Functional megacolon in children (etiology, pathogenesis, diagnosis). Review.

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Abstract The definition of "Megacolon" is still subjective since the methods of determination are not based on reliable limits of the norm. A radiometric method for determining the size of the colon, and anal canal length in children of different ages is described, which made it possible to establish that acquired megacolon is always of an obstructive nature. In about 5% of patients, the cause of constipation is congenital stenosis of the anus with a relatively wide diameter (secondary megacolon). In other cases, constipation is caused by dysfunction of the anal canal (functional megacolon - FM). FM occurs between 3 and 9 years of age, because of an involuntary delayed defecation, which leads to overflow of the rectum. A subsequent painful attempt at defecation causes a conscious delay. Thus, a vicious circle arises, because of which a functional megacolon (FM) develops. After 3-5 years, because of the bougie effect of the fecal stone, the pelvic floor muscles are stretched and weakened, which is manifested by a shortening of the anal canal. Measuring the length of the anal canal allows one to determine descending perineal syndrome during a routine barium enema. Due to the weakness of the puborectalis muscle encopresis occurs. Prompt treatment can lead to complete recovery. With third degree megacolon, treatment should be long-term and complex. Anorectal achalasia is also complicated by hypertrophy and sclerotic changes in the internal anal sphincter. In addition, because of stretching of the levator plates, during an attempt to defecate, the contraction of the stretched levator plates does not open the anal canal, which sharply increases the resistance to the movement of feces. In some patients, chronic obstruction causes an inflammatory process, which leads to an increase in the tone of the colon and a decrease in the diameter of the rectum. This leads to the normalization of bowel movements. It has been shown that megacolon is the key to understanding such pathophysiological phenomena as slow transit, changes in pressure in the colon, dyssynergic defecation, etc.

Keywords: Functional megacolon; chronic constipation; functional constipation; barium enema; anal canal; descending perineal syndrome; children.

Abbreviations

ARM – anorectal malformations.

C – constant.

FC – functional constipation.

FM – functional megacolon.

IAS – internal anal sphincter.

EAS – external anal sphincter.

PRM – puborectalis muscle.

RPR – rectal-pelvic ratio.

DPS – descending perineal syndrome

1. Introduction

The definition of "megacolon" suggests an increase in the size of the colon. To consider the colon enlarged, it is necessary to know its normal size. This is especially difficult, since it is obvious that all anatomical parameters depend on age and height. Currently, there are no generally accepted limits of the norm for the width of the colon and therefore the definition of megacolon is subjective. For example, Cuda et al considered megacolon if the width of the sigmoid colon is ≈10 cm [1]. Considering that the normal width of the sigmoid colon in adults is 2.2–2.6 (2.36±0.03 cm) [2], this means that the expansion of the intestine from 3 to 10 cm is not mistakenly diagnosed as megacolon. There is no consensus on the pathogenesis of megacolon and its association with chronic constipation. For example, Cuda et al consider constipation to be one of the symptoms of megacolon. It is natural to assume that megacolon results from the retention of feces in the colon. Thus, the functional constipation and megacolon are probably obligatory satellites. Different authors use different terms to describe the expansion of the colon: megarectum and megacolon. It is known from anatomy that the rectum is always wider than the sigmoid colon. Only in Hirschsprung's disease is the rectosigmoid ratio less than one [3]. With constipation of a different etiology, accompanied by severe chronic constipation, the rectosigmoid ratio may be less than one. But in such cases, the rectum is significantly expanded relative to the norm. This is since the rectum is in the small pelvis and its expansion is limited by the boundaries of the small pelvis, while the sigmoid

colon can expand much more, since it is not limited by anatomical boundaries [4]. There are no answers to the following questions: (1) why does the rectum expand? (2) can rectum expand without expanding the sigmoid colon? (3) in what cases can these two definitions be used?

2. Analysis of different methods for measuring of the colon and anorectum

2.1. Rectal-pelvic ratio (RPR) was the first attempt to make to objectively define megacolon (megarectum) [5, 6]. The RPR was obtained by dividing the diameter of the rectal width by the diameter of the linea transversa of the pelvis **(Figure 1.A).** The mean RPR in control group was 0.52 (range 0.31–0.61). This method was developed with serious methodological flaws. First, only 5 patients were selected as a control group. Secondly, they had a high incidence of painful defecation (50%) and straining (30%), which is typical for patients with constipation, which is always accompanied by dilatation of the rectum. Thirdly, barium X-ray in the control group was not performed. The diameter of the balloon, at which the child felt the need to defecate, cannot be equated with the normal rectal width since the width of the rectum in the time of the fecal retention less than at the time of the urge to defecate. Fourth, in patients with rectal fecal impaction, 5 ml of barium diluted in 40 ml of water was injected. In this way, the width of the fecal stone, but not the rectum, was determined. The mean RPR in patients was 0.68 (range 0.32–0.83). The mean intrarectal balloon width was obtained in 25 patients and five controls and the results (0.64 (range 0.37–0.75) and 0.52 (range 0.31–0.61)) were not significantly different. The authors suggested identifying to define megarectum by a RPR of 0.61 or larger. However, the proposed method revealed the expansion of the large intestine in severe forms of megacolon and therefore, with rare exceptions, was not used in the next 13 years [6,7]. Subsequent studies in adults have proven the effectiveness of X-ray examination to determine the megarectum [8,9]. The megarectum was determined based on a rectal diameter > 6.3 cm, regardless of the height and volume of the patient's pelvis [9]. This study did not take projection distortion into account. It is known that the further the object (rectum) is from the cassette, the larger its image on the radiograph. With a wide pelvis, the length of the image may be 2 times the true value **(Figure 1b).**

