The distal edge of the intestine is located far from the P-C line (drawn by me), near the perineal skin.

The portion of the intestine that is caudal to the pubococcygeal line (Figure 3d) is called the anal canal, not the rectum. The article by Kraus et al states: "It is extremely important in this

regard to understand that the lowest part of the rectum is usually collapsed due to the muscle tone of the funnel-like striated muscle mechanism that surrounds the rectum in 90% of cases (in 10% of cases, mostly bladder-neck fistulae, the fistula is above the sphincter muscles)" [13]. The authors demonstrate on radiographs (Figure 3c, d) and describe the function of the normal anal canal, which is constantly contracted because of sphincters contraction and opens wide under high rectal pressure because of contraction of the levator plates. They call it the rectum, although it is known that there are no muscles around the rectum. Nevertheless, these data provide convincing confirmation that the anal canal is present in the so-called high types of ARM. Secondly, as can be seen in Figure 3d, all urethral fistulas are located below the pubococcygeal line (red), i.e. in the anal canal.

4. ARMs with vaginal ectopy

While the urethra is in the path of ectopic anus in boys, in girls the ectopic anus extends into the vagina. The scientifically unsubstantiated change in the name of ectopic anus in vagina to persistent cloaca was a disaster for these patients. The diagnosis of cloaca implied a dysfunction of the urinary system. Without research or evidence in the literature, Peña proposed operations as if these were children with a true cloaca and a normal urethra was being transformed into a bloodless and denervated tunnel, rendering the patients disabled. PSARP simultaneously destroyed the anal canal [14]. The pathological anatomy of this defect can be explained by an embryological feature that leads to two types of ectopic anus in the vagina. The usual form occurs when the anus is inserted into the vagina after a cavity has already formed in it. As with all forms of imperforate anus, they have a normally functioning anal canal. If a cavity has not yet formed when the ectopic anus inserts into the vagina, then a narrow, rigid fistula develops from the insertion site to the vaginal opening, like that seen in boys with an anopenial fistula (see Figure 1b-c). **(Figure 4).**

