Congenital stenosis of the anus. Pathological anatomy, embryology. Rationale for treatment.

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Abstract

All authors recognize the presence of an anal canal in congenital anal stenosis (CAS). However, there are different views on the stenosis length, which influences treatment methods and functional outcome. Material and Methods. We analyzed radiographic studies of CAS from our own experience and from literature sources to determine the pathological anatomy of CAS using radiometric method. Results. It was shown that all forms of imperforate anus develop similarly during the embryological period, with the formation of the anal canal resulting from the craniocaudal advancement of the endodermal internal anal sphincter (IAS). The absence of anal opening, as well as its stenosis in its usual location, indicates a developmental disorder of the exogenous IAS. Penetration of the IAS through subcutaneous tissue and skin leads to the formation of a narrow, rigid ring. This narrowing applies only to the anus, as the anal canal is normally developed. To avoid confusion with membranous stenosis of the anal canal (previously mistakenly called rectal stenosis), this condition should be called congenital stenosis of the anus (CSA). Conclusion: Congenital stenosis of the anus is a form of imperforate anus, characterized by the presence of a normal anal canal, where the stenosis of the normally located anus ranges from 2 to 5 mm in length. Radiographic examination provides an accurate anatomical and physiological characterization of the defect. Treatment should be initiated as early as possible to prevent the development of megacolon. Dissection of the rigid ring like the cutback procedure can lead to complete recovery.

Keywords: anorectal malformations; congenital anal stenosis; pathological anatomy; congeninal stenosis of the anus; x-ray diagnosis; embryology.

Introduction Congenital anal stenosis (CAS) is defined as a rare condition in which the anus is narrow and located in its normal position, surrounded by the sphincter muscle complex. Sometimes a membrane near the dentate line may accompany [1]. The authors diagnosed stenosis based on the passage of a small-diameter bougie. None of the 27 patients had other accompanying malformations, and no membrane near the dentate line was diagnosed. Members of the Arm-Net Consortium came to a consensus that with CAS, the anus lies in a normal

position, completely surrounded by the sphincter muscle complex, but is too narrow. It may be partly covered by a median bar or membrane, usually located at the dentate line [2]. Instead of providing evidence, a reference is given to a review article, which also contains no studies of the pathological anatomy of this type of ARMs. This formulation contains several contradictions. First, the anus is the opening by which the anal canal ends. It cannot be surrounded by a muscular complex. There is no anatomical category in scientific literature known as "a muscular complex." Normally, the anus is surrounded by the subcutaneous portion of the external anal sphincter (EAS), the length of which is approximately 1/10 of the anal canal [3] **(Figure 1).**

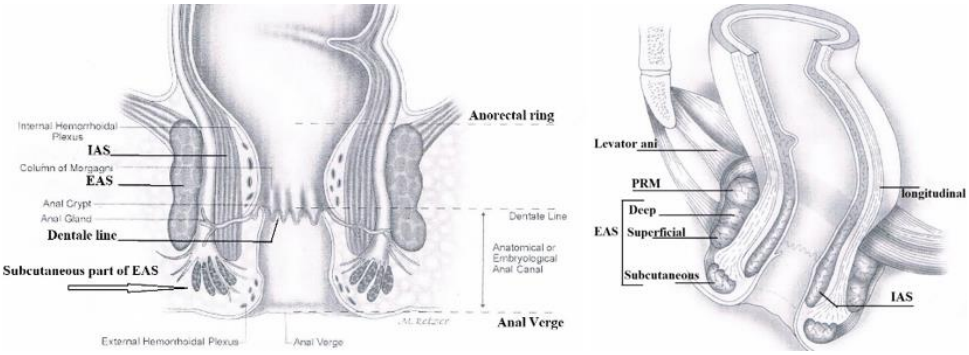

Figure 1. Diagram of the anatomy of the anorectum in frontal and lateral projection from the textbook by Jorge JMN and Habr-Gama A [3].