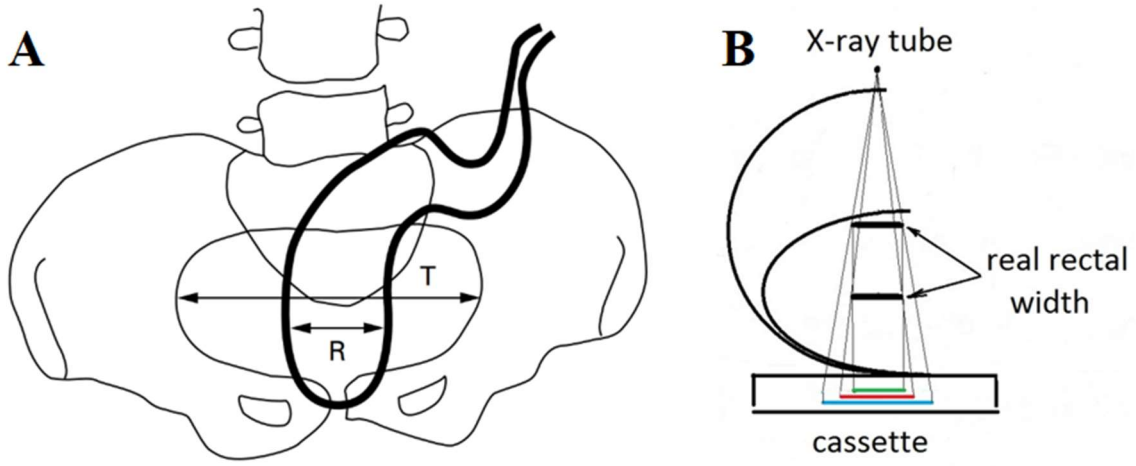

Figure 1. (A) Method for measuring the width of the rectum from van der Plas et al [6]. Explanations in the text. (**B**). The enlargement of the rectal image on the X-ray depending on the width of the pelvis (Scheme). Green line – the real rectal width; red line – its image in small pelvis; blue line – its image in large pelvis.

2.2. Koppen et al, made the following attempt to determine the border of the normal rectum [10]. To do this, they measured the width of the rectum on the frontal radiograph of children under the age of 6 years during desinvagination with pressurized air insufflation. They did not consider that the tone of the large intestine during intussusception is sharply disturbed, that desinvagination is performed under high pressure, that different parts of the large intestine are superimposed on the frontal image. Thus, they came to the paroxysmal conclusion the true width "r" is 4.7 cm. Because the dimensions on the roentgenogram are larger than the true anatomical dimensions, the value of the "rectal" width on the radiograph at standard conditions would be equal to 6.5 cm (**see Figure 1.B**). Thus, in a study by Koppen et al, the normal width of the rectum in the children of 2–6 years was greater than the upper limit of the norm determined for adults (6.3 cm) [8, 11]. Considering that the width of the rectum increases with age, the resulting normal limit does not make any sense.

2.3. Radiometric method. In our research, we proceeded from the following principles:

1) Since the rectum forms two bends in two projections, it cannot be differentiated on the frontal radiograph, which makes it impossible to measure its width **(Figure 2A).** On a lateral radiograph we measured the maximum width of the vertical portion of the rectum, as well as interval not containing contrast medium between the rectum and a contrast marker near the anus on the posterior contour of the tip of the enema (**Figure 2 B)**. This distance is due to the contacted anal canal.

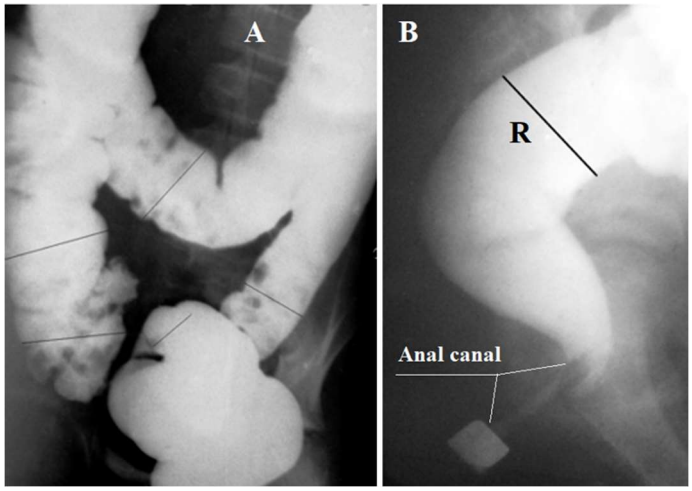

Figure 2. A. On the frontal radiograph, lines mark the measurement sites of all parts of the colon. In the conglomerate of loops of the sigmoid colon and rectum, the rectum is not differentiated and cannot be measured. **B).** Only on the lateral radiograph of the anorectum can one measure the width of the vertical part of the rectum and the length of the anal canal from the rectum to the marker near the anal dimple. Knowing the true diameter of the contrast marker, one can calculate the true width of the rectum (R) and the length of the anal canal.