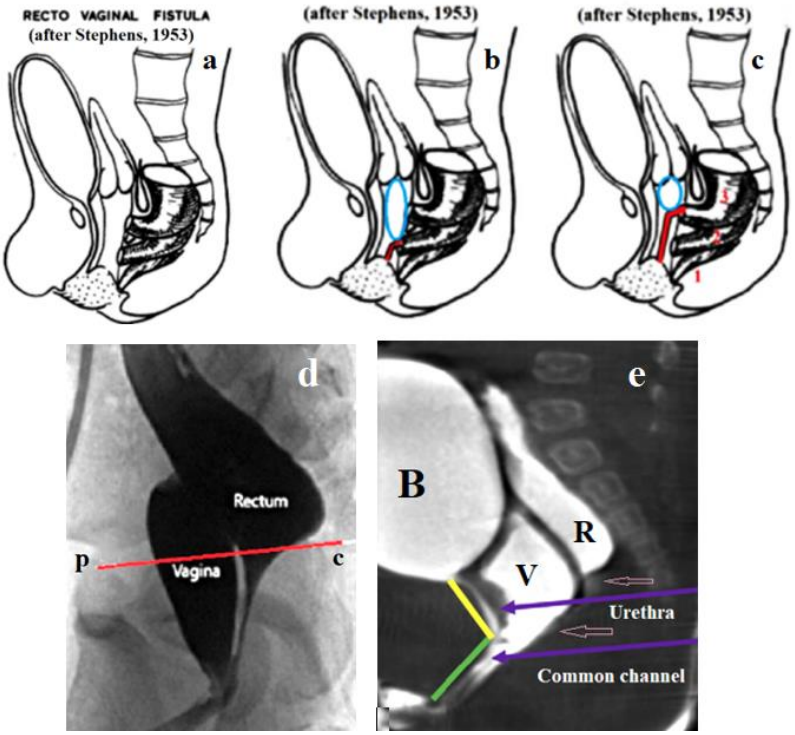

Figure 4. Diagrams and radiologic images of different types of ARMs with vaginal ectopy of the anus. **(a).** I used Stephens' diagram as a template. It shows ectopy ani in the vagina with at three different levels (1-vestibular; 2-vaginal low; 3-vaginal high). **(b)**. If a cavity has not yet formed in the vagina at the time of ectopy, in the lower part of the vagina forms a short, narrow canal that reaches the vagina. Sometimes it blocks the outflow of fluid from the proximal part, resulting in the formation of a closed cavity called hydrocolpos (blue ellipse). (c). With high ectopia ani, it forms a long and narrow fistula tract in the vaginal tissues. **(d)**. Ectopy ani in the lower part of a normally developed vagina. An incompletely closed anal canal is visible below the pubococcygeal line. **(e).** MRI examination from the article by Wood et al [15] with the caption "Short common channel cloaca with adequate urethra." The authors proposed that in the corner between the yellow and green lines, the urethra joins the vagina to form a common canal. The section, however, clearly shows that the urethra, which approaches the narrow vagina, continues to the perineum. A dark dividing line can be seen between the urethra and the narrow vagina, which represents the union of the urethral and vaginal walls. The pink arrow below the rectum shows the beginning of the anal canal. The lower pink arrow shows the ectopy of the anus in the vagina.

Analysis of radiological studies confirms that (1) penetration of the anus into the vagina, as with other types of imperforate anus, always occurs when the anal canal is already formed. (2) In all cases of ectopia of the anus, a normally functioning urinary system is present in the

vagina. (3) Depending on whether a cavity has already formed in the vagina during penetration of the anus, two types of ectopia occur: (a). When the cavity has already formed, a typical form with a wide vagina occurs; (b). If there is no cavity in the vagina, a long and narrow canal is formed, which opens near the urethra. The current situation is analyzed in detail in my article [14]. Below I will briefly dwell on the main evidence.

Pediatric surgeons encountered an unusual type of imperforate anus, where feces were released from the vagina, suggesting an ectopic anus within the vagina. However, in place of the vagina, there was a narrow canal. Practitioners did not engage in scientific research. Therefore, to date, no studies have been conducted on the function of the urinary system, anal canal, or embryology. By calling the defect a cloaca, surgeons a priori assumed that urinary function was the same as in a true cloaca. However, the presence of a urethra, vagina, and anal canal indicates that the defect developed in the postcloacal period. The idea that the three already formed canals reunited in a single location is not only unsubstantiated but also contradicts embryological principles. As some surgeons have shown, in cases of persistent cloaca where the urinary system was not surgically altered, no functional impairment was found. In cases where the operations proposed by Peña were performed, they were worse than in other forms of imperforate anus. For example, the article by Lane et al. presents the long-term outcomes of twenty-one patients who underwent cloacal surgery and met the inclusion criteria. Two patients have required renal transplants for congenital renal dysplasia, and two have developed chronic renal failure associated with the sequalae of vesicoureteric reflux - (19%). Eleven of the patients manage their bowels with an antegrade continent enema (ACE), and two of the LCC cloaca are defunctioned with a colostomy (62%). Clean intermittent catheterization is performed by 12 (57%) of the patients, either per urethra or via a Mitrofanoff channel. The authors concluded: -" The urinary and fecal continence are the main challenges in the management of cloaca patients. Many require surgical intervention to achieve social continence" [16]. These disastrous results do not correspond to reality, since those who were lost to follow-up, including those who died, were excluded from the total number of operated patients.

