

X-ray examination of the anorectum.

The state of the problem in pediatric colorectal surgery.

Normally, the rectum is in the pelvis retroperitoneally. It starts from the 3rd sacral vertebrae and ends at the level of the pubococcygeal line, where it borders on the anal canal. From a functional point of view, the rectum begins caudally to the rectosigmoid sphincter. While fecal retention, it performs a cumulative function, and during bowel movements, its strong peristaltic wave, which starts from the rectosigmoid sphincter, expels the stool through the open anal canal [1,2].

The anal canal is in the tissues of the pelvic floor. It is represented by the internal anal sphincter (IAS), which anatomically is a thickened continuation of the circular layer of the rectum, but functionally it fundamentally differs from the rectum, as it is in constant contraction. Unlike the rectum, it does not have intermuscular ganglia and does not peristaltic [3]. Around him are the muscles that, together with IAS, are involved in fecal retention, including the puborectalis muscle (PRM) and the external anal sphincter (EAS), as well as the levator ani muscle (LAM), the contraction of which during defecation causes a wide opening of the anal canal, which significantly facilitates the process of evacuation of feces. **Table 1** shows the normal width of the rectum and length of the anal canal in patients of different ages without anorectal pathology after filling the colon with barium at least to the splenic angle [1,4].

In 1953, Stephens proposed the concept of a pubococcygeal (P-C) line, which runs from the lower part of the pubic bone to the distal coccygeal vertebra. He showed that this line corresponds to the location of the PRM. It is located between the rectum and the anal canal. If in ARM patients the blind end of the intestine is located above this line, these cases are considered a high type of ARM,

if at the level of this line it is an intermediate type, and low type if the intestine is located more caudally than this line [5].

Table 1. The normal size of the rectum and anal canal in different ages.

| Ages | The width of the rectum (cm) | The length of the anal canal (cm) |
|--------------------|------------------------------|-----------------------------------|
| 5 days – 11 months | 1.3 – 3.0 (2.24±0.09) | 1.7 – 2.5 (2.21±0.15) |
| 1 – 3 years | 3.0 – 3.7 (3.21±0.11) | 2.3 – 2.8 (2.55±0.10) |
| 4 – 7 years | 3.0 – 3.9 (3.43±0.14) | 2.5– 3.6 (3.17±0.14) |
| 8 – 10 years | 3.2 – 4.1 (3.72±0.05) | 2.6 – 3.7 (3.11±0.10) |
| 11 – 15 years | 3.6 – 4.6 (3.95±0.07) | 3.1 – 3.9 (3.43±0.10) |
| 23 – 64 years | 3.5 – 4.8 (3.95±0.21) | 3.4 – 4.2 (4.08±0.07) |

Since then, it was believed that if the gut is located below the P-C line, it means the patient has an anal canal that needs to be preserved during surgery. "The results in low deformities operated by perineal rectoplasty showed "good" clinical scores, good preservation of the rectoanal reflex, and good electrical activity of the external sphincter [6]. Low type anomalies were treated by neonatal perineoplasty, anal transplants or cut-back. In patients with the vestibular ectopy "The sling of the puborectalis muscle was well identified by causing contraction of the puborectalis muscle with an electric stimulator". Patients with low type had good results in 100% [7].

In that period, an invertograms was doing for the diagnosis of ARMs type, i.e. lateral radiograph of the pelvis with the child's position upside down. It was assumed that in such cases the gas will rise to the highest possible level. It was not considered that the contraction of the anal canal prevents the penetration of gas into the anal canal. Only in the later dates, when high pressure was created in the rectum, was it possible to fix the opening of the anal canal and the penetration

of gas to the mark in the region of the anal dimple. However, surgeons evaluated the level of the defect at an earlier time, fearing bowel perforation. Because of this, most anomalies were erroneously rated as a high or intermediate type.

Manometric studies in all patients with an ARM with visible fistulas revealed normal basal anal pressure and a positive rectoanal inhibitory reflex (RAIR) in the terminal section of the intestine, which indicates the presence of a functioning anal canal, i.e. about the low type of ARM [8,9]. It should be emphasized that numerous articles by pediatric surgeons about the alleged presence of RAIR in patients after the removal of IAS mistakenly evaluated peristaltic wave (the Bayliss-Starling gut law) as RAIR. The decrease in pressure in the caudal segment of the intestine after stretching the wall in the cranial segment is mediated by the intramural nervous system and is not related to the reflex arc. The rectoanal inhibitory reflex is called inhibitory because, in response to the extension of the rectal wall, IAS relax, which through the reflex arc leads to a contraction of EAS and PRM, which inhibit (prevent) involuntary defecation.