2) The barium enema was performed after bowel cleansing to measure the width of the different part of the colon, rather than their contents. 3) It is necessary to fill the colon under the same hydrostatic pressure to avoid damage to the colon and to ensure that the filling is fast enough. A barium was introduced from the graded bag. The bottom of this bag at the beginning of the study was located 40 cm above the deck of the table. In the first period a barium was introduced into the colon up to the reflux into the terminal ileum. The difference of barium volume in the bag before and after the colon filling corresponds to the colon volume. Recently, when the standards are already known (Table 1), we fill the colon up to the splenic angle, because after filling the left half of the colon, the rectum stops increasing in size and its width corresponds to its capacity.

Table 1. The true rectal width and anal canal length.

Age	n	Rectal width (cm)	n	Anal canal length (cm)
5 days-11 months	12	1,3-3.0 (2.24±0.09)	7	1.7-2.5 (2.21±0.15)
1-3 years	9	3.0-3.7 (3.21±0.11)	7	2.3-2.8 (2.55±0.10)
4 – 7 years	9	3.0-3.9 (3.43±0.14)	8	2.3-3.6 (3.11±0.10)
8 – 10 years	9	3.2-4.1(3.70±0.06)	8	2.6-3.7 (3.07±0.11)
11 – 15 years	19	3.6-4.6 (39.5±0.07)	18	3.1-3.9 (3.43±0.10)

In children without signs of chronic constipation, we measured all sections of the colon, as shown by the black lines in Figure 2.a. [2]. According to the average indicators for people of different ages, the following graph was built (Figure 3). In newborns and children of the first months of life, the width of all sections of the colon is the same. It increases with age disproportionately and always, in each case, this proportion retains.

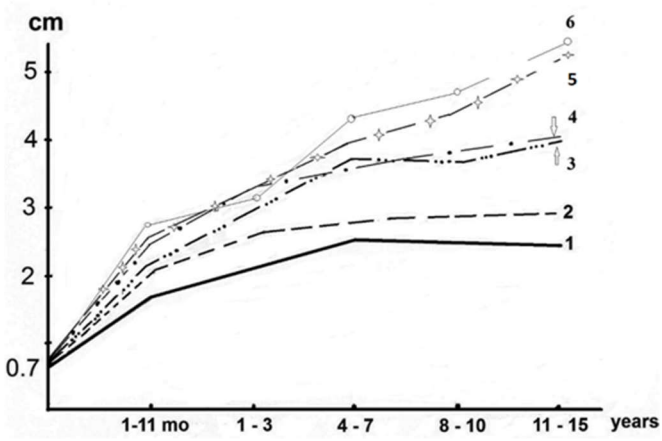

Figure 3. Scheme of expansion with age of the different parts of the colon and rectum: 1- sigmoid colon; 2- descending colon; 3- rectum; 4 - transverse colon; 5 - cecum; 6 - ascending colon.

To compare the different studies as well as studies of the same patient at different ages, we calculate the constant (C), which is the integral characteristic of the colon value.

It is calculated using the following formula:

$$C = \frac{V \times R \times \kappa}{h}$$

Where: **C** – constant, **V** – colon volume (ml).

R – rectal width (cm).

κ – projection distortion factor, which is the ratio of the true width of the marker to its image on the radiograph;

h – patient height (cm).

In healthy children, "Constant" was in the range of 17-31, regardless of age. Megacolon determined if «Constant», exceeds 31. It has been possible to differentiate megacolon varying degrees depending on the constant (C):1st degree - (C = 31- 45); 2nd degree - (C = 45-60) and

3rd degree - C> 60. For the first time on the radiographs the length of the anal canal has been measured, allowing to determine the state of the puborectalis muscle (PRM) [2,11].

3. Functional megacolon

Our study is based on the experience of diagnosing and treating 279 children aged 1 to 15 years with functional megacolon (FM), who were treated at the Belarusian Center for Pediatric Surgery in 1980-1985. [12,13]. In all patients the diagnosis of FM was established based on the barium enema method described above. The final exclusion of Hirschsprung's disease was made using manometric detection of the rectoanal inhibitory reflex in 129 patients, and in 11 cases based on the histochemical study of the of acetylcholinesterase activity. Anorectal malformations and surgical interventions of the anorectal zone were excluded.

We studied the relationship between the time of onset of symptoms and the time of presentation to a surgical hospital in patients with chronic constipation without encopresis (**Figure 4 A)**, as well as in a combination of constipation with encopresis (**Figure 4 B**).