Part 2. Pathogenetic treatment of different forms of imperforate anus

1. Ectopy of the anus with visible openings (perineal, vestibular)

As shown above, these patients have a normally functioning anal canal. The distance from the anal canal wall to the anal pit is 2 mm in newborns and 4 mm in infants. This distance is determined by the thickness of the skin and subcutaneous tissue. The subcutaneous tissue contains the subcutaneous portion of the EAS. The anteriorly displaced opening is usually narrowed relative to normal and is represented by a rigid fibrous ring. Depending on the diameter, constipation may occur soon after birth or when stool becomes hard. Timely diagnosis and treatment are essential, as delayed treatment leads to megacolon.

The cutback procedure is the ideal surgical procedure for perineal ectopy of the anus. In the original so-called "cutback operation," one blade of a scissors was placed in the fistula and the other across the perineum; upon closing the scissors, the lower orifice was enlarged by cutting backwards in the midline of the perineum. To prevent a blind pocket from forming posteriorly the newly created anus, the ring of the subcutaneous part of the EAS must be transected into two parts along its diameter. According to pediatric surgeons, fecal incontinence is never observed despite complete transaction of the subcutaneous part of the EAS [5].

Since "an ectopic anus is a true anus with normal nerve control for opening and closing," Browne recommends cutback surgery for vestibular ectopy in girls and perineal ectopy in both sexes. To prevent stenosis, he warns: "No attempt should be made to suture the raw surfaces thus produced, and after a month or two they will be covered with supple and satisfactory new skin" [17]. In such cases, dilation begins 2 weeks after surgery to prevent stenosis. My research has confirmed the validity of Browne's recommendation. In three newborns without visible fistulas, in whom I perforated the perineum and inserted a tracheostomy tube into the anal canal, the diastasis between the anal canal wall and the skin healed without developing stenosis [18].

There's an unwritten rule among surgeons to close the surgical wound, creating an anatomy as close to normal as possible. Almost all surgeons perform an anoplasty after a cutback procedure, which leads to a narrowing of the newly created anus and an inflammatory reaction that results in the formation of fibrous tissue. Dilation of the stenosis enlarges the opening but does not eliminate rigidity. Therefore, with age, repeated dilations or surgeries are required. Moreover, in some patients with vestibular ectopy after the cutback procedure, the anus is situated very close to the vagina, presenting a significant aesthetic defect, despite a positive functional outcome. This accounts for pediatric surgeons opting for anorectoplasty, despite the notable loss of anorectal function. Although the potential risk of urinary tract inflammation due

to the proximity of the urethral and anal openings is often cited as an additional reason for avoiding the cutback procedure, no evidence of this complication has been reported yet.

Based on a literature review and my own research, I proposed a new version of the cutback procedure for children with perineal and vestibular ectopy of the anus [19, 20]. Tissue dissection is performed as described by Browne, starting from the ectopic anus until the ring of the subcutaneous part of the EAS is completely transected. A tracheostomy tube is inserted into the rectum. Its balloon is inflated in the rectum to secure the tube in the anal canal. In case of vestibular ectopia, to move the newly created anus away from the fourchette, the skin and subcutaneous tissue are sutured from the tube to the site of ectopy. Bowel movements are performed through the tube for 2 weeks. During this time, the diastasis between the anal canal wall and the skin is replaced by new elastic tissue **(Figure 5).**