X-ray studies of anorectum in patients with visible fistulas are presented in **Figure 1**. They allow the following conclusions.

First, the injection of contrast medium into the rectum at rest reveals a narrow channel located caudal to the pubococcygeal line equal to the length of the normal anal canal (**Figure 1.c**). This channel ensures the preservation of the contrast medium in the rectum, i.e. provides normal stool retention. This canal is not a fistula since during bowel movements it opens to the width of the rectum, like a normal anal canal (Figure 1. d).

Secondly, in most cases (except for Figure 1.a) there is convincing evidence of stenosis of the fistulous opening, which led to the appearance of megarectum. In Figure 1.c, severe stenosis led to the development of proctitis.

Thirdly, the distance between the wall of the opened anal canal and the mark near the anal dimple ranges from 2 to 5 mm, depending on the thickness of the skin and subcutaneous tissue, i.e. from the age of the patient.

Fourth, the anal dimple is located near the axis of the rectum at a distance of the length of the anal canal from the P-C line.

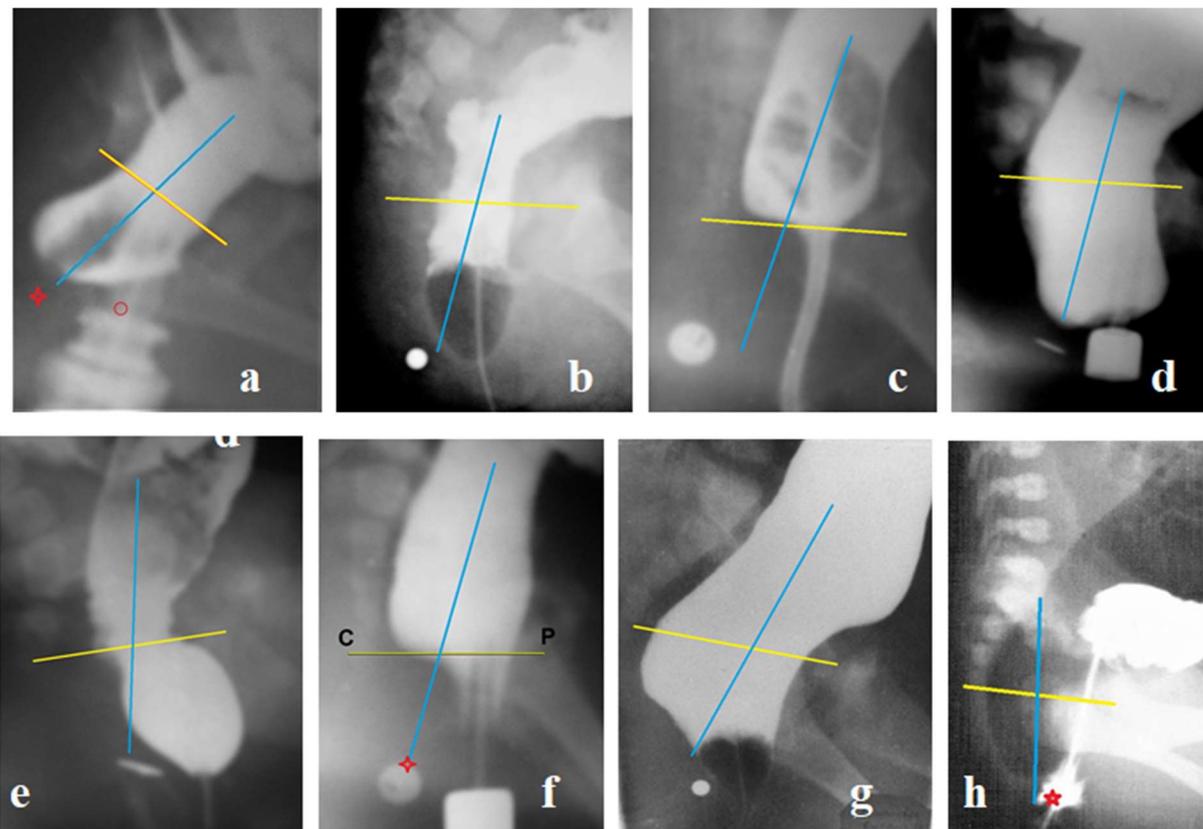


Figure 1. Lateral radiographs of anorectum in patients with an ARM with visible fistulas (a-g), and without a visible fistula in a newborn (h). The Pubococcygeal line (P-C) (yellow line) was drawn approximately, as there are no precise bone landmarks on radiographs. Therefore, it is impossible to accurately measure the length of the anal canal. The axis of the rectum is the blue line. Location anal dimple - red star (a) A barium enema in a 3-month-old baby with a vestibular fistula. The distance from the sprocket to the P-C is equal to the length of the anal canal. The distance from the anal dimple to the wall of the opened anal canal is ≈ 2 mm. (b) In a girl of 8 months with a vestibular fistula, after the introduction of barium, an uneven expansion of the retrorectal space is determined, which indicates proctitis. The balloon of the Foley catheter lingered over the rigid ring of the ectopic anus. The distance from the wall of the anal canal to the mark in