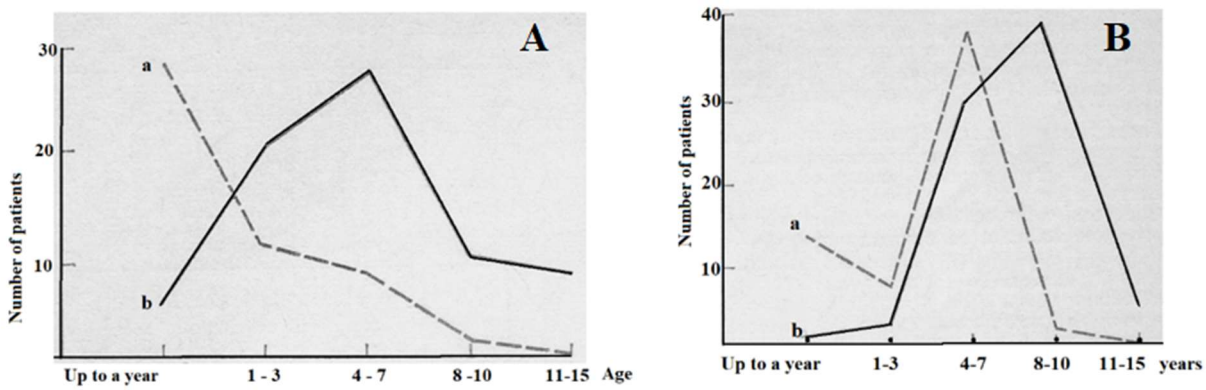

Figure 4. A) Graphic representation of the frequency of megacolon depending on the time of occurrence of constipation (without encopresis) (a) and the time of going to the surgical hospital (b). **B).** Graphical representation of megacolon frequency depending on the time of onset of constipation (a) and the time of onset of encopresis

The graph in Figure 4 A shows that in the first year of life, chronic constipation occurred in 27 patients. However, in most children the constipation was not severe, because only 4 (15%) sought help. In infancy, parents especially scrupulously monitor the condition of the baby, who is under the close attention of pediatricians. The number of new cases of chronic constipation without the development of encopresis sharply decreases with age, but the symptoms of constipation worsen up to 7 years (time of presentation). The graph in Figure 4 shows that a small number of children who developed constipation in the first year of life came to us with

encopresis. However, in most patients, constipation appeared at the age of 3 - 7 years, during potty training and during admission to the children's group. After 2-4 years, constipation was complicated by encopresis. These data suggest two causes (etiologies) of megacolon in this group, which is supported by the results of the study by Duhamel [14] and Clayden and Lawson [15]. Duhamel, in a histological study of the internal anal sphincter (IAS), found that in certain cases, the smooth muscle was dissociated by sclerosis and intimately mingled with striated muscle fibers. In some cases, there was even complete absence of smooth muscle which had been replaced by striated fibers which were more or less fibrotic. These histological aspects were the same as for congenital anal stenosis or rectovulvar fistula biopsy [14]. Clayden and Lawson, in four (5%) of the 79 cases on anal dilatation found a minor degree of anal stenosis, which is a string stricture at the mucocutaneous junction. This string stricture, approximately 1 cm from anal verge, accepted only 2 fingers of the operator's hand during anal dilatation [15]. Thus, minor forms of anorectal malformations, such as anal stenosis, which is mistakenly called rectal stenosis, during the newborn period, especially in the presence of unformed stools, can occur without severe constipation. However, as the child grows, the diameter of the rectum increases, but the diameter of the hole in the membrane does not change. The discrepancy between the width of the feces and the throughput of the rigid opening in the membrane of the anal canal leads to fecal retention in the rectum and the occurrence of megacolon. It follows from this that some patients in whom megacolon is considered functional, it is secondary.

We divided all patients into 3 groups depending on the main clinical symptom.

3.1. In the 1st group there were 95 patients (M:F - 48:47) with an average age of 5.2 years with complaints of chronic constipation. In 27 (28%) children, constipation began in the first year of life, and in the rest from 1 to 5 years (average 2.5 years). In most patients, the feces were large in diameter, and defecation was plentiful. The severity of megacolon is presented in Table 2. In 18% of patients, megacolon, calculated by constant, was absent. These were children under 4 years of age with a recent onset of constipation.

3.2. In the second group there were 129 patients with an average age of 7.5 years (M: F - 2:1). The main complaint was daytime fecal incontinence. Chronic constipation was in each of them. It appeared at an average age of 3.5 years. Encopresis arose at a mean age of 5.4 years. After a cleansing enema, the soiling disappeared and reappeared when the stool is held. Each of them had megacolon predominantly 2nd and 3rd degree. In 5% of cases, penetration of barium into the upper part of the anal canal was observed, the length of which was within the normal range.

This testified to the weakness of the puborectalis muscle (PRM), which could not press the posterior wall of the anal canal to the tip of the enema [16]. In 23% of patients, a significant shortening of the anal canal was observed, which indicated of the PRM failure. This condition is referred to in the literature as Descending perineum syndrome (DPS), which is manifested by incontinence of feces [17, 18, 19] **(Figure 5).**

