Dear GEORGE W. HOLCOMB, III, M.D., MBA Editor-in-Chief Journal of Pediatric Surgery,

The article by Zoe et all "Can Anorectal Stenosis be Managed with Dilations Alone? A PCPLC Review" was recently published as Journal Pre-proof. The authors define congenital anal stenosis (CAS) as the narrowing of the anal canal is usually located at the dentate line and lies within an intact sphincter muscle complex. Congenital rectal stenosis (RS) defined "as a well-developed, normally positioned anus within an intact sphincter complex but with a pathological narrowing located proximal to the dentate line". These definitions do not correctly describe the pathological anatomy of these defects. Firstly, the authors combine in one article different diseases, which are united only by the word "stenosis". Scientists distinguish CAS from rectal atresia or rectal stenosis (RA/RS) [1, 2]. Secondly, in the sources cited by the authors {2,3}, there are no studies confirming these anatomical definitions. Thirdly, with CAS the anus is an opening that opens within the subcutaneous portion of the external anal sphincter (and not the sphincter complex) and is always represented by a narrow rigid ring, the length of which is equal to the thickness of the skin and subcutaneous tissue from 2 mm in newborns to 4-5 mm in infants [3] (Figure 1a).

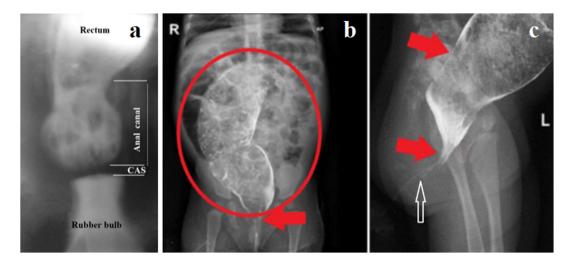


Figure 1. (a). A 7-month-old child with CAS with secondary megacolon. The introduction of barium from the rubber bulb provoked a defecation reflex, resulting in a wide opening of the anal canal with the penetration of barium and feces into the anal canal. The enema tip completely obstructed the narrow opening of the anus. The length of the constriction (CAS) is 3 mm (the gap without contrast between the anal canal and the pear). (**b-c**). Radiographs from the article by Aldabbab et al, where the authors with a red arrow "showed evidence of a stenotic short segment at distal rectum" [4]. However, this segment is the closed anal canal. The location of the anal stenosis is shown by me with a white arrow at the end of the contrast agent traces in the anal canal next to the catheter. This error in understanding the physiology of the anorectum is a formal justification for pull-through surgery that destroys the anal canal.

The dental line is the junction of the endodermal and ectodermal rudiments of the anal canal and divides the anal canal into upper (two-thirds) and lower (one-third) parts. Thus, with CAS there is a normally functioning anal canal. Stenosis does not occupy the lower part of the anal canal (at the dentate line), but only the site of perforation of the internal anal sphincter of the skin and subcutaneous tissue. As correctly stated by some of these authors in another article: - "The aim of the definitive surgical repair in such cases is to preserve the anal canal" [5]. As shown in our study, rectal stenosis and rectal atresia are in the anal canal, and not in the rectum, as previously thought. X-ray analysis of RA/RS radiographs revealed a membrane in the anal canal at 1 cm from the anus, which allows for correction through the anus [6]. The authors further write that "Recent studies have now proposed the use of dilation as the primary treatment modality to potentially defer or eliminate the need for surgical repair" {4}. In the article to which the authors refer (1997) an unusual case is described, which is not a scientific study. The link to it is not correct.

The authors state that they «performed a multi-institutional retrospective review of patients with anorectal stenosis to characterize management and describe outcomes using the Pediatric Colorectal and Pelvic Learning Consortium (PCPLC) registry".

In the discussion indicates that "most patients with anal stenosis 44 (77%) of 57 and rectal stenosis 6 (85.7%) of 7 were successfully treated with dilations alone without requiring surgical intervention during the follow up period. However, in the section "Results" there is no information about the condition of the colon and constipation, and there are no comparisons of treatment results after different operations (anterior anorectoplasty, cutback anoplasty, and the posterior sagittal anorectoplasty). In the article modified PSARP was recommended with a link to an article in which was presented «case of rectal atresia and anal stenosis" {9} [7]. The results of this operation are not described anywhere. It was not used by the authors of the Pediatric Colorectal and Pelvic Learning Consortium.

It is surprising that the authors do not mention the main problem of treating stenosis - the difficulty of preventing the development of megacolon and associated severe constipation, which require long-term treatment throughout life. Any anorectal stenosis causes expansion of the rectum and left half of the colon. With a long course, the muscles of the perineum (puborectalis muscle and levator plates) are damaged. The authors do not mention the X-ray method of examination, without which it is impossible to determine whether there was megacolon at the time of dilatation or not (which is unlikely). In the results of the study: -

"Specific details such as initial and final dilation size and duration of intervention before considered a failure of management were not available". However, the text contains an unreasonable and practically impossible estimate of severe stenosis, as being more than 50% narrower than would be expected for the patient's age. In addition, dilation is more successful if the dilator is only slightly smaller than normal. These discussions make no sense without indicating age standards and the actual width of the rectum.