Second, the remaining 9/10 portion of the terminal intestine, located below the pubococcygeal line, called the anal canal, which surrounded by the internal anal sphincter (IAS), which is responsible for 45% of fecal continence [4]; the puborectalis muscle (PRM), which plays an important role in fecal continence [4,5], and the deep and superficial portions of the EAS, which significantly exceed the subcutaneous portion of the EAS in length and width. The subcutaneous portion of the EAS, ich the authors call ""the muscle complex", is not involved in fecal continence. This is evidenced by the absence of fecal incontinence after its dissection during the "cutback procedure" [6,7,8]. In the diagnostic algorithm proposed in the article by Amerstorfer et al, the diagnosis is based solely on the location of the anus and the maximum caliber of the bougie that freely passes through the anus. This examination allows one to determine the degree of anal narrowing, but not the length of stenosis. The algorithm does not contain recommendations for identifying stenosis at the dentate line level [2]. In the discussion of this article, it is stated that "According to present knowledge, the “fistula” in ARM represents an ectopic anal canal and should be preserved as far as possible to improve the chance for fecal continence" [2]. Unfortunately, this declaration, which was a generally accepted idea until

1982, is not supported by any research or references in this article. Modern research confirms that "The fistula, currently resected during surgical reconstruction for ARM, contains vital structures like the IAS, normal epithelial transition zone and normal ganglion cells... Our results indicate that the fistula has a normal anal canal morphology and should be spared during ARM reconstruction if possible" [9]. Saenz et al., on behalf of the Pediatric Colorectal and Pelvic Learning Consortium (PCPLC), agreed on two types of anorectal stenosis. "Congenital anal stenosis is defined as an anus that lies within an intact sphincter muscle complex but is pathologically narrow. The narrowing of the anal canal is usually located at the dentate line, and patients often present with a skin-lined, 'funnel anus' and is frequently associated with a sacral anomaly." Congenital rectal stenosis (CRS) can be similarly defined as a well-developed, normally positioned anus within an intact sphincter complex but with a pathological narrowing located proximally to the dentate line [10]. There is no evidence for these definitions in the article by these authors, nor in the references they cite. In the definition of CAS, the authors consider the stenotic anus to be at the level of the dentate line. However, the dentate line is formed during the embryonic period at the junction of the endodermal and exodermal rudiments of the internal anal sphincter (see Figure 1). It is located at the border between the upper two-thirds and lower one-third of the anal canal. In a full-term newborn, the true length of the anal canal is 1.7 cm [11]. Therefore, the dentate line is located 0.6 cm above the anal stenosis. This line is not visible or palpable during digital examinations and therefore cannot serve as a diagnostic landmark. Obviously, these authors mistakenly called the cutaneous mucosal junction the dentate line. Secondly, the definition of CAS as "the stenotic anus" contradicts the definition of CAS as "the narrowing of the anal canal", because anus is an opening, and the anal canal is a long channel from the rectum, located above the pubococcygeal line, and the anus.

The definition of "Congenital rectal stenosis (CRS)" as a stenosis "located proximal to the dentate line" is not correct, due to incorrect localization of the "dentate line". As shown by radiographic analysis of 15 published cases of rectal atresia and stenosis, in all observations the membrane was found in the anal canal near the dentate line [12]. Normally, according to Nobles (1984), the anal membrane, arising at the junction of the endodermal and exodermal primordia BAC, ruptures during embryos with a length of 13.5-135 mm [13]. At the site of membrane destruction, a dentate line is determined. If the anal membrane is not destroyed, anal canal atresia (but no rectal) occurs. If the membrane is partially destroyed, anal canal stenosis (but no rectal) occurs, which is an opening in the membrane of varying diameter with rigid fibrous walls [12].

The purpose of the present study is to analyze the literature on congenital stenosis of the anus (CSA) to determine the pathological anatomy and physiology of it.