the anal dimple is ≈ 2 mm. (c-d) A patient with a vestibular fistula. (c) At the age of 9 months, a barium enema was performed through a tube inserted through the fistula. The anal canal contracted around the tube. (d) At the age of 1.5, during a barium enema, an attempt was made to defecate with the opening of the anal canal. The distance between the blind wall of the anal canal and the button in the anal dimple is 0.5 cm. (e) In a boy with a perineal fistula, after a barium enema, a Foley catheter was inserted into the rectum. A soluble contrast medium was introduced into its balloon and the catheter was pulled up to stop. Megarectum is determined. The distance between the wall of the anal canal and the button is ≈ 2 mm. (f) In a patient with a vestibular fistula and megarectum, barium penetrates in front and behind the tip of the enema, which indicates the weakness of the puborectalis muscle. (g) A patient with a perineal fistula has a pronounced megarectum. A balloon of Foley catheter by small diameter stuck above a narrow fistulous opening. (h) A newborn with an ARM without a visible fistula. The surgeon tried to inject the contrast medium into the rectum but introduced it near the intestine. The picture shows the opening of the anal canal. The distance from its wall to the injection site is ≈ 2 mm.

In ARM patients without a visible fistula 30 hours after birth, compression of the abdomen causes the opening of the anal canal if there is a low type of defect (Figure 2. C-D). In patients with a colostomy, the opening of the anal canal occurs under the influence of high pressure in the rectum. Augmented pressure colostogram (Figure 2. A-B) is an accurate but not safe diagnostic method. Compression of the abdomen after filling the rectum with contrast medium is no less accurate and completely safe since the opening of the anal canal is not a result of mechanical pressure, but a reflex reaction. As can be seen from Figure 2, the distance between the rectal fossa and the wall of the opened anal canal is the same 2-5 mm.

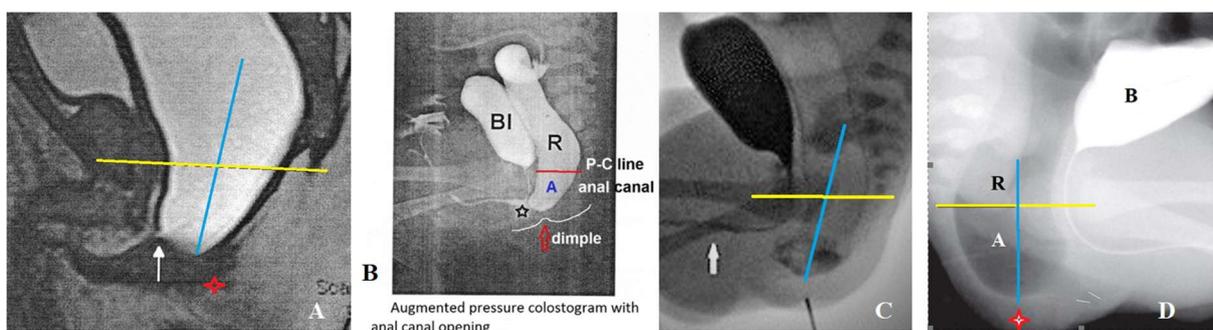


Figure 2. Lateral radiographs of anorectum in boys with ARM without visible fistula. (A-B) Augmented pressure colostogram in patients with urethral fistulas. (C-D) Radiographs of newborns with perineal fistula. (C) at the root of the scrotum: (D) A very narrow perineal fistula.