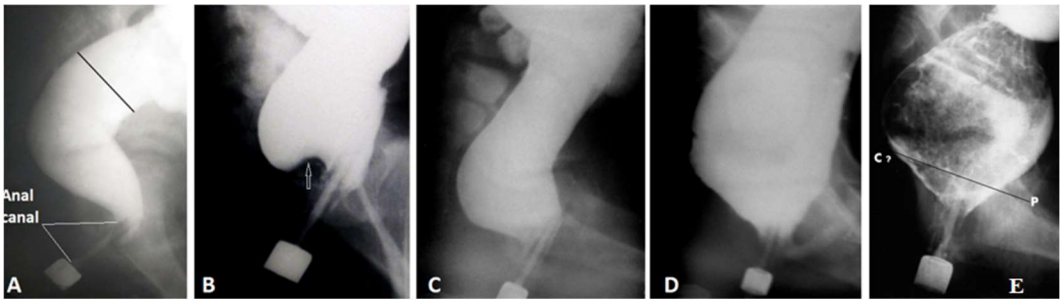

Figure 5. Lateral radiographs of the anorectum in patients with FM at different stages of megacolon development and PRM damage. **(A).** In a 13-year-old patient without constipation (an example of a norm), the vertical and horizontal branches of the rectum are determined. The width of the rectum and the length of the anal canal are normal. On radiographs B-E, the horizontal branch of the rectum is absent due to rectal dilatation. **(B).** A 3-year-old patient has concavity at the base of the rectum (arrow) because of PRM edema. It gives the impression of an elongation of the anal canal. This is the initial phase of FM. **(C).** In a 4-year-old patient, the penetration of barium into the upper part of the anal canal behind the enema tip is determined due to the weakness of the PRM. **(D).** In a 12-year-old patient, a sharp expansion of the rectum is combined by the sharp shortening of the anal canal due to insufficiency of PRM. This is a picture of the descending perineal syndrome, which is often characterized by fecal incontinence. **(E).** The mechanism of anorectal obstruction is shown. The peristaltic wave creates a large force in an attempt to push a large diameter fecal stone through the anal canal. However, the anal canal cannot skip a stone of this diameter. As a result of the efforts of the rectum, the stone bougienages and stretches the muscles of the pelvic floor, which leads to stretching and weakening of the PRM and levator plates.

3.3 The third group included 55 patients with a mean age of 8.8 years (M:F-22:33). They were admitted for examination for recurrent abdominal pain. Constipation was reported in only 33 case histories. In 4 patients, constipation was combined with encopresis. In 5 cases, occasionally there was an unformed stool with mucus. In some patients, the feces had an unusually large diameter, but in some it was in the form of sheep's feces. The constant was calculated in 38 cases. The degree of megacolon was markedly less than in the other groups

(Table 2). Although in 13% of patients the constant was within the normal range, the sigmoid colon was significantly elongated. In it, narrowed or spastic segments were combined with segments much wider than the maximum normal parameters. Slow colon filling during barium enema due to intermittent spastic contractions indicated of high colon tone. Only in one case the PRM was weakened.

Table 2. Dependence of the degree of megacolon and the state of PRM on the main clinical symptoms, age, and sex of patients.

Groups	Average age. yrs.	M:F	Degree of megacolon (%)				PRM weakness	DPS
			No	First	Second	Third		
1st- 95	5.2	48:47	18	39	33	10	5	0
2nd- 129	7.9	86:43	0	19	40	41	18	23
3rd- 55	8.8	22:33	13	50	24	13	3	0

Comparison of indicators in the three described groups shows two options for the development of the disease. It always begins with constipation, which eventually leads to the development of megacolon (group 1). In the absence or insufficiently effective treatment, the number of patients with megacolon grade 2 and 3 is increasing. After about two years, a significant proportion of patients develop fecal incontinence, which is explained by weakness or complete insufficiency of PRM (group 2). There are 2 times more males in this group than females. In 13% patients of the 3rd group, megacolon was absent at the time of the study, but the lengthening and segmental expansion of the sigmoid colon indicated that in the earlier period the sigmoid colon was expanded and lengthened. Most of the patients had first degree megacolon, and there were no patients with PRM insufficiency. In this group, the majority were females. The clinical and radiological picture indicated an increase in the tone of the colon and the presence of signs of nonspecific inflammation (colitis), which was the cause of recurrent abdominal pain (**Figure 6)**.