Characteristics of the pathological anatomy and physiology of CSA according to literature data

1. All authors state that CSA is characterized by a normally functioning anal canal [1-10].
2. The anal opening is narrow. It is surrounded by fibrous rigid tissue. It is initially dilated with a #7 dilator and expanded to the width of a #12 dilator [7]. There are no studies determining the required size of anal dilation [10].
3. Kiely et al. showed that functional outcomes were good when treatment was initiated 4 months of age and poor at older ages. A study of poor outcomes revealed megarectum [14].
4. Saenz et al., for the treatment of CSA, dilation was used in 77% of patients, and surgery was performed in 23% of cases, including posterior sagittal anorectoplasty in 61% of them [10]. Some authors recommend dilation alone [1,2].
5. I did not find any studies about the long-term results of treatment.

Characterization of CSA according to embryology

"Normally, in the post-cloacal period, the endodermal internal anal sphincter migrates in the craniocaudal direction to meet the ectodermal portion" [15]. It passes within the pelvic floor muscles, through the puborectalis muscle (PRM) loop, and within the deep and superficial parts of the external anal sphincter (EAS). The largest group of anorectal malformations is imperforate anus. This means that the anal opening is absent or severely narrowed due to impaired development of the exodermal IAS. The anatomy and physiology of the anorectum with imperforate anus is shown in radiographs with visible fistulas in **Figure 2.**

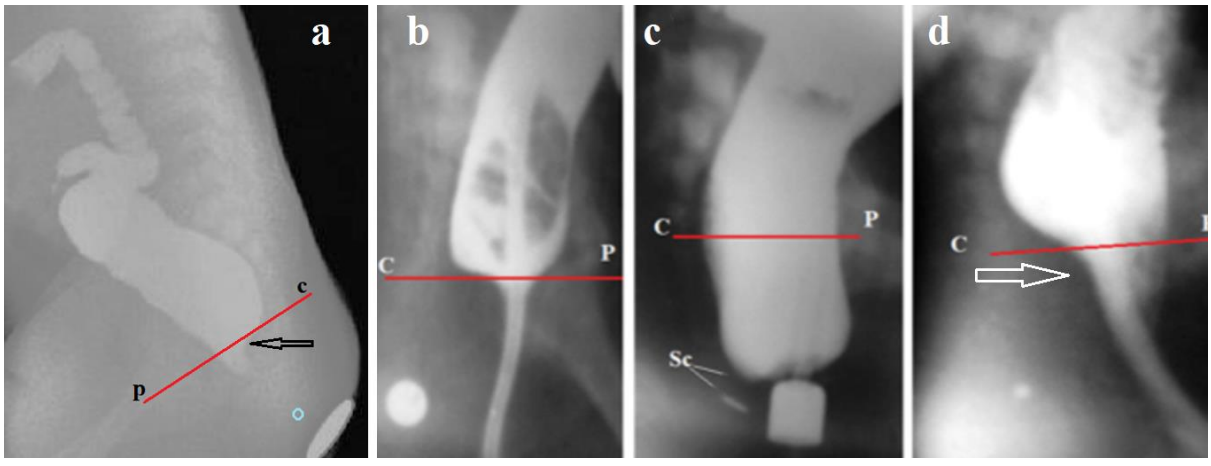

Figure 2. (a) From Menon et al [16]. A 22-day-old full-term male child with constipation; he was found to have a normally positioned but small anal opening. The authors believed that the

distal colostogram demonstrated dilated rectosigmoid with anal stenosis and an atretic anal canal extending from the anorectal junction to the anal verge, measuring 3.9 cm in length. Therefore, pull-through was done. My analysis. The disc marker is located on the buttocks. However, the anus is located deeper between the buttocks (blue circle). The length of the anal canal between the pubococcygeal line (red) and the anus in infants is no more than 2 cm, not 3.9 cm. The arrow indicates the depression resulting from contraction of the puborectalis muscle. This is normal anal canal, which is in a contracted state and which the authors of the article mistook for a long stenosis and removed. **(b)** Lateral radiograph of the anorectal area of a 9-month-old patient, performed after barium was introduced into the rectum through a catheter passed through 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. **(c)** 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 (b), its absence around the ectopic anus does not affect the function of barium (feces) retention. **(d)** During the introduction of barium through the endotracheal tube, relaxation of the IAS occurred, resulting in barium entering from the rectum into the upper part of the anal canal in front of the enema tip. At the same time, the posterior portion of the IAS was pressed against the tube due to contraction of the PRM. At the same time, the deep and subcutaneous parts of the EAS also contracted to prevent barium leakage. This is the radiographic equivalent of the rectoanal inhibitory reflex [11].