All radiographs with urethral fistulas have the same characteristics.

Firstly, the so-called fistula is very narrow and short (1 mm) communication between the anal canal and the urethra (**Figure 2.A-B**). What surgeons call fistula is the contracted IAS. During surgery under anesthesia, the rectal pressure is low, so IAS is in a contracted state, just as in healthy individuals.

Secondly, the distal wide intestine, and its anastomosis with urethra located caudal of the P-C line. This defect more correct to call as anal ectopy into the urethra.

Thirdly, the anal fossa is always located posterior to the fistula, near the axis of the rectum, and at 2-5 mm from the wall of the anal canal.

Fourth, the presence of a functioning anal canal does not depend on concomitant abnormalities, including in the spine.

Pathogenesis of ARM. The fistulous opening, wherever it is always, to one degree or another, is narrower than the normal anus. While the baby has loose stools, bowel movement is not impaired. However, when the stool becomes thick, the emptying of the rectum is disturbed, the stool accumulates in the rectum, gradually causing it to stretch – megarectum (**Figure 3.a**). If the stenosis is not eliminated, then there is a stretching and weakening of PRM, which is defined as descending perineum syndrome. This leads to fecal incontinence (**Figure 3.b-c**). Anorectum status after a pull-through operation is shown in a girl at the age of 2 years (**Figure 3.d**). The true diameter of the contrasting marker near the anus is 1.6 cm. Therefore, the width of the rectum is 6 cm (the maximum normal limit for this age is 3.7 cm). IAS was excised as a fistula. PRM was dissected. There is no contraction of the deep and superficial parts of the external anal sphincter. However, the injected contrast medium does not spill out of the intestine, which

is most likely due to a narrowing in the area of the subcutaneous part of the EAS. If the delay was due to a contraction in the sphincter, then it would have stopped 1 minute after the introduction of the barium. The presence of megarectum also indicates stenosis.

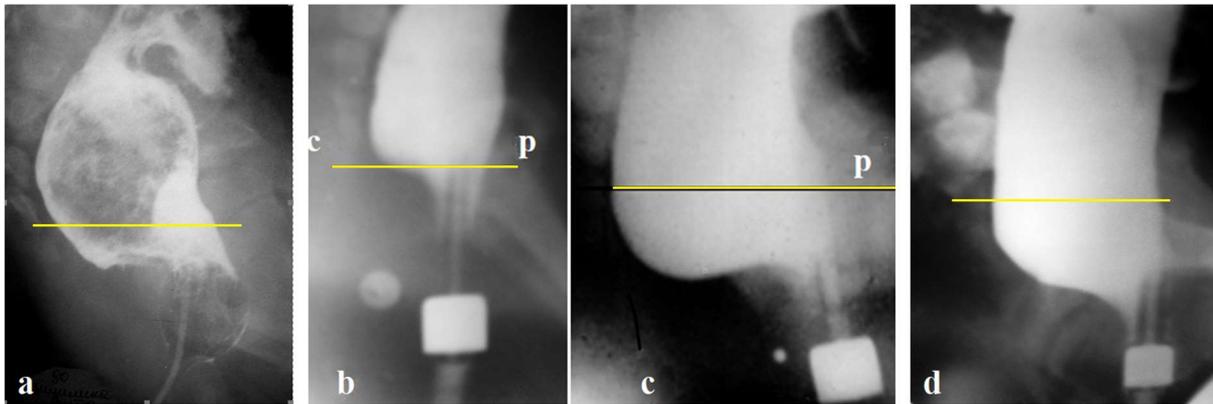


Figure 3. Anorectum studies in patients with ARM. **(a)** As a result of stenosis of the ectopic anus, feces accumulate in the rectum, causing megarectum. A strong peristaltic wave is trying to expel fecal stone, which bougie the muscles of the pelvic floor and stretches PRM (descending perineum syndrome). **(b-c)** Study of a girl with a vestibular fistula: **(b)** Under the age of one year, the penetration of barium into the anal canal (length -2.5 cm) is determined behind the tip of the enema, which indicates a weakness of PRM. The width of the rectum is 3.4 cm (maximum norm 3 cm); **(c)** In a year megarectum (5 cm) is determined (maximum norm 3.7 cm) and descending perineum syndrome. Due to PRM failure, the upper part of the anal canal is not involved in stool retention. **(d)** After the pull-through operation, the megarectum and stenosis of the neoanus are determined. There is no function of IAS, PRM, and EAS.

