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10-YEARS OF EXPERIENCE IN SURGICAL CORRECTION OF CHILDREN WITH ANORECTAL MALFORMATIONS IN THE REPUBLIC OF BELARUS

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Summary. Anorectal malformations (ARM) are a significant problem in pediatric surgery due to their prevalence (1:1500–1:5000 newborns), complexity of diagnosis, treatment, and high risk of postoperative complications, which significantly affect the quality of life of children.

Objective. to systematize and analyze the experience of diagnostics and surgical treatment of children with anorectal malformations in the Republic of Belarus over the past 10 years.

Materials and methods. From 2015 to 2024, 220 children with various forms of anorectal malformations were treated at the Republican Scientific and Practical Center for Pediatric Surgery (Minsk, Belarus). The age of patients ranged from 0 to 13 years, primary proctoplasty was performed at the age from the first day of life to 6 months.

Results. Primary proctoplasty was performed in 53 patients with a low-type malformation, accounting for 24.1%. A total of 167 (75.9%) children underwent a three-stage surgical approach, including an anorectoplasty using A. Pena's technique. In 7 patients with high-type atresia, mobilization and fistula closure were performed via an abdominal approach, while in 2 cases, video-assisted separation and bowel descent were utilized. Four girls with persistent cloaca underwent posterior sagittal anorectourethrovaginoplasty.

There were no intraoperative complications. Among patients who underwent a three-stage surgical approach, complications were observed in 12.5% (21/163) of cases, with 4 instances related to stoma formation and subsequent surgeries for colonic anastomosis. In 9 out of these 21 cases, repeat proctoplasty was required due to the development of anal stenosis (4/9), secondary ectopia (3/9), bowel retraction (1/9), and a pararectal fistula (1/9). Among children who underwent a single-stage procedure, complications were observed in 7.5% (4/53) of cases, consisting of wound dehiscence.

Conclusion. The choice between primary and three-stage correction depends on the type of ARM, the presence of associated anomalies, and the patient's age. The developed approach (primary PSARP for low-type cases without associated anomalies, three-stage correction for all other cases) has proven effective but requires adaptation in regions with delayed diagnosis. For children with low-type ARM, single-stage posterior sagittal proctoplasty is the preferred procedure, as it reduces treatment and recovery duration, eliminates the need for a colostomy, and minimizes related complications and financial costs.

Keywords. *Anorectal malformations, posterior sagittal proctoplasty, colostomy.*

Anorectal malformations (ARMs) represent a significant challenge in pediatric surgery due to their prevalence (1:1500–1:5000 newborns) [1], difficulty in diagnosis and treatment, and high risk of postoperative complications, which significantly impact children's quality of life. Modern advances in surgical correction, including posterior sagittal anorectoplasty (PSARP) [2] and minimally invasive laparoscopic techniques [3], have improved outcomes, but challenges in choosing the optimal timing of surgery, minimizing complications (fecal incontinence, chronic constipation, anal stenosis) [1, 4], and ensuring long-term functional and social adaptation of patients remain.

APRs are often accompanied by associated anomalies, such as cardiac, urinary, spinal, and limb defects, which occur in 40–70% of cases [5]. This requires a multidisciplinary approach that includes pediatric surgeons, urologists, cardiologists, geneticists, and rehabilitation specialists. Prenatal diagnosis using ultrasound and MRI increases the likelihood of early detection of APRs [6], but the limited availability of these methods in some regions and low diagnostic accuracy (less than 50%) complicate treatment planning.

The relevance of this topic is enhanced by social and economic aspects: treatment of APD is associated with significant costs for surgical interventions, long-term monitoring, rehabilitation, and psychological support for families [7]. The increase in the incidence of APD in some regions may be due to environmental factors, genetic mutations, or improved diagnostics [8]. Research emphasizes the importance of genetic counseling and the study of the molecular mechanisms of APD for the development of targeted therapies [9]. Personalized protocols that take into account the type of de-