Analysis of Figure 2 demonstrates that congenital anal stenosis is a pathology caused by a disruption in the formation of the exodermal rudiment of the anal canal. During embryonic development, craniocaudal progression of the endodermal IAS creates a normally functioning anal canal, which reaches the subcutaneous tissue, which contains the subcutaneous portion of the EAS. Without meeting the exodermal anal canal along the way, in 1-2% of casest penetrates the skin of the anal fossa. This penetration creates a narrow, rigid canal, the length of which is equal to the thickness of the skin and subcutaneous tissue. In newborns, their length is 2 mm, and in infants, 4 mm. In 45% of cases, the anal canal shifts forward and slightly upward along the midline, creating a perineal ectopia ani with a narrow, rigid opening. Figures (a, b) show that the anal canal contracts at rest, serving as a fecal continence barrier due to contraction of the IAS, the deep and superficial portions of the EAS, and the PRM. It also opens wide (c)

during defecation due to contraction of the levator plates. The subcutaneous portion of the LES occupies a minimal amount of space, and these patients do not experience fecal incontinence until megacolon develops. Transection of the ectopic anus along with the subcutaneous portion of the EAS during the cutback procedure does not cause fecal incontinence [6, 7, 8].

The endodermal IAS continues to move until it emerges (CSA, perineal and vestibular ectopy) or penetrates a cavity (the urethral ectopy in boys and the vaginal in girls). However, by the time of penetration, in all cases, a functioning anal canal has already been formed. For example, in an article Kraus et al, to the augmented-pressure distal colostogram, the authors emphasized: "It is extremely important in this regard to understand that the lowest part of the rectum is usually collapsed from the muscle tone of the funnel-like striated muscle mechanism that surrounds the rectum in 90% of cases…" [17]. These authors describe the anatomy and physiology of the anal canal, calling it the rectum. However, it is known that there are no muscles around the rectum. They exist only around the anal canal, and they contract, performing the function of fecal continence. Thus, the authors confirmed that in urethral fistulas there is a functioning anal canal, which, as normal, opens under high rectal pressure [11]. (see Figure 2) **(Figure 3).**

