Archives of Gastroenterology and Hepatology

ISSN: 2639-1813 | Volume 4, Issue 1, 2021

DOI: https://doi.org/10.22259/2639-1813.0401001

Diagnostics and Treatment of Fistula Forms of Rare Regional Variants of Anorectal Malformations in Children

Otamuradov F.A.

Termez branch of the Tashkent Medical Academy, Uzbekistan.

**Corresponding Authors:* *Otamuradov F.A., Termez branch of the Tashkent Medical Academy, Uzbekistan.*

Abstract

Aim: *The aim of the study was to study the frequency of nosological forms, features of clinical and anatomical variations, the nature of combined anomalies and the choice of diagnostic and treatment methods for fistulous forms of rare regional variants of ARM in children.*

Materials and Research Methods: *In the clinical bases of the Department of Hospital Pediatric Surgery of TashPMI in 2009 - 2019, 504 children with ARM were examined and treated. At the diagnostic stage, clinical and laboratory, instrumental and special research methods.*

Results: *In our observation, 121 children had rare regional variants of anorectal malformations. These forms accounted for 24.1% in the general structure of ARM with fluctuations from 0.83% to 24% among various nosological forms; prevailed among girls - 95 (78.5%), among boys - 26 (21.5%). Ectopia of the anus was observed in 25.6% of cases; H-form rectogenital fistulas with a normally formed anus in 24%; atresia with rectovaginal fistula in 18.2%. Cases of rectal duplication, exstrophy of the cloaca, and urogenital fistula are presented. The nature and frequency of concomitant pathology in various nosological forms have been analyzed.*

Keywords: *anorectal malformations, rare regional variants, diagnosis.*

Introduction

The frequency of anorectal malformations (ARM), according to various authors, ranges from 1.66 to 9.94 per 10,000 children and does not tend to decrease [1,2]. The large variation in the ARM frequency can be explained by the organization and registration of malformations. It should be noted that individual forms of anorectal anomalies differ in frequency in different regions of the world. In France, Sweden and the United States, the average frequency of ARM was 3.4 per 10,000 children, with a larger difference in populations[3]. According to H.H. Almaramhy (2012), the incidence of anorectal anomalies in Saudi Arabia for 1998-2010 was 9.94 per 10,000 births, which is higher than in Europe, America and China[4]. The international conference dedicated to the standardization of the assessment of the outcomes of anorectal anomalies in Krikenbeck (Germany) in 2005, a classification of the ARM was adopted, which distinguishes between the main clinical forms and rare regional variants[5,6,7].

The aim of the study was to study the frequency of nosological forms, features of clinical and anatomical variations, the nature of combined anomalies and the choice of diagnostic and treatment methods for fistulous forms of rare regional variants of ARM in children.

Materials and Research Methods

In the clinical bases of the Department of Hospital Pediatric Surgery of TashPMI in 2009 - 2019, 504 children with ARM were examined and treated. There were 258 girls (51.2%), 246 boys (48.8%) aged from 1 day to 15 years. The patients were distributed in accordance with the international classification adopted in Krikenbek (2005), highlighting the main

(383-76%) and rare regional forms (121-24%). At the diagnostic stage, clinical and laboratory, instrumental (X-ray, ultrasound, MSCT) and special research methods were carried out, aimed at determining the clinical and anatomical forms of the ARM, assessing the anatomical and functional state of the anorectal zone and the obturator of the rectum.

Results and its Discussion

Among 504 children with ARM, the main clinical forms of pathology were observed in 383 (76%). Of these, there were 220 boys (57.4%), and 163 girls (42.6%). Rare regional forms were diagnosed in 121 (24%) patients; 95 (78.5%) girls and 26 (21.5%) boys. By the nature of the pathology, the frequency of individual nosological forms, depending on the sex of the patients, differences were revealed. In girls, the frequency of ARM moderately prevailed with a greater variety of nosological forms, some of them were not observed in boys; in girls, rare regional variants of the anomaly significantly prevailed (95 - 36.8% of 258) compared with boys (26 - 10.6% of 246). With an anomaly occurring in individuals of both sexes, differences in their numbers were noted.

Rectovaginal fistula (22 girls) accounted for 4.3% and 18.2%, respectively, in the structure of rare regional forms with ARM. 17 of them were initially hospitalized in our clinic, 5 were admitted from other medical institutions after operations (sigmastoma imposition - 1, fistula recurrence - 4). Mucopurulent discharge from the vulva between acts of defecation in girls after discharge from the maternity hospital with a diagnosis of rectovestibular fistula was the main reason for going to a surgical hospital. One girl was 3 months old, 8 - up to one year, 8 - up to three years old with severe symptoms of vulvovaginitis. That is, children with rectovaginal fistulas were sent to a surgical hospital with a delay. In 13 (59.1%) girls, due to the sufficient diameter of the fistulous course, there was no stool retention. In 9 (40.9%) cases, obstructed bowel movements were periodically noted. In all cases, the correct diagnosis was made in the clinic. The main diagnostic method is a thorough examination of the perineum in order to differentiate pathology with a rectovestibular fistula in which, as a rule, it is possible to determine the fistulous passage in the vestibule of the vagina. In 8 (42.1%) patients, the rectum with a fistula opened into the distal part of the vagina. Isolation of intestinal contents from it in the directional diagnosis was mistakenly regarded as a rectovestibular fistula. In 6 (27.3%) cases, the fistula was localized in the proximal vagina with a characteristic discharge of intestinal contents through the hymen, which was also mistakenly taken as a rectovestibular fistula. The diagnosis of a rectovaginal fistula is an indication for a preventive colostomy.