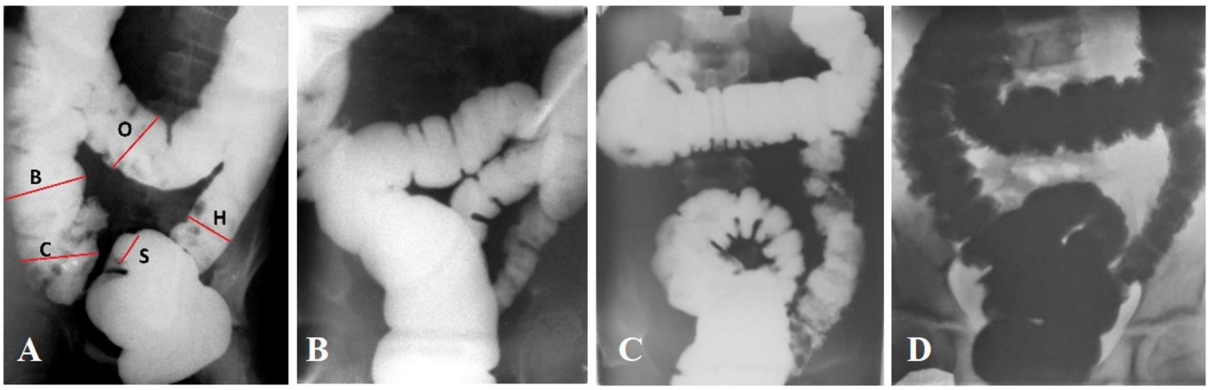

Figure 6. To the pathogenesis of FM in patients of the 3rd group. **(A).** Barium enema in a patient without colon pathology (example of a norm). The rectum is not visible and cannot be measured. Normal ratio of diameters of different departments. The sigmoid colon is the narrowest and at the age of over 5 years is always in the small pelvis. (**B**). In a patient with FM, the expansion of the rectum and sigmoid colon is determined. The sigmoid colon is elongated and located in the abdominal cavity. **(C).** Due to multiple spasms, it was not possible to fill the entire large intestine. All sections of the colon have a normal diameter, except for the sigmoid colon, which extends beyond the pelvis, narrow sections are replaced by wide ones. Very fine asymmetrical haustration. **(D).** All departments are of normal size, except for the sigmoid colon. The fuzziness (fluffiness) of the contours of the intestine is determined, indicating an inflammatory process.

The pronounced sex difference between groups can be explained by the fact that girls are more open for mothers and receive pathogenetic treatment earlier. Boys complain late when they have a sharp increase in the volume of the colon and damage to the muscles of the pelvic floor (group 2). The third group represents a variant of a favorable outcome, despite abdominal pain. Three factors contribute to the normalization of the stool. First, with age, the width of the anal canal increases, which increases the likelihood of restoration of the passage of feces. Secondly, the example of Hirschsprung's disease shows that a serious violation of the passage of feces causes enterocolitis [20]. Smith and Gill described a clinical syndrome of prestenotic enteritis and enterocolitis consisting of abdominal distention and pain, intermittent diarrhea, and constipation. They considered that "The pathogenesis of this enteritis and enterocolitis apparently is related to partial mechanical obstruction of the bowel with proximal dilatation, stagnation, and capillary stasis [21]. This nonspecific inflammatory process, which was observed in patients of the 3rd group, causes an increase in tone colon, which helps to reduce the size of the intestines, including the rectum, and improves the evacuation of feces. Thirdly, the use of stimulant laxatives also increases intestinal tone, which leads to the same favorable

result. However, it is important to stress that the untoward effects which may result from laxative abuse could be greater than those of constipation [22, 23,24].

Discussion

Functional megacolon (FM) is a disease characterized by chronic constipation and enlargement of the colon in the absence of an organic cause (Hirschsprung's disease, secondary megacolon, including anorectal malformations, and acquired constrictions. Currently, this type of constipation is called functional constipation (FC). Authors using this name refer to the Rome IV criteria, which divide chronic constipation into four subtypes: (a) functional constipation, (b) irritable bowel syndrome with constipation, (c) opioid-induced constipation, and (d) functional defecation disorders, including inadequate defecation stimulation and dyssynergic defecation [27]. It should be kept in mind that the recommendations of the Rome IV criteria, which are accepted by practitioners by vote, and have changed over time from Rome I to Rome IV, have no scientific status (significance). Secondly, irritable bowel syndrome with constipation and opioid-induced constipation are different diseases with the symptom of constipation. However, such diseases also include hypothyroidism, taking large amounts of laxatives, cystic fibrosis, acid-dependent diseases (inflammatory processes of the stomach, duodenum, GERD), etc. The fact that these diseases are not included in the differential series is surprising. Eliminating the cause and/or treating the disease leads to a cure. constipation. Third, functional constipation and functional defecation disorders, including inadequate defecation stimulation and dyssynergic defecation, are the same disease, which the authors distinguish based on different research methods. Fourth, the Rome IV criteria state that the pathophysiology of functional constipation is not fully understood, although the prevailing hypothesis relates to disruption of the gut-brain interface [27]. This contrasts with extensive evidence that this type of constipation occurs due to an inability to defecate timely in social settings during early childhood, as discussed below. Fifth, these authors argue that in FC the rectum does not always dilate [27]. This statement is based on a study in which, due to a violation of the methodology, the authors came to the absurd idea that the normal limit of rectal width in children under 6 years of age is the same as in adults [10, 11]. Thus, the name FC does not reveal the causes of constipation, which gives rise to authors who do not know the etiology of FC to call it idiopathic megarectum (IM) [25], or idiopathic constipation (IC) [23]. A situation has arisen when the same authors, including those promoting the Rome IV criteria, in different articles call the same disease by different names (FC, MI, IC and Segmental dilatation of the colon) [28, 29, 30].

The X-ray analysis of barium enema that we developed is based on measuring the width of all parts of the rectum and colon, the length of the anal canal and the volume of the colon in children of different ages. It is based on standardization of hydrostatic pressure, calculation of true dimensions considering the projection distortion of the image on radiograph. First, normal parameters were determined, measured in patients without chronic constipation, in whom barium enemas were performed to diagnose the cause of acute and chronic abdominal pain, anemia, etc. [2]. The idea of the absence of chronic constipation based on a survey of patients and their parents is the only unreliable link in this study. It is possible that among the so-called control persons there could be children with the initial form of the disease. However, this method of diagnosing megacolon has proven highly reliable in practice. For example, the integral value of the colon, which is determined by a constant, was within the normal range only in 18% of patients in group 1, and only in those cases where the children applied no more than six months after the onset of constipation. In all patients with encopresis (group 2), megacolon was determined by a constant. Among patients of the 3rd group, the constant was normal in 13%, which was accompanied by an increase in the tone of the colon and rectum and normalization of bowel movements.