Dilation of the rigid annulus results in a gradual increase in diameter, but the annulus remains fibrous. Since dilation begins late, when megacolon has already appeared, then already at this stage a discrepancy arises between the width of the feces and the throughput of the fibrous ring. Secondly, the diameter of the dilated but rigid fibrous anus does not increase with age, while the width of the rectum, in which feces are formed, increases with age from 1.3 - 3.0 (2.24 ± 0.09 cm) in the first year of life to 3.6 - 4.6 (3.95 ± 0.07 cm) at 11-15 years [8]. This discrepancy is the cause of the development of megacolon and severe constipation.

Solving the problem is in understanding pathological physiology. In CAS, in which there is a normal anal canal, the narrow rigid ring has a length of 2 to 5 mm. Cutting the ring in at least two places and inserting a tube into the anal canal with a balloon inflated in the rectum to secure the tube will result in the development of elastic tissue in the spaces between the fibrous areas. This will allow the anus to expand as the diameter of the stool increases. This operation is functionally like the cutback procedure for perineal fistulas in boys, which leads to good functional results [9]. The PSARP modification that the authors recommend has never been used, its results are not described anywhere, theoretically it is not rational, since it destroys the anal canal.

As shown in our study, the pathology, which is commonly called RA/RS, is a membrane in the anal canal at ≈ 1 cm from the anus. It is easily excised from the anal approach, as described in the article of Chowdhury et al in a patient with rectal stenosis [10].

Note. In curly brackets { } are links from the article being reviewed. In square brackets [] are my links.

References

- Hamrick M, Eradi B, Bischoff A, Louden E, Peña A, Levitt M. Rectal atresia and stenosis: unique anorectal malformations. J Pediatr Surg. 2012 Jun;47(6):1280-4. doi: 10.1016/j.jpedsurg.2012.03.036.
- de Beaufort CMC, Gorter RR, Iacobelli BD, Midrio P, Sloots CEJ, Samuk I, van Rooij IALM, Lisi G; ARM-Net Consortium. Rectal atresia and rectal stenosis: the ARM-Net Consortium experience. Pediatr Surg Int. 2023 Jul 28;39(1):242. doi: 10.1007/s00383-023-05518-7.
- 3. Levin MD. The pathological physiology of the anorectal defects, from the new concept to the new treatment. Eksp Klin Gastroenterol. 2013; (11):38-48.
- Aldabbab HY, Al Ghadeer HA, Alnosair AA, et al. Complete Currarino Triad Presenting With Chronic Constipation. Cureus. 2022 Apr 1;14(4):e23743. doi: 10.7759/cureus.23743.
- Lane VA, Wood RJ, Reck C, Skerritt C, Levitt MA. Rectal atresia and anal stenosis: the difference in the operative technique for these two distinct congenital anorectal malformations. Tech Coloproctol. 2016 Apr;20(4):249-54. doi: 10.1007/s10151-016-1435-5.
- Levin MD. X-ray studies of rectal atresia and rectal stenosis. <u>https://www.anorectalmalformations.com/_files/ugd/4d1c1d_e299b7ebc8ce447daf57b</u> <u>3c0cd6423c5.pdf</u>.
- Lane VA, Wood RJ, Reck C, Skerritt C, Levitt MA. Rectal atresia and anal stenosis: the difference in the operative technique for these two distinct congenital anorectal malformations. Tech Coloproctol. 2016 Apr;20(4):249-54. doi: 10.1007/s10151-016-1435-5.
- Levin MD. Radiological anatomy of the colon and rectum in children. Gastroenterology & Hepatology. 2019; 10 (2):82-6.
- Kyrklund K, Pakarinen MP, Taskinen S, Rintala RJ. Bowel function and lower urinary tract symptoms in males with low anorectal malformations: an update of controlled, long-term outcomes. Int J Colorectal Dis. 2015 Feb;30(2):221-8. doi: 10.1007/s00384-014-2074-9.
- Chowdhury TK, Hoque MM, Kabir M, Banu T. Delayed presentation of congenital rectal stenosis associated with Down's syndrome and hypothyroidism: Case report. Clin Case Rep. 2021 Nov 16;9(11):e05083. doi: 10.1002/ccr3.5083.

Dear author,

Your manuscript titled "Analysis of recommendations of the pediatric colorectal and pelvic learning consortium on the diagnosis and treatment of anal and rectal stenosis", has been carefully evaluated by our editorial consultants.

I regret to inform you that this will not be published in the Journal of Pediatric Surgery. Unfortunately, the manuscript received a low priority score. Many manuscripts are submitted to the Journal. However, only those that achieve a high content rating and priority scoring can be published.

We appreciate your submitting your work to the Journal.

Sincerely,

GEORGE W. HOLCOMB, III, M.D., MBA Editor-in-Chief Journal of Pediatric Surgery (05/18/24)