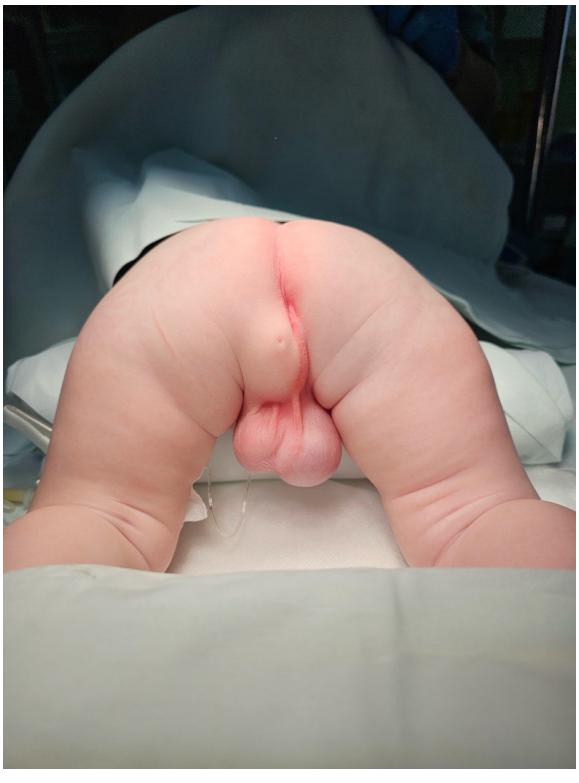

Figure 1. Rectal atresia in a boy with Currarino syndrome

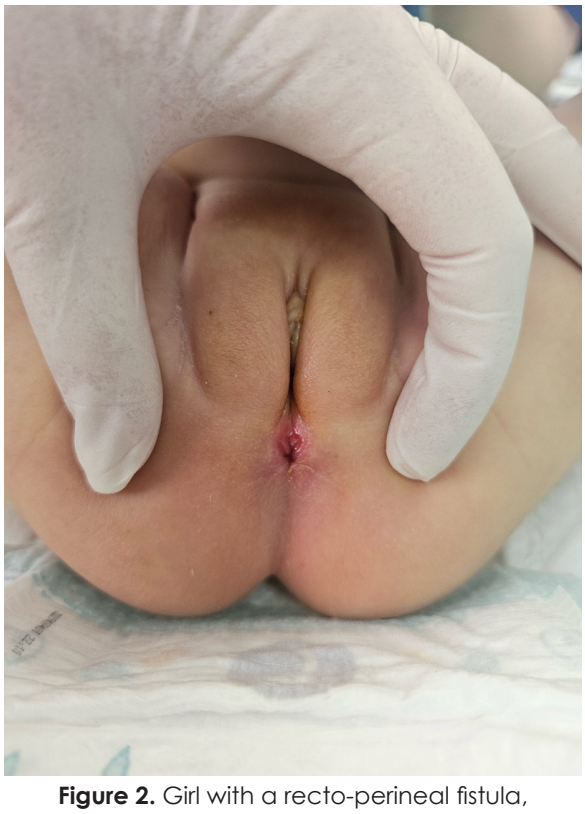

Figure 2. Girl with a recto-perineal fistula, diagnosed at 3 months of age

fect, associated anomalies, and genetic profile, as well as long-term rehabilitation programs aimed at restoring bowel and urinary system function, remain priority areas [2, 5]. Thus, improving diagnostic, surgical, and rehabilitation strategies for children with APD is a key task in modern medicine.

Purpose of the study. To systematize and analyze the experience of diagnosis and surgical treatment of children with anorectal malformations in the Republic of Belarus over the past 10 years, to highlight the main achievements, problems and prospects of surgical care for this category of patients.

Materials and methods. The study included data on 220 children with various forms of AP who underwent surgical treatment at the Republican Scientific and Practical Center for Pediatric Surgery (Minsk, Belarus) from 2015 to 2024. Of these, 114 (52%) were boys and 106 (48%) were girls. The pa-

Table 1. Characteristics of operated patients

	Primary PSARP	3-stage treatment
Total	53 / (100)	167 / (100)
Girls	37 / (69.8)	69 / (41.3)
Boys	16 / (30.2)	98 / (58.7)
Rectovestibular fistula	2 / (3.8)	34 / (20.3)
Rectoperineal fistula	47 / (88.7)	61 / (36.5)
Rectourethral fistula	–	38 / (22.7)
Rectovesical fistula	–	4 / (2.4)
Atresia without fistula	2 / (3.8)	19 / (11.4)
Anal stenosis	1 / (1.9)	5 / (3.0)
Cloaca	–	4 / (2.4)
Rare forms	–	2 / (1.2)

Note: Here and in Table 2, percentages are in brackets.

tients' ages ranged from 0 to 13 years; primary proctoplasty was performed at the age from the first day of life to 6 months. Forty-one children (or 18.6%) had severe concomitant malformations (VACTER association, congenital heart disease, Down syndrome, Currarino syndrome, etc.) (Figure 1).