H-type fistulous forms were detected in 30 patients - this is, respectively, 5.9% and 24.8% in the general structure and among regional forms of ARM. Among them, girls predominated - 29 out of 30. It is difficult to establish the exact causes of the H-type fistula. They can be congenital or due to the postponed, inflammatory process, complications of the postponed perineal surgery, or due to a combination of these phenomena. In 28 (93.3%) patients, the pathology was regarded as congenital. Of these, 24 (85.7%) girls were born with a rectogenital fistula with a normally formed anus, 1 (3.3%) - with a stenotic anus and rectum. Only in 1 (3.3%) boy, before the operation, a rectourethral fistula was diagnosed with a normally formed anus.

2 girls in connection with a perianal abscess at the age of 1 and 2 months. conservative treatment was carried out at the place of residence. In three cases, the fistula arose against the background of the transferred inflammation. The simultaneous presence of rectovestibular and pararectal fistulas was noted in one patient. Indirect signs confirming the onset of the process as a complication of the postponed purulent inflammation in the perineum are deformation of the labia and sclerotic changes in the mucous membrane in the vestibule of the vagina and adjacent areas of the perineum. In the rest of the girls, the mouth of the fistula in the rectal mucosa was localized at the level of the dentate line and closer to the labia minora on the right or left. Histological examination of the resected fistulous tract in most children revealed an epithelial lining with squamous epithelium, a circular arrangement of muscles with nerve plexuses, corresponding to the intestinal structure and indicating a congenital nature.

In most cases, the clinical diagnosis of H-type fistulas in girls was not difficult. The main importance in establishing a diagnosis is examination of the perineum, external genital organs and rectum. In 28

(96.6%), the anus was formed normally; 1 (3.4%) girl had stenosis. In 24 (82.8%) patients, the localization of the anus was typical, in 5 (17.2%) - with mixing; in 3 it was insignificant (index of anal position - 38), in 1 - noticeable (index of anal position - 22). In 18 (62.1%) patients with a fistulous passage more than 5 mm in diameter, the site of the fistula opening in the vestibule was clearly visible. In 6 (20.7%) patients with a narrow fistulous course and in 5 (17.2%) patients with a rectovaginal fistula, the localization of the fistula could be established using a button-like probe inserted through the mouth of the fistula from the vestibule of the vagina or in the lumen of the rectum.

The mouth of the fistula in the lumen of the rectum was localized at different distances from the mucocutaneous junction of the rectum: to the dentate line - 7 (24.1%), at the level - 13 (44.8%), above - 9 (31.1%). In 6 (20.7%) patients, the localization corresponded to the anovastubular H-type - "perineal canal"; in 11 (37.9%) rectovubular - intermediate form; in one of them, with rectovestibular communication, a pararectal fistula was also noted; 12 has a tall form.

Of 52 boys, 1 (1.9%) had a rectourethral fistula before surgery with a normally formed anus of a congenital nature. The diagnosis was confirmed by contrast urethrocystography. In 51 (98.1%) cases, rectourethral fistulas were established after primary perineal surgery for anorectal anomalies (presumably; as not diagnosed before and during surgery or iatrogenic damage to the urethra during rectal mobilization). Our data on the extreme rarity of H-forms of fistulas in boys correspond to the literature.

Rectoscrotal Fistula (RSF)

Rectourethral fistulas that open along the midline suture of the scrotum (Fig. 2) are rare forms of ARM. (Endo M. et al. 1999). We observed 6 (6.5%) boys out of 93 with perineal fistulas out of 59 (63.4%) boys and 34 (36.6%) girls. As with recto-perineal fistulas, the opening of the rectosochonal fistula was narrowed. The degree of constriction was determined with Gegar's dilators. The diameter of the fistula corresponded to a narrow fistula - in 5 (80.3%) patients, a fistula of medium latitude - in 1 (19.7%) patients. A wide fistula was not observed among these patients. The diagnostic complex included ascending fistuloirrigography and studies aimed at identifying associated anomalies of other organs and systems.