Etiology The reason for the expansion of the rectum and the left half of the colon is the retention of feces above the anal canal, because of a discrepancy between the width of the feces that form in the dilated rectum and the throughput of the anal canal. The present study confirmed the studies of other authors [14,15], who proved that megacolon, which is commonly called FM, is not a homogeneous disease. Constipation that begins in the first year of life most likely results from congenital anal stenosis or membranous stenosis in the anal canal at 1 cm from the anal verge. If the width of the anus with congenital anal stenosis or rectal stenosis causes defecation problems in infants, they are treated promptly in infantcy. However, the diameter of the anus and the opening in the membrane may be wide enough to allow acceptable evacuation of feces in newborns. If the opening is wide enough, constipation occurs when the width of the formed stool is wider than the opening. No matter how wide the opening, its rigid ring does not increase with age, while the width of the rectum increases from 1.3-3.0 (2.24±0.09 cm) in the first year of life to 3.0-3.9 (3.43±0.14 cm) at the age of 4-7 years (see Table 1). For example, Clayden and Lawson, during anal dilatation, which was performed under general anesthesia by introducing 4 fingers of the operator’s hand, in 4 out of 79 patients they could not insert more than 2 fingers due to the presence of a rigid ring, which was at 1 cm from the anal verge. During histological examination of the IAS Duhamel found in some patient fibrous

changes in the anal canal, which were like congenital anal stenosis [14]. It follows from this that when examining patients with megacolon, one must consider the possibility of relatively wide anal stenoses, which cannot be determined by bougienage. Thus, anal distension, which is effective in treating FM, may also be useful for diagnosing secondary megacolon, i.e., anal stenosis.

True FM begins between the ages of 3 and 8 years in results intentionally withholding of bowel movements, due to the stress of entering in a new social group or not paying attention to the rectal urge during games. During a subsequent attempt to defecate, the implementation of wide feces into the anal canal causes pain, which forces the child to abandon this attempt. This is how a closed circle arises, в результате чего длительная deliberate delay in defecation, result in stagnation of large amounts of feces in the rectum, causing it to stretch [31, 32]. When symptoms are more than six months old, the rectum always increases (megarectum), the sigmoid colon not only always increases, but also lengthens, since it is not fixed behind the peritoneum. The descending colon dilates more than normal in 72% of cases, while the right parts of the colon most often do not dilate in children. Dilation of the rectum, sigmoid and descending colons is called megacolon. Since the rectum never expands in isolation from other parts, this pathology is correctly called megacolon, and not megarectum.

Pathophysiology. Retention of feces in the rectum causes increased peristalsis of the rectum and sigmoid colon, trying to push large feces through the anal canal. This causes an increase in pressure in the rectum and left colon, leading to muscle hypertrophy. "The thickness of the rectal wall in children with FC is significantly greater than in healthy ones" [33]. Duhamel showed evidence of internal sphincter hypertrophy on anorectal manometry and the need for vigorous anal dilatation (to accommodate 4 fingers) under general anesthesia in FM [14]. Multiple strong contractions of the rectum, unsuccessfully trying to push wide feces through the anal canal, lead to stretching of the pelvic floor muscles. It has been shown that the PRM first becomes hypertrophied and/or edematous, manifested by an indentation on the inferior wall of the rectum (see Figure 5B). Further bougienage the PRM leads to its stretching and weakening of function, which is manifested by the penetration of barium behind the enema tip, because of the PRM is not able to firmly press the posterior wall of the anal canal to the tip **(see Figure 5 C).** As shown on Figure 5 D, E, 3 to 5 years after the onset of constipation, stretching of the anal canal leads to complete failure of the PRM, which is manifested by a sharp shortening of the anal canal. This is a picture of descending perineum syndrome (DPS), which is usually diagnosed during defecography [17,33,34]. Defecography has two major

disadvantages. First, it is associated with a high dose of ionizing radiation, which is unacceptable for the examination of children. Secondly, the measurement of the anorectal angle or perineal descent with defecography has poor reproducibility, and differences exist between examiners [35]. The barium enema described above with measuring the length of the anal canal on a lateral photograph allows you to accurately measure the length of the functioning anal canal in centimeters. If at the beginning of filling the colon with barium the length of the anal canal is normal, but at the end of the filling it becomes short, we are talking about a milder form of DPS, which is called non-fixed form of DPS. If the anal canal is 2 times shorter than normal from the very beginning of its filling, this form is called fixed DPS. This means that the upper part of the anal canal, which is surrounded by the PRM, does not function to retain feces. In these cases, constipation in children is often complicated by encopresis.