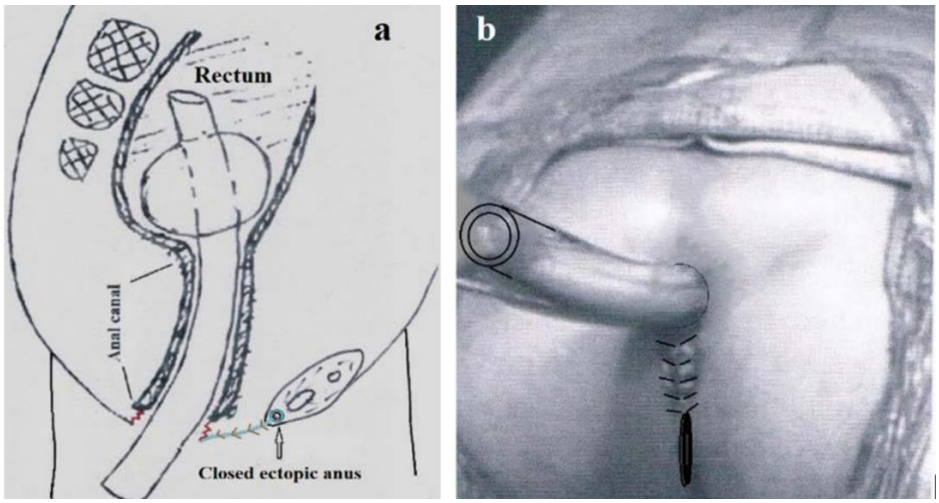

Figure 5. Schemes of modification of the cutback procedure in vestibular ectopy. **(a)**. Central sagittal plane. Initially, the skin, subcutaneous tissue, incised between the scissor blades from the location of the ectopic anus in the vestibule (arrow) to the complete intersection of the annulus of the subcutaneous part of the EAS. A tracheostomy tube is inserted into the rectum. At its end, a balloon is inflated to secure the tube in the anal canal. The skin with subcutaneous tissue is closed with single sutures from the ectopic anus to the anterior wall of the tube (blue line). A diastasis had formed between the wall of the anal canal and the skin (red). **(b).** Frontal view.

2. Congenital stenosis of the anus

Congenital anal stenosis differs from perineal and vestibular ectopy by the absence of displacement. The fibrous ring, fused to the subcutaneous portion of the EAS, is 2 to 4 mm thick. Above it lies a normally formed anal canal. Treatment, like the cutback procedure, involves dissection of the ring anteriorly and posteriorly, followed by insertion of a tracheostomy tube, as described above.

3. Ectopy of the anus without a visible opening (into urethra)

All cases are divided into two types, depending on the results of the X-ray examination. It is performed on the second day after birth, when swallowed air enters the rectum and its volume, along with meconium, allows the anal canal to open. The examination is performed with the patient in a horizontal position on their side. During fluoroscopy, the doctor compresses the newborn's abdomen between two palms. If the fistula is not functional, the pressure in the rectum will reach the pressure of the defecation reflex, leading to the anal canal opening **(Figure 6).** In such cases, the anal canal remains open while pressure is applied. This allows us to perform a "perineal perforation procedure" (PPP). If, despite abdominal compression, gas disappears from the anal canal, a colostomy is performed.

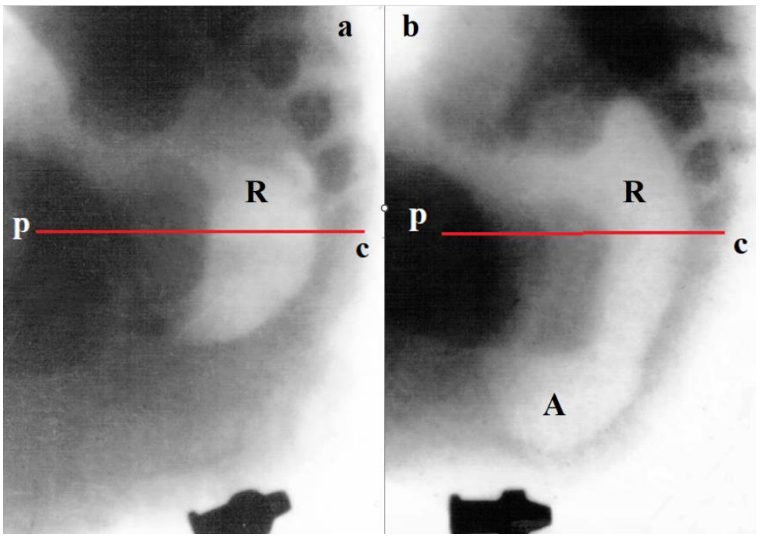

Figure 6. X-ray study of a newborn with abdominal compression. The cannula from needles is taped near the anal dimple. **(a).** At the beginning of compression, gas from the rectum (R) began to enter the anal canal. **(b).** And immediately the caudal wall of the anal canal (A) approached the marker near the anal dimple. The anal canal contracted and gas returned to the rectum as soon as abdominal compression stopped. This image indicates the absence of a functioning fistula and the possibility of performing a perineal perforation procedure (PPP).