The diagnosis was established on the basis of clinical examination, antenatal diagnosis, radiography, ultrasound examination (US) and, if necessary, magnetic resonance imaging (MRI).

Classification of the defect type was performed retrospectively based on patient medical data using the Crickenbeck classification, which provides a unified approach to assessing and categorizing these anomalies. Patient data are presented in Table 1.

Results and discussion: Prenatal ultrasound can help suspect AP if polyhydramnios, lack of gastric visualization, or associated defects are detected, but diagnostic accuracy remains low (less than 50%) [10]. According to our data, only 15% of fetuses were suspected of having rectal atresia based on ultrasound imaging—signs of low intestinal obstruction in the second and third trimesters of pregnancy.

Given the low prevalence of prenatal diagnosis, timely detection of ARP is the responsibility of the neonatologist and should be performed within the first day of life during a perineal examination in the delivery room. The presence or absence of the anus in its proper location, the prominence of the anal pit, its hyperpigmentation, the presence of radial folds, and the development of the gluteal muscles are assessed. In boys, the median perineal raphe, scrotum, and the location of the meatus are examined in detail. In girls, attention should be paid to the anatomy of the external genitalia: small labia minora and the inability to locate the vaginal opening may indicate the presence of a cloaca. Normally, in a full-term newborn, the anus is located in the middle of the line connecting the ischial tuberosities, and a 14 FR Foley catheter is passed through.

However, even in this case, diagnostic errors are not uncommon today, as APRs include more than 30 types of anomalies, making accurate classification difficult without additional research [1]. According to our data, APRs are diagnosed in the first few days of life in only 92% of cases. Diagnostic errors are especially common for low-grade defects, where anatomical changes are minimal (Figure 2).

In our practice, 12 children with rectoperineal fistula were diagnosed after reaching one year of age. In cases of anal atresia without fistula, a thorough examination of the newborn usually helps avoid diagnostic errors. However, in four cases, complete atresia was not detected until 2–3 days of life. The key factor in such situations is the presence or absence of meconium passage.

It should also be remembered that rectal atresia can be part of a complex of multiple malformations, and the absence of meconium passage in the first day of a newborn's life may be associated with the presence of a concomitant gastrointestinal defect. According to international literature, concomitant malformations are detected in 40–80% of children with rectal atresia [8]. According to our data, concomitant defects were noted in 18.6% of cases at the time of radical correction. Therefore, a perineal examination and a catheter-based test for anal patency are mandatory in all newborns within the first hours of life [2], and additional examination in questionable cases using an electrical stimulator to determine the clear boundaries of the external sphincter.