Clayden and Lawson during manometry found that in patients with megacolon the rectoanal inhibitory reflex was detected not with the introduction of 150 ml as is normal, but with the introduction of 200-300 ml due to the expansion of the rectum [15]. A significant increase in the volume of air administered to detect the rectoanal inhibitory reflex was found in every case of megacolon in our studies [12,13]. These data prove that the reflex is excited at a certain (threshold) pressure in the rectum. The wider the intestine, the larger the volume of the balloon (air, feces) is necessary to achieve the threshold pressure. Fathy et al, referring to the Rome criteria, performed manometric studies in FC without mentioning the presence of megacolons. They found that maximal tolerable volume was significantly higher in constipated than in control children ($p=0.03$) [36]. This observation supports the idea that the main cause of this phenomenon is an increase in the volume of the colon. With a long course of FM, which, as shown above, causes an inflammatory reaction, the sensitive elements of the rectal wall can be damaged, which can lead to hyposensitivity to stretching. However, the main factor in the described phenomena is obviously megacolon.

Small volumes of feces constantly arrive in the rectum, which causes an increase in rectal pressure. They stimulate reflex relaxation of the IAS and contraction of the EAS and PRM. After this, the rectum, adapting to the new volume, relaxes and the pressure in it decreases. This retention reaction occurs approximately 18 times per hour [37]. The defecation reflex is possible only when a volume of feces accumulates in the rectum that causes a threshold defecation pressure. The defecation reflex includes the simultaneous relaxation of the IAC, EAS, PRM and contraction of the levator plates, which create a canal in the pelvic floor, which facilitates the passage of feces under the influence of contraction of the rectum [37]. Those

volumes of the rectal balloon, which, during a manometric study in control individuals, cause a defecation reflex, in patients with FM cause a retention reaction, i.e., relaxation of the IAS, and contraction of the EAS and PRM, which prevents defecation. To stimulate the defecation reflex in FM, it is necessary to inflate a rectal balloon of a much larger volume, depending on the degree of megacolon. This is one of the causes of evacuation disorder, and dyssynergic defecation, the cause of which is considered unknown [38, 39 However, defecation dysfunction in FM may also be due to damage to the function of the levator plates in DPS, resulting in a lack of wide opening of the anal canal, which sharply increases the resistance to the passage of stool through the anal canal. In addition, impaired stool evacuation causes changes in the IAS. Its thickens and fibrous changes cause rectoanal or anal achalasia [14,15, 40]. The literature also describes anismus, which is considered as a change in the PPM [41, 42]. Shafik's study showed that rectal distension causes a decline in the intestinal pressure. The author suggested that this reflex inhibits the intestinal transit, thus giving the rectum time to evacuate itself [43]. Thus, slow transit in conditions of long-term retention of large volumes of feces in the left half of the colon is not the cause of constipation, but a physiological response to FM and it is inevitably accompanied by a change in pressure in the colon which depends on the degree of megacolon. Thus, the discrepancy between the throughput of the anal canal and the size of feces that forms in the dilated rectum (anorectal outlet obstruction) is a key factor in the pathophysiology of FM. All other characteristics are secondary. Their diversity depends on the degree of this discrepancy and duration of the disease.

Pathogenesis. Since the width of the anal canal increases with the age of the child, timely treatment (laxatives and enemas) that prevents further expansion of the rectum leads to a situation where the stool formed in the rectum comes out unhindered, which means a complete cure. Delayed treatment leads to a sharp expansion of the rectum, which requires long-term conservative treatment (laxatives, enemas, anal distension). 3-5 years after the onset of constipation, encopresis often appears because of damage to the PRM. Complex treatment may also include the introduction of Botox into the anal canal [44], or into the rectum [45]. In some patients, chronic obstruction causes an inflammatory process, which leads to an increase in rectal tone. As a result of colitis, the diameter of the rectum decreases, which leads to normalization of the act of defecation.

Conclusion A radiometric method for determining the size of the colon, and anal canal length in children of different ages is described, which made it possible to establish that acquired megacolon is always of an obstructive nature. In about 5% of patients, the cause of constipation

is congenital stenosis of the anus with a relatively wide diameter (secondary megacolon). In other cases, constipation is caused by dysfunction of the anal canal (functional megacolon - FM). FM occurs between 3 and 9 years of age, because of an involuntary delayed defecation, which leads to overflow of the rectum. A subsequent painful attempt at defecation causes a conscious delay. Thus, a vicious circle arises, because of which a functional megacolon (FM) develops. After 3-5 years, because of the bougie effect of the fecal stone, the pelvic floor muscles are stretched and weakened, which is manifested by a shortening of the anal canal. Measuring the length of the anal canal allows one to determine descending perineal syndrome during a routine barium enema. Due to the weakness of the puborectalis muscle encopresis occurs. Prompt treatment can lead to complete recovery. With third degree megacolon, treatment should be long-term and complex. Anorectal achalasia is also complicated by hypertrophy and sclerotic changes in the internal anal sphincter. In addition, because of stretching of the levator plates, during an attempt to defecate, the contraction of the stretched levator plates does not open the anal canal, which sharply increases the resistance to the movement of feces. In some patients, chronic obstruction causes an inflammatory process, which leads to an increase in the tone of the colon and a decrease in the diameter of the rectum. This leads to the normalization of bowel movements. It has been shown that megacolon is the key to understanding such pathophysiological phenomena as slow transit, changes in pressure in the colon, dyssynergic defecation, etc.

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