The functioning anal canal forms from the rectum up to the subcutaneous tissue opposite the anal dimple, even before the onset of anal displacement, regardless of the height of the displacement. However, only urethral ectopy can result in obstruction of the anus due to a narrow opening being formed in the narrow urethra if it is sealed with a tight meconium plug. Therefore, PPP, with rare exceptions, can only be performed in boys with urethral ectopy.

A) Perineal perforation procedure (PPP).

On the X-ray table, under general anesthesia, a cross-shaped skin incision is made in the anal fossa area. After stretching the subcutaneous muscles of the EAS, the child is placed in a lateral position. While compressing the abdomen, and opening of the anal canal, the needle is inserted from the center of the EAS through the open anal canal into the rectum (Figure 7, A-B). Only this stage is performed under fluoroscopic control. The sound of escaping gas indicates that the needle is inside the rectum. Then, a guidewire with a soft floating tip is inserted through the needle into the rectum (Figure 7, C). The needle is removed, and a dense conical bougie with a maximum diameter of approximately 0.8 cm is inserted through the guidewire into the rectum (Figure 7, D). After this, a tracheostomy tube with a diameter of approximately 0.8 cm is inserted into the rectum, and the guidewire is removed (Figure 7, E). The balloon of this tube is inflated with 5 cm^3 of air, which allows the tube to be fixed for 7-10 days. After 10 days, the tube is removed. After two weeks, the wound heals without scarring, and the mother begins to dilate the anus by inserting her little finger.

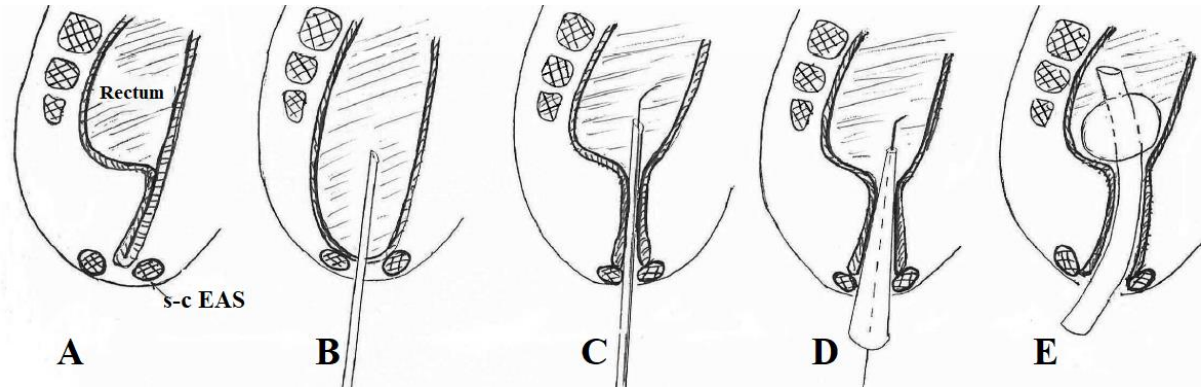

Figure 7. Diagram of the PPP stages. Explanations in the text. **s-c EAS** - subcutaneous portion of the external anal sphincter.