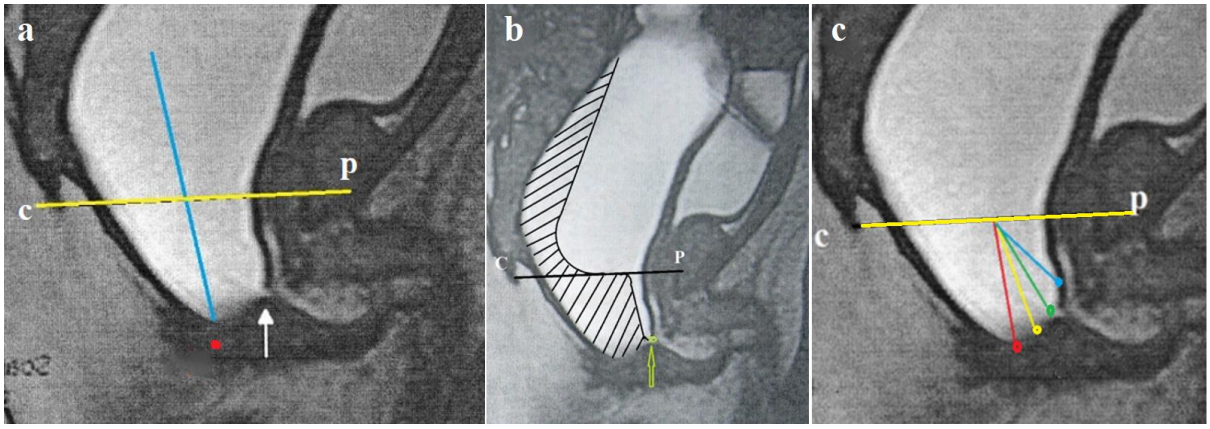

Figure 3. Augmented-pressure distal colostogram during MRI. **(a)** The anal canal located distally to the pubococcygeal line opened under high rectal pressure. The caudal wall of the IAS approached the anal dimple. The subcutaneous part of the EAS is in space between the wall of the IAS and marker near the anal dimple (red spot). The arrow shows the opening between the anal canal and urethra – bulbar ectopy of the anus. **(b)** Schematic diagram of anal canal contraction with low rectal pressure. The IAS is attached to the fistula opening. Therefore, IAS is displaced anteriorly and shorter than normal. **(c)** ARM variants: congenital stenosis of the anus – red, perineal ectopy – yellow, fistula into the bulbar urethra – green, fistula into the prostatic urethra – blue.

The above radiographic findings of normal anorectal anatomy and physiology allow for a new assessment of the described cases of the Currarino Triad. Below is an analysis of two well-examined patients.