Thus, in both manometric and radiological examinations, a functioning anal canal was detected in all ARM patients with visible fistulas and in the vast majority ($\approx 90\%$) of boys with urethral fistulas [10,11].

In modern pediatric colorectal surgery, erroneous postulates are accepted on which the false understanding of the pathological physiology, diagnostic methods, and treatment of some colorectal disease are based.

1. As a preamble to all articles on the ARM, we can read: "In 1982 the first posterior sagittal exploration of a child with an anorectal malformation was performed, allowing the first direct observation of the intrinsic anatomy of these

defects. It was found that the “puborectalis muscle,” traditionally considered an important landmark, actually was not the “rectal sphincter” [10,12].

First, PRM is difficult to detect even with an anatomical study. With surgical dissection, it is not visible, but this is no reason to believe that it is not a sphincter. Obviously, the author of this invention tried to justify the security of the PRM intersection during the posterior sagittal approach.

Secondly, (outside of pediatric surgery), it is known that PRM plays an important role in stool retention [13]. As a result of this deception, children's surgeons have no idea about descending perineum syndrome. This syndrome shows pelvic floor muscle weakness, including PRM, in severe chronic constipation, which leads to fecal incontinence [14]. Such articles are not allowed for publication, as they contradict this false concept. On the same basis, there is no research in pediatric surgery about the role of paradoxical puborectalis contraction as the cause of functional constipation (FC). [15]. Ignoring this cause of FC, pediatric surgeons came to the false conclusion about the possibility of segmental expansion of the colon and produce a resection of the dolichosigma, leaving the expanded rectum and damaged anal canal.

2. With no evidence, there was stated that patients with ARM have no anal canal [16]. For the sake of avoiding doubts about the value of posterior sagittal access, ridiculous efforts are made to not use the concept of “anal canal”. The authors write: "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" and further, in the signature to the figure it is written: " Distal colostogram in a 2-month-old boy shows rectal sphincter that is closed ..." [10].

Firstly, there is no rectal sphincter in anatomy. Secondly, there are no muscles around the rectum. Thirdly, that part of the intestine, which is in constant contraction as a result of the function of the surrounding sphincters and opens

under the influence of high pressure in the rectum, as described in this article, is a functioning anal canal.

The questions arise:

(1) how could such errors, which contradict fundamental scientific knowledge, could be printed in a scientific journal and not provoke the indignation of scientists?

(2) If most patients with ARM have a functioning anal canal, why do children's surgeons destroy it? What will they say to their patient in the courthouse to the question: - Doctor, why did you destroy my anal canal?

See how augmented pressure colostogram analysis (Figure 4) looks like in the article Helleran et al [17].

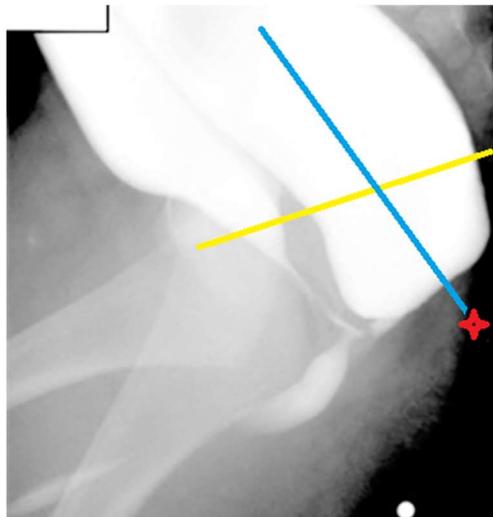


Figure 4. (Original signature). Distal colostogram of male born with an ARM. The pubococcygeal line may be useful in predicting the need for laparoscopy or laparotomy at the time of posterior sagittal anorectoplasty. If the rectum is the first structure encountered from a posterior sagittal approach, then the case can more likely be performed using only a posterior sagittal approach. If the urinary tract is the first structure that would be encountered via a posterior sagittal incision, than it is the more likely that laparoscopy or laparotomy likely be needed [18].