In one case during insertion of a needle into the rectum, no gas escape was heard. Contrast medium was injected to confirm that the needle was in the rectum **(Figure 8).**

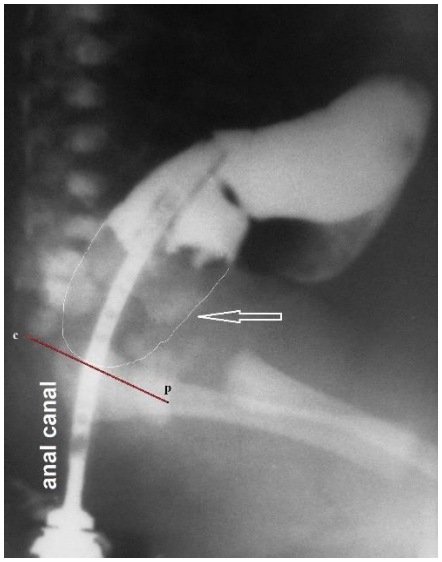

Figure 8. A newborn with an imperforate anus without visible fistula. After needle insertion, no release of gas was heard, leading to doubt that the needle was in the intestinal lumen. A tube was passed through the guidewire, through which a contrast agent was injected. A large amount of meconium (arrow) was in the rectum, blocking the lumen of the needle.

PPP was performed on four newborns without visible fistulas. One of the four children had ARM combined with esophageal atresia. He died from aspiration pneumonia after esophageal correction. Three patients had an uneventful postoperative period. They were recalled for examinations at 1.5, 1.9, and 2.2 years of age, and continence and defecation were normal in each of them. I recommend this method, despite the small number of children operated on, because the method described is scientifically justified. Secondly, the treatment outcomes are strikingly different from all other pull-through operations.

B) Perforation of the perineum from the anal canal using an endoscope inserted through a colostomy

If radiographic examination confirms fistula function, performing a PPP is not possible. In such cases, a colostomy must be performed, and reconstruction should be performed after one year.

In pediatric surgery, there's an axiom that surgery is best performed at an older age rather than in the neonatal period. However, pediatric surgeons performing pull-

through procedures from different approaches prefer to perform definitive correction in the neonatal period, as functional outcomes are independent of the age of the patient's undergoing surgery. This is because complete destruction of the anal canal equalizes functional outcomes in patients undergoing surgery at different ages, as it can't be worse.

The diagnostic method proposed by Pakarinen and Rintala is completely safe and can be used for surgical treatment. The anal canal visualized in the intestinal lumen using retrograde flexible endoscopy through a previously performed sigmoid fistula. The distal end of the anal canal is defined by the convergence of the anal columns. Bright light of the endoscopic from the anal canal indicates that the end of the endoscope is near the anal dimple (Figure 9) [23].

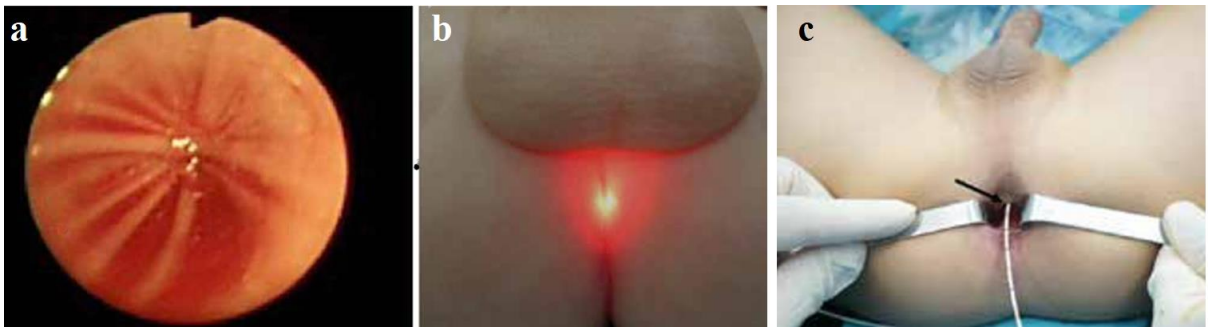

Figure 9. (a-b). Examination of the terminal segment of the intestine using a flexible endoscope inserted through a sigmoid fistula from the article by Pakarinen and Rintala [23]. **(a)**. The image shows a blind-ending anal canal with the confluence of the anal columns. **(b).** The bright light from the endoscope is visible externally because it passes only through the blind-ending, wall of the IAS, subcutaneous tissue, and skin above the anal dimple. **(c).** Examination under anesthesia: the appearance of the anus and sphincter function are normal. The anus opening into the urethra (black arrow) is located 1 cm above the anal verge without visible scar.