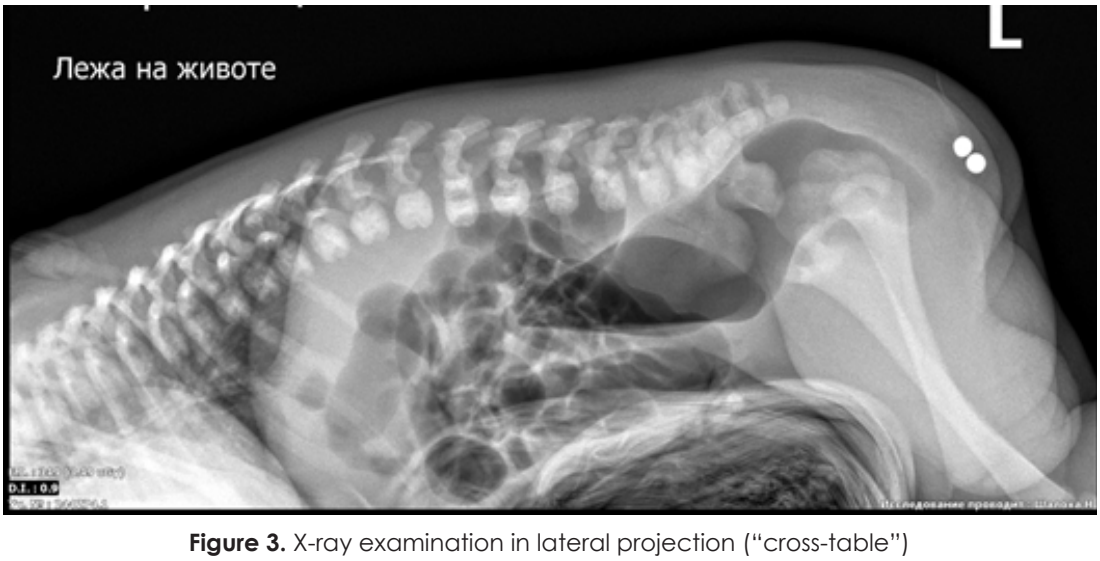

Figure 3. X-ray examination in lateral projection ("cross-table")

One of the priority tasks at this stage is organizing the timely and safe transfer of the newborn to the surgical hospital. We recommend transporting the newborn from the maternity hospital no earlier than 10–12 hours after birth to minimize the risk of deterioration in the baby's condition during the early adaptation period. The transfer should be performed exclusively by a specialized transport team. In perinatal centers, earlier transfer is possible.

If APR is confirmed in newborns, vital signs are monitored, clinical and laboratory tests are performed, and an ultrasound examination of the abdomen, heart, and brain is performed to rule out concomitant pathologies or genetic syndromes. In cases of multiple congenital anomalies, a customized treatment plan may be developed for each individual patient. Until the type of APR is determined and a surgical strategy is chosen, the child receives parenteral nutrition.

In case of anal atresia without a fistula on the perineum, it is recommended to perform an X-ray examination in the lateral projection ("cross-table") no earlier than 18–24 hours after birth (Figure 3).

Ultrasound diagnostics is currently considered a safer method. The first ultrasound, like radiography, should be performed by the end of the first day of a newborn's life. It allows for accurate measurement of the distance from the blind end of the rectum to the perineal skin and the detection of fistulas, such as rectoperineal or rectovaginal [6].

It should be noted that the accuracy of ultrasound examination results is largely determined by the level of training and experience of the specialist performing the procedure, as well as the quality of the equipment used [6]. To date, our center has performed perineal ultrasound in preoperative examinations on only 7 patients.

Children admitted to the Republican Scientific and Practical Center for Pediatric Surgery over the age of three months with a rectoperineal fistula underwent mandatory fluoroscopy through the fistula opening to rule out secondary megacolon and determine the surgical approach. Secondary megacolon was detected in 7 of 15 patients during barium enema, ruling out the possibility of primary anorectoplasty.

Despite the development of surgical technologies, a number of authors continue to consider the classical three-stage method of correction of ARP as preferable, using it regardless of the type of defect, starting with the formation of a preventive colostomy at the first stage.

In our clinic, we performed primary proctoplasty, posterior sagittal and abdominoperineal proctoplasty with the imposition of a preventive colostomy, depending on the anatomical variant of the defect.

Based on accumulated experience and analysis of world literature, we perform primary posterior sagittal anorectoplasty using the A. Peña technique without forming a protective sigmoid colostomy in children with rectoperineal fistula, rectovestibular fistula, anal stenosis, or anal atresia without fistula, provided that the distance from the blind end of the rectum to the perineum is less than 10 mm, and there are no concomitant malformations or complications associated with APR.

A. Peña's posterior sagittal anorectoplasty technique provides precise visualization of the perineal anatomical structures, allowing dissection to be performed strictly in the sagittal plane. This approach minimizes trauma to the rectal nerve plexuses and pelvic structures. Performing radical surgery in the neonatal period prevents the development of secondary changes, such as megacolon, and promotes optimal tissue healing, restoration of neural connections, and development of sensation and function. This ultimately improves the child's social adaptation. Primary anorectoplasty was performed on 51 patients.