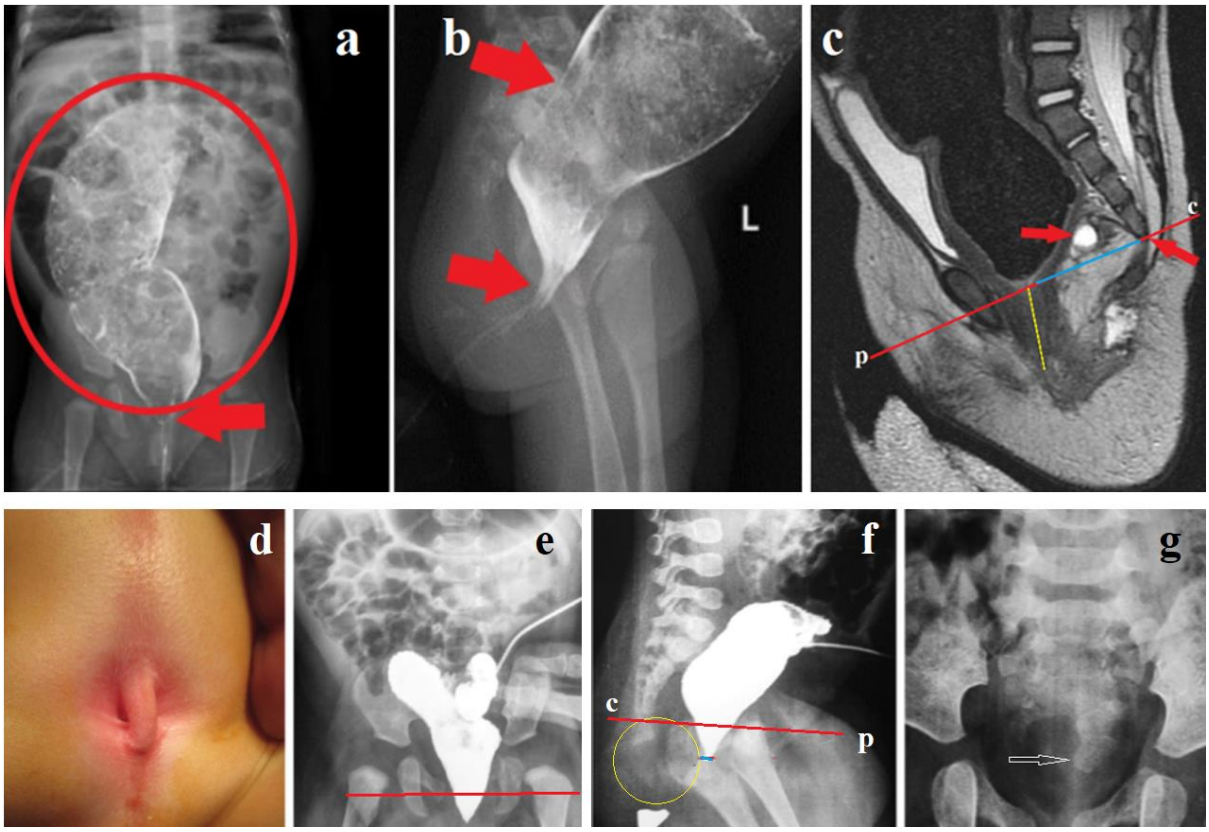

Figure 4. Two cases of patients operated on the Currarino Triad using PSARP.

Case 1(a-c), described in the article by Aldabbab et al [18]. An eight-month-old baby girl had progressive chronic constipation for the last two months. "Per rectal examination revealed stenotic anus with no gush of stools upon withdrawal of the examiner's finger". On the radiograph after the introduction of barium, the authors describe "a stenotic short segment at the distal rectum with a significantly distended sigmoid colon" (**Figure 4 a,b**). "MRI impressive of presacral lipoma-myelomeningocele" (Figure 4c). Biopsy of the presacral mass confirmed a mature teratoma. A patient was diagnosed with Currarino syndrome and managed surgically with excision of the presacral mass and an anorectoplasty via a posterior sagittal midline incision. My analysis confirms marked dilation of the rectum and sigmoid colon. The dilated retrorectal space is caused by anterior displacement of the rectum and anal canal by the teratoma. However, the "stenotic short segment at the distal rectum" that led to the anorectoplasty is a contracted, anteriorly displaced anal canal. **(c)** Chronic constipation is due to the anterior displacement of the rectum (blue part of the pubococcygeal line), and of the normal anal canal, resulting in incomplete opening of the anal canal during defecation. There

is no evidence of ARM because penetration of a finger through the anus is generally accepted evidence of the absence of stenosis.

Case 2 (d-g), described in the article by AbouZeid et al [19]. A male patient was diagnosed with an anorectal malformation at birth (Figure 4d) and underwent a colostomy. At 3 months of age, a distal colostogram was performed, which, according to the authors, showed a "blind termination of the rectum," which was classified as "anorectal atresia without fistula." His six-year-old sister underwent surgery for Currarino's triad. The boy underwent removal of a retrorectal cyst approximately 2 cm in diameter and anorectoplasty through a posterior sagittal approach. My analysis. The diagnosis of Currarino's triad is certain, as all three features are present: (1) a covered anus, (2) a space-occupying lesion approximately 2 cm in diameter, displacing the rectum and anal canal anteriorly (Figure 4f – yellow circle ≈ 2 cm in diameter), and (3) a saber-shaped notch of the sacrum, which is displaced to the left (Figure 4g – arrow). However, no classification system includes a diagnosis such as "anorectal atresia." ARMs such as rectal atresia and imperforate anus are known. A covered anus is one of its forms. The authors claim that the rectum terminates blindly, meaning that the entire distance from the rectum to the anal pit represents an atretic segment of the intestine, but such cases have not been described. Figures 4e and f show relaxation of the IAS with penetration of barium into the upper part of the anal canal in the form of a cone while the posterior wall of the anal canal is pulled forward by a contracted of PRM. This is the radiographic equivalent of the rectoanal inhibitory reflex (see Figures 2a and 4b). As shown above, imperforate anus occurs due to impaired development of the exogenous IAS. However, the endogenous IAS always reaches the subcutaneous tissue. This means that the patient had a normally functioning anal canal, which should have been preserved, rather than warning parents about the possibility of chronic constipation and fecal incontinence.