These observations confirm the evidence presented above that, in a non-perforated anus, no matter where the ectopic anus opens, there is always a functioning anal canal. Secondly, the ectopia always occurs below the pubococcygeal line. This

means that closure of the ectopic anus is possible through the anal approach. Thirdly, this is a narrowed opening in the wall between urethra and anal canal. The long fistula tract, called a fistula and removed during PSARP, is an IAS, without which fecal continence cannot be normal.

Theoretical justification for reconstruction of ectopic anus using colostomy

If a newborn with an imperforate anus without a visible opening is diagnosed with a functioning fistula and a colostomy is performed, reconstruction with preservation of the anal canal can be performed after one year of age. The anal canal is opened by passing an endoscope through the sigmoid colostomy. The distance from the confluence of the anal columns to the anal dimple is approximately 4 mm. The piercing instrument of the endoscope is inserted externally. The guidewire is retracted into the rectum. The newly created canal is distended using a conical bougie (Figure 10b). The bougie is then removed, and a tracheostomy tube is inserted through the guidewire. The balloon is inflated to secure the tube in the anal canal (Figure 10c). After removal of the tube and completion of anal bougienage with the little finger, the ectopic anus is sutured through the anal approach (Figure 10d).

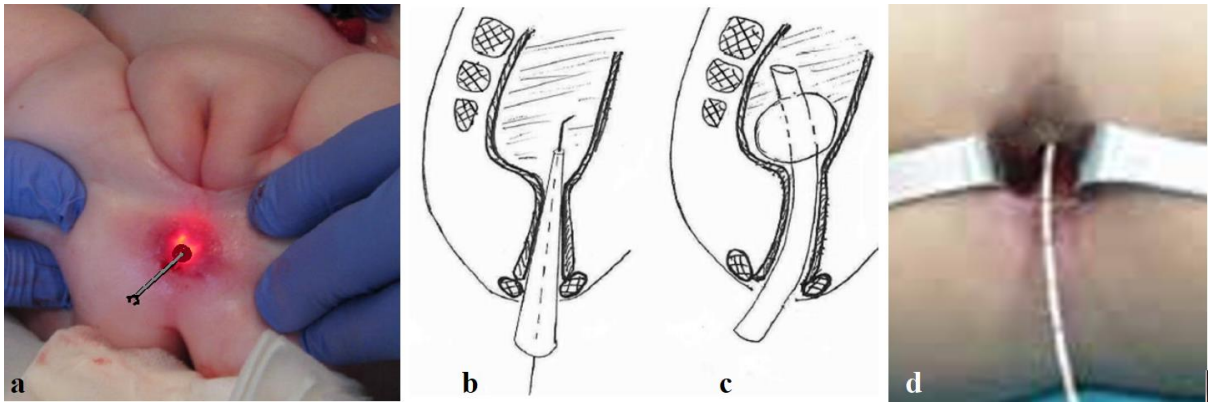

Figure 10. Schematic representation of perineal perforation from a colostomy and closure of the ectopic anus into the urethra or vagina via an anal approach. Explanation in the text.

C) Theoretical justification for the reconstruction of ectopy of the anus with a narrow vagina, erroneously called persistent cloaca.