Particular attention should be paid to children with rectovestibular fistulas. With this malformation, the rectum is intimately adjacent to the posterior vaginal wall, requiring a more extensive surgical ap-

Table 2. Types of surgical correction

Type of operation	Boys	Girls	Total
Primary PSARP	16 (14)	37 (34.9)	53 (24.1)
PSARP	89 (78)	65 (61.3)	154 (70)
Video-assisted colon pulldown (VAARPT)	9 (8)	–	9 (4.1)
Posterior sagittal anorectourethrovaginoplasty (PSARVUP)	–	4 (3.8)	4 (1.8)
Total	114 (100)	106 (100)	220 (100)

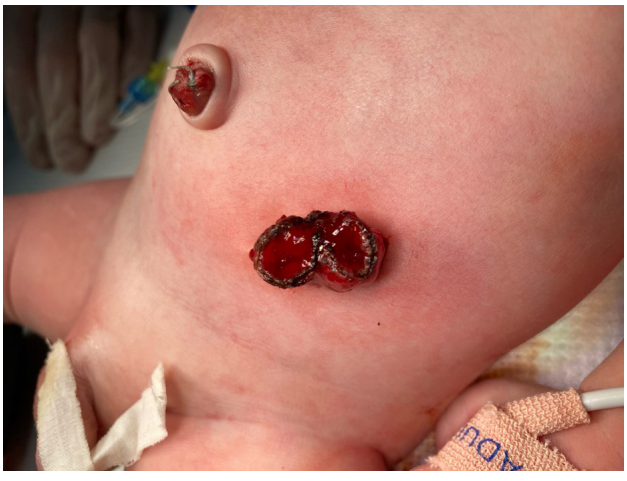

Figure 4. Double sigmoid colostomy in a newborn boy with a high form of APR

proach and greater bowel mobilization, which in turn leads to a higher risk of intra- and postoperative complications during primary anorectoplasty.

Eleven patients with low-grade colostomies and no associated pathology underwent three-stage surgery. We believe this is due to late diagnosis (7 patients) and poor awareness among regional surgeons (4 patients had undergone colostomies at other medical institutions). In all other cases, we begin correction with a three-stage preventive colostomy.

Previously, with APR, a double-ended, separate sigmoid colostomy was considered preferable to prevent intestinal contents from refluxing into the distal stoma. However, based on our experience, we believe a double sigmoid colostomy with an outlet in the left mesogastrium is the optimal option (Figure 4).

This approach ensures convenient care, ease of use of the ostomy bag, ease of subsequent stoma closure, and a better cosmetic outcome. We have not recorded any cases of intestinal contents refluxing into the distal stoma. Exceptions include girls with a persistent cloaca; in these cases, we perform a transverse colostomy (Figure 5).

In cases of anal atresia without a perineal fistula, distal colostomy is mandatory for fistula diagnosis and verification. It is important to remember to create sufficient contrast pressure to ensure its passage through the fistula and obtain a reliable anatomical structure, which allows for predicting the surgical course and choosing the optimal intervention, such as laparoscopic mobilization for rectovesical fistulas.

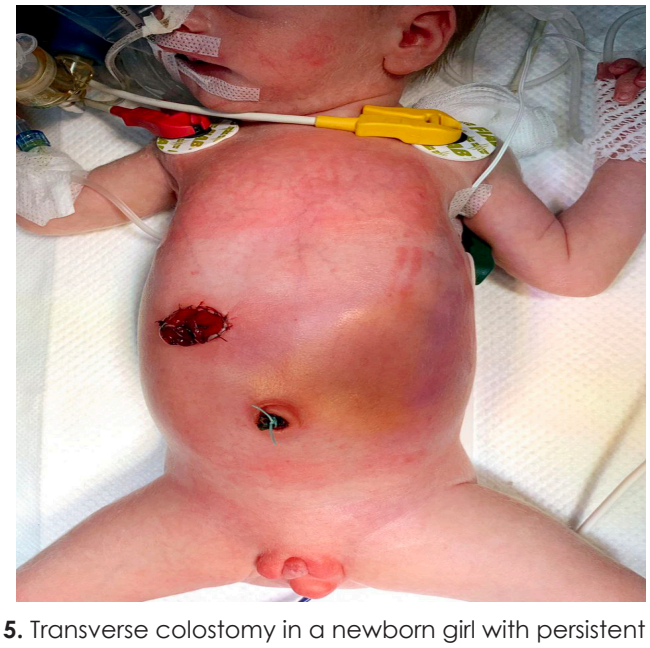

Figure 5. Transverse colostomy in a newborn girl with persistent cloaca

Figure 6. Surgical intervention tactics for isolated defects

The second stage involved anorectoplasty, also using the A. Pena technique. In seven patients with high-grade AP, fistula mobilization and closure were performed through an abdominal approach, and in two cases, video-assisted bowel dissection and lowering was used. In four girls with persistent cloaca, posterior sagittal anorectourethrovaginoplasty was performed.