Pouch colon was found in 9 (1.8%) of 504 patients with ARM and in 7.4% among patients with rare regional variants of the anomaly. There were 6 boys (66.7%), and 3 girls (33.3%). Regardless of gender, the newborns were operated on with a diagnosis of "non-varicose form of ARM" with symptoms of low intestinal obstruction in 1-2 days of life; the operation was completed with the imposition of a sigmastoma. The reason for the progressive low intestinal obstruction in boys was an excessive accumulation of intestinal contents in the rectal sac due to narrow fistulous passages that form a communication with the bladder. Out of 3 girls, 2 had a cystic enlarged section of the colon communicated with the vagina, and 1 with the bladder. A characteristic radiological sign of pouch colon in rectal atresia is the presence of gas in the bladder cavity. Due to the rarity of pouch colon in 2 patients, this indicated X-ray sign remained unnoticed. In all cases, the final diagnosis was established at the subsequent stages of diagnosis and surgical correction at the age of: -1 month - 2; -2 months-2; 3 months-2; 6 months -3. In the course of elimination of the sigmastoma, a saccular enlargement of the shortened colon filled with intestinal contents was found with thickened walls without haustrats and fatty suspensions, with hypertrophy of the mucous membrane. In the absence of a transition zone between the normal section of the intestine and abundant blood vessels of cystic expansion up to 12 cm, the fistula flowed into the bladder or vagina

Our patients corresponded to the complete type of lesion (4) - the absence or insufficient for bringing down the length of the colon, requiring coloplasty from the rectal sac: a) type I - complete lesion of the colon - 4; b) Type II - complete defeat of the large intestine with preservation of the ileocecal flap -3. 2 patients corresponded to an incomplete type of lesion (the length of the colon proximal to the dilated sac, sufficient for descent without coloplasty: type III - subtotal involvement of the proximal and transverse parts of the colon (1); type IV - with cystic enlargement of the left parts of the colon (1).

In conclusion, it can be noted that the variety of ARM consists not only of simple single anomalies, but also complex anatomical variants; manifests itself in a different frequency of individual forms depending

on the sex of the patients, which complicates the diagnosis and treatment. A significant predominance of fistulous forms in the reproductive system in girls (51 versus 7), correlates with the frequency of rectourinary fistulas in boys with the main clinical forms of ARM (60 versus 0). This circumstance in the literature is recognized as the fact that "congenital perineal fistulas can be defined as the" male version "of the rectovascular fistula, then the rectourethral and vesical can be attributed to the male version of the rectovaginal fistula" confirm our observations. Rare regional forms accounted for 24.1% in the general structure of ARM in children with fluctuations from 0.83% to 24% among various nosological forms. This indicates that our data correspond to intermediate indicators between the regions, where the indicated forms of ARM are observed often or rarely. To diagnose rare regional forms, an integrated approach is required with the identification of associated anomalies. To improve the accuracy and specificity of the verification of individual nosological forms, it is necessary to use special research methods. In our patients with rare regional variants of ARM, surgical tactics and methods of surgical treatment correspond to the principles of the Crikenbeck Consensus.

References

[1] Endo M, Hayashi A, Ishihana M, et al (1999) Analysis of 1992 patients with anorectal malformations over the past 2 decades in Japan. J PediatrSurg 34:435-441

[2] Holschneider A., Hutson J., Pena A., Bekhit E., Chatterjee S., Coran A. et al. Preliminary report on the International Conference for the Development of Standards for the Treatment of Anorectal Malformations// Journal of Pediatric Surgery. – (USA), 2005;(40)5:1521-1526.

[3] Holschneider A., Hutson J. Anorectal Malformations in Children. Embryology, diagnosis, surgical treatment, follow-up. - Heidelberg: Springer, 2006:251.

[4] I.de BLaauw, Wijers C.H.W., Schnriedeke E., Holland-Cunz S., Gamba P., Marcelis C.L.M., Reutter H., Aminoff D., Schipper M., Schwarzer N., Grasshoff-Derr S., Midrio P., Jenetzky E., van Rooij I.A.L.M. First results of a European multi-center registry of patients with anorectal malformations// Journal of Pediatric Surgery. – (USA), 2013;(48):2530-2535.

[5] Lawal T.A., Chatoorgoon K., Bischoff A., Pena A., Levvit M.A. Manegment of H-type rectovestibular and rectovaginal fistulas. J Pediatr Surg. 2011; 46(6):1226-1230.

[6] Levitt M.A., Pena A. Вкн.: Ashcraft's Pediatric Surgery 6th Edition. George W. Holcomb III. 2014:492.

[7] Pena A, Levitt M: Anorectalmalformations . In Pediatric Surgery and Urology: Long term outcomes 2nd edition. Edited by: Stringer M,Oldham K, Mouriquand PDE. //Cambridge: Cambridge University Press; 2006: p. 401-415

Citation: **Otamuradov F.A.** *Diagnostics and Treatment of Fistula Forms of Rare Regional Variants of Anorectal Malformations in Children. Archives of Gastroenterology and Hepatology. 2021; 4(1): 01-04. DOI: https://doi.org/10.22259/2639-1813.0401001*