As demonstrated above, girls with an imperforate anus and supposedly a "single" opening in the vestibule have normally functioning urinary tracts (bladder and urethra), as well as an anal canal. The vagina is represented by a narrow channel because the IAS penetrated the vagina before a cavity had formed in it. Therefore, the IAS's progression through the vagina created a long, narrow fistula tract that opens near the urethra. Sometimes, the IAS, at the point of its penetration into the vagina, blocks the opening from the upper vagina, resulting in a closed cavity where fluid accumulates (hydrocolpos). This mass can compress the urinary tract and cause hydronephrosis and renal failure. Chia described the results of treatment of 59 children with persistent cloaca, in whom, instead of total urogenital mobilization (TUM) and/or urogenital sinus separation (UGS), he used reconstruction involving in situ preservation of the anterior vaginal wall-urethra-bladder neck-introitus complex with selective reconstruction of the posterior vaginal wall and introitus [24]. Thus, he, like some authors before him, proved that this was not a cloaca, and the urinary tract was functioning normally [14].

1. If hydrocolpos is present, its drainage is an emergency procedure. After catheterization of the urethra, a metal pigtail probe should be inserted into the opening near the external urethral sphincter, where feces exit. It should be advanced, screwed under fluoroscopic guidance, until it enters the hydrocolpos cavity. Several dense Hegar bougies can be successively passed through the resulting channel to widen the channel and to leave a Foley catheter into the upper vagina and inflate the balloon to secure it.

2. In newborns, a colostomy operation should be performed, since все corrective surgery in newborns leads to destruction of the anal canal.

3. At the age of over one year, it is proposed to perform perforation of the perineum using an endoscope inserted through the colostomy, as described in the previous text (B).

This evidence-based treatment plan for ectopy anus with narrow vagina, which was called persistent cloaca without any evidence, will preserve the function of the urinary system and anal canal, because of which these children can have normal urinary and fecal continence, without any complications or abnormalities. Gynecologists should perform vaginal expansion or plastic surgery.

Instead of a conclusion

I believe that every pediatric surgeon must ask themselves questions, without answers to which it is impossible to honestly fulfill their professional duty:

1. Why did Alberto Peña's experience prevail over scientific evidence?

2. Why do pediatric surgeons perform pull-through surgery for ARMs with a visible opening, destroying the anal canal, if before the Krickenbeck classification, the anal canal was preserved in these patients, the pathology was called ectopy of the anus, and a simple "cutback procedure" led to normalization of defecation and fecal continence?

3. Why do Kraus et al. in ARMs with urethral fistula describe normal function of the anal canal, call it the rectum, and destroy it using PSARP?

4. Why, in cases of so-called persistent cloaca, without research or scientific justification, are normally formed urethra, bladder, and anal canal still being destroyed, despite scientific evidence that their function is not impaired?

I investigated the reason for the discrepancy between Peña's experience and scientific facts. Peña staged a scientific conference in Krickenbeck (2005). Of the 25 participants (excluding Peña), 23 pediatric surgeons were invited, who had no scientific publications. Probably they had attended Peña's course. He used the support of three surgeons who had publications. However, Rintala claimed the presence of an anal canal in ARMs. Holschneider supported the idea of PSARP based on the erroneous belief that the innervation of the anal canal should be the same as in the rectum. The decision that determined PSARP to be the best procedure for ARMs was unscientific, illegal, and meaningless. However, Peña formally took all pediatric surgeons' hostage. Since then, surgeons cannot publish scientific studies that contradict Peña's experience, which creates the general impression that all scientists support Peña's experience. Secondly, surgeons are hesitant to change treatment tactics because they fear they will be held accountable if failure occurs. Breaking this vicious cycle is only possible by recognizing that Peña's experience contradicts scientific evidence.

5. As a result of the activities of Peña and his associates, patients were taken hostage. As a result of iatrogenic damage to the anal canal and urinary tract, they suffer lifelong fecal incontinence, chronic constipation, and dysfunction of the urinary system, even leading to renal failure.

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