The breakdown of surgical interventions is presented in Table 2.

In the postoperative period, all patients underwent bladder catheterization for 5 days, and 2 weeks after anorectoplasty, mandatory daily anal bougienage began until the age-appropriate bougie size was reached.

Based on our experience and literature review, we developed a strategy for treating children with APR without severe developmental defects (Fig. 6). In the presence of concomitant defects, we perform correction in three stages, regardless of the type of APR.

Complications. There were no intraoperative complications. In patients undergoing three-stage surgery, complications were observed in 12.5% (21/163), with 4 cases related to stoma placement and subsequent colonic anastomosis procedures. Nine of the 21 cases required repeat proctoplasty due to anal stenosis (4/9), secondary ectopia (3/9), bowel retraction (1/9), and perirectal fistula (1/9).

In children undergoing single-stage surgery, complications occurred in 7.5% (4/53) of cases and included wound dehiscence, two of which had a concomitant pathology, such as congenital heart defects. Although wound healing occurred by secondary intention after sigmoid costomy, a satisfactory cosmetic and functional outcome was achieved.

Follow-up examinations of patients were conducted at 3, 6, and 12 months after surgery to evaluate the functional and cosmetic results. Forty-seven patients who underwent single-stage surgery had no complaints from their parents, which accounted for 88% of this group. In the second group, 73.6% (123 patients) of parents had virtually no complaints and were completely satisfied with the treatment outcome: the frequency of bowel movements in the operated children corresponded to the age norm, and there was no fecal soiling. Thirty-six (21.1%) of all operated patients complained of constipation and the need for cleansing enemas. Fecal soiling was observed in six children who underwent single-stage surgery and in 15 who underwent three-stage surgery. In the second group, severe fecal soiling with elements of encopresis was observed in children operated on for high rectal atresia and persistent cloaca. When examining the perineum and stimulating the external sphincter, in 99% of cases the exact location of the rectum in the center of the sphincter apparatus was noted.

In our opinion, the cosmetic outcome depends on the surgical correction method and is assessed based on the appearance of the perineum, the presence of scars, the location and shape of the anus, and the condition of the surrounding skin. Primary anorectoplasty achieved the best cosmetic outcome (achieved in 92.5%), while the three-stage technique yielded good results in the perineum in 76% of cases, but the overall cosmetic outcome was compromised by scarring on the anterior abdominal wall.

Since primary proctoplasty is performed in a single stage without a colostomy, usually in the neonatal period and with low forms of APR, the anus looks anatomically correct without significant scarring, which is associated with better tissue restoration due to the high regenerative capacity of infant skin and an anatomically correctly formed perineum, characteristic of low forms of the defect. In turn, the absence of a preventive stoma increases the risk of postoperative complications in the form of suppuration of the postoperative wound, which can lead to secondary healing and the formation of more pronounced scars. A study by Bischoff A et al. (2017) showed that in 80% of patients with high APR, operated on in three stages, the anus looked aesthetically pleasing after PSARP, but scars from colostomy closure reduced parental satisfaction in 20% of cases [2].

Conclusion:

1. Treatment of children with anorectal malformations requires a multidisciplinary approach involving specialists, including neonatologists, pediatricians, geneticists, and pediatric surgeons. The importance of accurate diagnosis is emphasized in clinical guidelines and medical literature, as it determines the choice of the correct treatment strategy and the prevention of intra- and postoperative complications.

2. Low accuracy of prenatal diagnosis (15%) and diagnostic errors in low forms of defects (for example, rectoperineal fistula) highlight the need to improve the qualifications of neonatologists and the availability of MRI/ultrasound.

3. The choice between primary and three-stage correction depends on the type of PSARP, the presence of associated defects, and the patient's age. The developed strategy (primary PSARP for low-grade forms without associated anomalies, three-stage for other cases) has proven effective but requires adaptation in regions with late diagnosis.

4. For children with low forms of APR, one-stage posterior sagittal proctoplasty is preferred because it reduces the duration of treatment and recovery, eliminates the need for a colostomy, and minimizes associated complications and financial costs.

5. Formation of a colostomy is a very important and responsible operation in relation to possible complications and all subsequent stages of surgical correction of anorectal malformations.

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