Discussion

Congenital anal stenosis (CAS) is a rare condition. In 2024, the European Anorectal Malformation Network (ARM-Net) recorded a 2% incidence of CAS over a 10-year period [20]. In a population-based cohort study on the diagnosis and early management of anorectal malformation in the UK and Ireland, no cases of CAS were recorded among 174 cases from 26 centers during 2015–2016 [21]. In an article by Hageman et al., the incidence of different types of ARMs in Europe and Australia was compared. The incidence of CAS was 3.7 times higher in Australia than in Europe. Only perineal fistulas were 6.6% higher in Europe than in Australia. The incidence of other types of ARMs was virtually identical [22]. Except for perianal fistulas

in females, the incidence of different types of ARMs was similar in Italy and South Africa [23]. An analysis of the literature reveals clear patterns. (a) The frequency of different types of ARMs is identical across continents and among different races, with minor deviations in perineal fistulas and sharp differences among CAS. (b) Visible fistulas (perineal and vestibular) account for approximately 60%. The higher the ectopia of the anus, the less common these anomalies are. (c) The frequency of CAS not only varies sharply across population studies but is also significantly less common than other types of ARMs with visible fistulas (20 times less common than perineal ectopia of the anus). These data indicate that the genetic factor is decisive in the etiology of ARMs. Some difference in the frequency of perineal fistula (6.6%) can be explained by the fact that some authors adhere to the Krickenbeck classification [22], while others use the diagnosis of "anterior anus", which is not included in the Krickenbeck classification [2,21]. There is no clear distinction between perineal fistula and anterior anus. In the former case, the incidence of CAS is 2.1% [22], while in the latter, it is 0% [21]. A contradiction to the patterns described above is evident when comparing two articles. None of the 26 medical centers in the UK and Ireland reported CAS over the course of a year [21], while another center, not specialized for colorectal patients, reported 27 cases of CAS over 4 years (6.7 cases per year) [1].

All authors provide the same definition of CAS, which has remained unchanged since the mid-20th century. Congenital anal stenosis is defined as an anus that lies within an intact sphincter muscle complex but is pathologically narrow. The narrowing of the anal canal is usually located at the dentate line» [1, 2, 10]. All authors repeat this formulation, sometimes without evidence adding the possibility of vertebral anomalies or a combination with a skin-lined, 'funnel anus'. However, there are no studies in the literature that precisely localize the site of obstruction. The ARM-Net Consortium and the Pediatric Colorectal and Pelvic Learning Consortium (PCPLC) publish literature reviews that define the status quo. They do not contain any studies or new hypotheses. To diagnose CAS, a perineal examination and determination of the degree of anal stenosis by inserting Hegar dilators are recommended. These studies cannot determine the length of the stenosis. It is surprising that all authors mistakenly call the mucocutaneous junction of the anus the dentate line. This can only be explained by the fact that consensus reports are written by one author, and others sign off on the finished text. It is impossible to imagine another option: that dozens of the surgeons operating children with ARMs did not open textbooks on anorectal anatomy. The definition described above contains another error. On the one hand, CAS is defined as a narrowed anus and, at the same time, as a narrowing of the anal

canal. In this definition, as in all literary sources, the anus is mistakenly identified with the anal canal.

In fact, all authors refer to two different types of ARMs by the term CAS, which have different embryonic development and require different surgical treatments. Normally, in the post-cloacal period, the endodermal internal anal sphincter migrates in the craniocaudal direction to meet the ectodermal portion [15]. In the 5th week, the ectoderm invaginates to form the anal pit. In the 6th week, a membrane forms between the endodermal and ectodermal portions of the IAS. In the 7th week, the anal membrane ruptures to establish continuity between the upper and lower anal canal [13, 24]. There is reason to hypothesize that absence of the anal membrane to rupture leads to anal canal atresia at the level of the dentate line, while incomplete rupture of the anal membrane leads to anal stenosis. These conditions were previously called atresia and stenosis of the rectum, although it is known that the dentate line is in the anal canal (see Figure 1). It cannot be confused with stenosis of the anus, since in anal canal atresia the anus and lower part of the anal canal are normally developed. Congenital stenosis of the anus (CSA) develops during the embryonic period because of impaired formation of the exodermal part of the anal canal, as in all types of imperforate anus. Importantly, the endogenous bud of the anal canal always creates a normally functioning anal canal, except for the distal 2-4 mm. This is the length of congenital stenosis of the anus (CSA) [25].