Signature shows that the surgeon's actions will depend solely on what he will see after the intersection of the puborectalis muscle. The role of the pubococcygeal line is completely perverted. Obviously, under the influence of high pressure, the anal canal, which is located caudal to the pubococcygeal line is open. The fraction is located far from the anal fossa (not behind, but in front of the fistula and far from the axis of the rectum). A red asterisk is a more likely site of the anal fossa. It is located a few (2-3) mm from the wall of the anal canal. It seems that the fraction was drawn on a radiograph to show the remoteness of the intestinal wall from the anal fossa. Conclusion: ectopy of the anal canal into the urethra, i.e. low type of the ARM. The IAS during PSARP will be in a closed state and will be removed as a fistula. The rectum torn from the levator plates will be lowered to place of the IAS. The nervous connection of the rectum with the perineal muscles will be destroyed, because of which a perineal fistula will be created instead of a functioning anal canal.

A psychological phenomenon is known that when people keep hearing the same statement multiple times, they start perceiving this statement to be true. The following are false statements that are not confirmed by any research, contradict well-known scientific facts and are nevertheless replicated as truth.

- a) Puborectalis muscle does not play an important role in stool retention,
- b) In patients with ARM, the anal canal is absent,
- c) The results of the posterior sagittal anorectoplasty are remarkable.

Since in most cases of the ARM patients have a functioning anal canal, which completely collapses during PSARP, an acceptable fecal retention and defecation function after this surgery is theoretically impossible. This is evidenced by the long-term results of this surgery. «A large proportion of the patients have persistent fecal incontinence, constipation and sexual problems that have a negative influence on their quality of life. The ARM-patients had an inferior

outcome ($P < 0.05$) for all incontinence parameters, 8 of 10 parameters for constipation, 2 of 6 for urogenital function and 7 of 13 quality of life parameters. Forty-four patients (32%) reported incontinence of stool at least once a week and 16 (12%) had a permanent colostomy" [19]. Keshtgar et al "...studied 62 children with intractable fecal incontinence after repair of anorectal malformation between January 1991 and January 2005. Sixteen (26%) children had excision of megarectum with median age of 9 years (range, 2-15 years) and postoperative follow-up of 5 years (range, 1-10 years). Patients with a poor IAS or neuropathy will often require artificial means of fecal continence" [20].

The literature research shows that children and young people with anorectal malformations have a worse quality of life than their healthy peers. They are ashamed, feel different than others and are afraid that they smell bad. They see themselves as unattractive and are often dissatisfied with their bodies. The multiple problems in the social, physical and psychological areas make it necessary for children and adolescents with anorectal malformations to get early support (bowel management program, psychologist, group therapy). For example, affected children may begin already with 3^{1/2} years with a bowel management program to prevent negative consequences and/or to avoid social exclusion. For parents, it is important that they learn to accept the child's illness and to help the children to cope with their illness better. With this support in childhood, it will be possible to accept the disease better and to integrate it into their self-image [21]. From childhood, these children were convinced that they were born without the anal canal and they were operated on in the best way so that the stool does not involuntarily fall out of the rectum. Can you imagine their quality of life when they find out (they will know) that they were born with a functioning anal canal?

This situation was possible due to the "divide and rule" policy. After numerous repetitions about the superiority of PSARP to the authors managed to take a

leadership position, which allowed them to "tear" children's colorectal surgery from the general scientific direction. Thus, they defended themselves from criticism of their anti-scientific insinuations.

Since then, the number of invented, unreasonable methods of treating colorectal problems has been multiplied.

Functional constipation.

1) The role of the anal canal in the pathogenesis of FC is ignored, as a result of which the idea arose of the possibility of segmental colonic dilation. The authors advertise a resection of the sigmoid colon, which, firstly, is not physiological, because the cause of the disease remains (affected anal canal and megarectum). Secondly, as Duhamel showed, this operation does not fix the problem [].

2) In 2005, the authors reported 17 patients with FC after a sigmoid resection. "Following sigmoid resection, 10% of patients did not require any more laxatives, have bowel movement every day and no soiling. Thirty percent of patients decreased the laxative requirement by 80%. The remaining 60% of patients decreased the laxative requirement by 40%" [22].

15 years have passed since then. The experiment with resection of the sigmoid colon continues, even though the long-term results of this method have not been studied have not been published. A preliminary report of a new approach using a laparoscopic sigmoid resection combined with a malone appendectomy was published in 2017 [23]. Six months later, an intermediate report was published. During this time, the number of patients increased by 25 with the following "intermediate" results: «Ten (32.3%) are clean with no flushes (1 takes no laxatives, 8 are on low dose laxatives only, and 1 patient was clean on laxatives but chose to switch back to flushes). Of the 21 patients that remain on antegrade flushes, 20 (95.2%) are clean, and one patient (4.8%) continues to soil [24].