In all articles, the term "anal" refers to both the anus and the anal canal, leading to misconceptions about the defects. If we exclude the phrase "Sometimes a membrane near the dentate line may accompany" from the generally accepted CAS definition, as it denotes stenosis of the anal canal, we are left with a completely different type of ARMs: "the anus is narrow and located in its normal position," i.e., "congenital stenosis of the anus." (CSA).

In CSA, the anal canal functions normally. The problem lies in the narrowing of the anus, which impedes normal rectal emptying. As a result, large amounts of fecal material accumulate in the rectum, causing its dilation and megacolon, leading to severe constipation. Due to the force of rectal peristalsis, large fecal stones distend and weaken the pelvic floor muscles (PRM, levator plates), leading to descending perineum syndrome, which clinically manifests as fecal incontinence [26,27]. Early diagnosis and treatment of CSA can prevent chronic constipation and fecal incontinence. With CSA, a narrow opening is a fibrous, rigid ring that is histologically indistinguishable from ectopic anus with visible fistulas.

Numerous articles convince us that dilation can treat CAS. However, there is no evidence that this method can cure patients, as not a single article reports long-term results after dilating the stenosis with Hegar dilators. Furthermore, this is impossible from an anatomical and

histological perspective. Treatment begins with inserting a #7 Hegar dilator into the narrow, rigid opening, increasing its diameter by 1 mm weekly, and ends with the free insertion of a #12 or #17 dilator. The opening diameter gradually increases due to ruptures of the fibrous fibers. However, the rigid ring does not become elastic and does not increase in size with age. With age, the diameter of the rectum, where feces are formed according to its capacity, increases. Since the anus does not widen with age, feces cannot pass through the opening that previously ensured normal bowel movements. This inevitably leads to severe constipation. Anterior or posterior anorectoplasty damages elements of the anal canal, which negatively affects the function of fecal continence and defecation for the rest of the patient's life. For example, Gauthier and Valayer presented a series of 8 cases of congenital anorectal stenosis with a follow up of 1 to 7 years. Three of them had fecal incontinence [28].

There are three compelling scientific facts that must be considered to preserve the anal canal. (a) Anal dilation of the narrow anus increases its diameter but does not eliminate rigidity. Therefore, it leads to megacolon and unnecessary delay in surgery. (b) The reports on the use of the cutback procedure have shown that transecting the subcutaneous portion of the EAS along with a rigid ectopic anus does not lead to canal incontinence [6, 7, 8]. (c) Suturing the edges of the wound formed after cutback leads to narrowing of the opening, an inflammatory reaction, and the formation of scar tissue. Therefore, after the cutback procedure, it is necessary to dilate the anus for several weeks. Browne was the first to show that the edges of the wound should never be sutured, since after a few weeks it will fill with elastic tissue [7]. My experience confirms this phenomenon [29]. These facts provide the scientific basis for double cutting of the anal stenosis ring along with the subcutaneous part of the EAS like cutback procedure.

Conclusion

Congenital stenosis of the anus is a form of ARMs characterized by the presence of a normal anal canal, where the stenosis, surrounded by the subcutaneous portion of the external anal sphincter, measures 2 to 5 mm in length, depending on age. Radiographic examination provides a precise anatomical and physiological characterization of the defect. Treatment should be initiated as early as possible to prevent the development of megacolon. Dilation of the stenosis does not resolve the rigidity of the fibrous ring. Dissection of the rigid ring with insertion of a tube into the anal canal can lead to complete recovery.

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