Obviously, functional results after sigma resection are independent of approach. From the laparoscopic approach, a reduction in surgery time and a reduction in infectious complications can be expected. Therefore, justification by the authors of laparoscopic sigmoid resection seems completely no logical. “Previously reported (1) open sigmoid resection as a surgical option, but this did not sufficiently reduce the laxative need, then (2) a transanal approach (with resection rectosigmoid), but this led to a high rate of soiling...” [23]. The authors do not provide evidence that their assumptions were justified, and the need for laxatives significantly decreased. In order to evaluate the results shown, they would need to be compared with the results of those centers where children with FC are treated with exclusively conservative methods. It is noteworthy that the authors use methods that have no scientific justification and can be considered as unreasonable experiments.

Firstly, the use of high doses of Senna has no justification. It causes severe irritation of the colon, cramping, and abdominal pain. At a time when there are different laxatives and different methods of conservative treatment of FC (biofeedback, electrical stimulation, botulinum toxin injections, as well as a retrograde enema etc.), the appointment of high doses of Senna is an irresponsible mistake.

Secondly, if the patient cannot withstand the pain due to the high dose of Senna, or within a week of such treatment, there is no clinical effect, this should not be an indication for surgery. [25, 26]. In such cases, the logical solution is to change the laxative or the use of other methods of treatment.

Thirdly, as a result of unreasonable indications for surgery on the basis of the lack of effect from Senna's treatment, or a manometric study, or a false assessment of the width of the rectum, young children are operated at the initial stage of the disease and with a high probability of complete recovery without the ability to test the accepted methods of conservative treatment.

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An example of unfounded experimentation was published in 2005. The authors recommended a method for treating chronic constipation in children. "An alternative could be to resect the rectosigmoid including the rectum, down to the pectinate line in a similar manner as for patients with Hirschsprung's disease, and anastomose the non-dilated colon (that is assumed to have normal motility) to the rectum above the pectinate line" [22]. In 2009, the same authors published data on 15 patients who underwent a transanal rectosigmoid resection. They showed that this operation " ... resulted in a dramatic reduction or elimination in laxatives use while preserving continence. It is a useful alternative to surgical options such as other colonic resections, antegrade enemas, and stomas [27]. The pectinate line (dentate line) is a line that divides the upper two thirds and lower third of the anal canal. Consequently, the authors removed and recommended to other pediatric surgeons to remove most of the anal canal. No wonder "this led to a high rate of soiling" [23]. It should be noted that after removal of the anal canal, these people are doomed to incontinence of feces for life.

It is hard to believe that pediatric surgeons who do not know the anatomy of anorectum operate on children with severe illnesses in this area.

The same authors, not engaged in scientific research (anatomy, histology, manometry, and radiation diagnostics) once upon a whim, the concept of a rare until then cloacal malformation was sharply expanded. They offered surgical treatment, not having an idea about the internal and external urethral sphincters. When operating patients with a vestibular and vaginal fistula, they not only

destroy the anal canal, but also the urethral sphincters created by nature, and explain the poor results with the severity of the defect [28].

An analysis of the published articles of these authors did not reveal a single scientifically based result. It follows that all proposals for the diagnosis and treatment of ARM and FC are unreasonable experiments, which, as shown above, damage the function of the anal canal. The question is, how could pediatric surgeons who are not versed in the anatomy of anorectum dictate their experimental essays to the world community of pediatric surgeons?

This is the psychological strategy of victory:

a) You can lie (about the excellent results of the PSARP, about the absence of the anal canal, and the role of PRM, etc.) When people keep hearing the same statement multiple times, they start perceiving this statement to be true;

b) Divide and rule. Children's colorectal surgery has become their patrimony. They decide what to consider correct, even though this contradicts scientific evidence and even common sense.

c) He who is not with us is against us. People with charisma, who can provide attention and help, evoke reciprocal gratitude, which turns into dependence on the principle of political correctness. Through these people, winners control publications in scientific journals.

This state of affairs cannot continue indefinitely, firstly, because it has nothing to do with science, secondly, because patients should not suffer from unreasonable experiments.

Dear colleagues! For many years, you have believed the publications of these authors. However, faith and science are incompatible. I urge you to check all the provisions, which I presented in this analysis of the situation in pediatric colorectal surgery.

respectfully

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