DIAGNOSIS AND TREATMENT OF CONGENITAL MEGALODOLICHOCOLON

The results of diagnosis and treatment of congenital anomalies of the colon in 107 children were examined. Twenty-nine patients underwent surgical correction: 24 intra-resection of the colon by Rehbein and 5 abdominoperineal rectoplasty by Soave-Lenushkin and Soave-Boley. In extending of colon in combination with megarectum the preference is given to Soave operation and its modifications.

Keywords: Chronic constipation, dolichosigmoid, dolichocolon, multispiral computed tomography, Rehbein’s and Soave surgery.

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Introduction

The constipation is one of the most widespread disturbances of the function of digestive system, from 12% up to 19% of the population suffer from this disease (Locke et al., 2000; Higgins and Johnson, 2004). According to the Roman criteria III, the constipation is considered to be chronic, if symptoms are observed during period not less than 6 months. Typical symptoms of constipation are rare and hard feces, exertion, and and feeling of incomplete emptying of the bowels (Longstreth et al., 2006). The hard feces, exertion and increase of intraperitoneal pressure can become the reason of such complications, as hemorrhoids, anal fistula and, probably, prolapse of the rectum, formation of the feces ulcer and feces accumulations with feces incontinence due to overflow bowel.

In spite of the fact that chronic constipations (CC) do not seem to be serious surgical diseases, the harm, which they put growing body, is very great. Practically in all children with long-term CC there is seen delayed physical development, connected with constant endotoxicosis, caused by the increased permeability of the intestinal barrier. In these children marked anemia is found, nutrition is damaged, frequent allergic expressions, and endotoxicosis can result in changes of biochemical parameters of blood and other changes (Kirghisov et al., 2005).

According to the data of sociological interviews, 16-32% of the parents note the tendency to constipations in their children. Before the pubertal period the constipations occur with equal frequency both in the boys and girls (Weaver and Steiner, 1984).

The constipations may be found in organic lesions of the colon, in connection with this they are divided into organic and functional. In a number of cases the excessive lengthening (dolichocolon) and distention (megadolichocolon) of the colon are underlying the chronic constipations. The term “dolichocolon” means unusually large length of sigmoid, that is interpreted as inherent anomaly of the colon development and can not express clinically during all life of the human. There are different opinions about formation of dolichocolon. The lengthening of the bowel is considered as a defect, or variant of development, or as anomaly (Henry and Svosha, 1985; Romanov, 1987; Vorobey et al., 2005). According to the data of the different authors, dolichocolon accounts for from 9 up to 11% of all anomalies of development of the colon (Lenushkin et al., 2002; Achkasov, 2003; Vorobey et al., 2005; Podolskaya-Devochko et al., 2006). At the same time dolichosigma, the frequent type of the colon lengthening, meets 15% of healthy children (Baranov and Klimanskaya, 1999). The appearing linical picture quite often simulates such diseases, as Hirschsprung’s disease, acute appendicitis, gastroduodenitis, chronic intestinal obstruction, the colon tumou. Many researchers consider that dolichosigma, megadolichosigma, dolichocolon and megadolichocolon are stages of the same defect, and disturbance of the moto-evacuating function of the colon is...
the basis of this malformation. Lengthening or lengthening with distention can be located within the limits of separate anatomic segments of the colon as megarectum, dolichosigma, subtotal or total megadolichocolon (Baranov and Klimanskaya, 1999; Lenushkin, 1999; Prudnikova, 2003). Today one of the multiple methods offered for conservative and surgical treatment has no 100% efficacy. Many authors show that the rate of unfavourable results after operative interventions achieves 27.3-45.9% (Romanov, 1987; Yaremchuk et al., 1990).

During the last decades in the treatment of chronic colostasis the colorectal surgery has passed the stages from performance of segmentary resections of the sigmoid bowel, including resection of dolichosifma and left side hemicolectomy, to expansion of volumes of the sigmoid resections, including subtotal and total colectomy to colproctoectomy with formation of intestine reservoir. However, precise indications to surgical treatment have not been developed yet, and the volume of surgical intervention for children has not still determined (Grigoriev and Yakovenko, 1998; Sukhova, 1998; Lenushkin et al., 2002; Achkasov, 2003; Vorobey et al., 2005).

**Materials and methods**

During the period from 2000 to 2010 on the clinical base of the Faculty of Hospital Children’s Surgery of TashPMI 107 children of the age from 3 years to 14 years with lengthenin and distention og various segments of the colon were under inpatient treatment: sigmoid bowel (dolichosigma) - 83 (77.6%), two and more segments (subtotal dolichocolon) - 14 (13.1%), lengthening of all segments of the colon (total dolichocolon) - 10 (9.3%). 29 (27.1%) patient was carried out surgical treatment.

Taking into account the complex character of pathology of the colon in children, for statement of the diagnosis the complex examination was performed. Except for clinical researches (anamnesis, examination, palpation examination of the rectum) the important place was allocated to instrumental investigation: ultrasonograpy of the abdominal cavity, kidneys and the colon at the ultrasound scanner FF-sonic UF-4100 of firm Fukuda Denshi. Contrast irrigography or virtual colonoscopy was performed in all children, paying attention to the sizes of the colon: its length, uniformity of a diameter, sites of expansion and additional loops. Besides there was done endoscopic investigation (recto- and colonoscopy) and morphological study of the removed segment of the colon.

**Results and their discussion**

Among the patients suffering from the chronic constipation owing to dolichocolon, the correct diagnosis at referring was made only in 27 (25.2%) patients. Others 80 (74.8%) were reffered to consultation and treatment due to various diseases, which final diagnosis was established at complex examination in the hospital.

The damage of bowels emptying (meteorism, constipations) was noted in 100 % of the patients, it may be considered as early manifestati on of illness. The duration of constipation was from 2 to 7 days. In 5 children with total dolichocolon and in 24 patients with dolichosigma the constipation expressed in the age of 4 months, and later with introduction of additional artificial feeding and transition of the the child to mixed feeding the constipation increased. Strong meteorism is induced by prolonged stagnation of feces in the colon, and sometimes by bending of the bowel loops, and also by valve-shaped folds of mucosa, detaining gas and chimus evacuation. Because of excessive length of the bowels and formation of bendings the considerable disturbance of blood circulation in the organ of abdominal cavity occurs quite often. Above the bending the bowel is distended by gases, and the vevous stasis is formed, that even more worsens conditions of the bowels feeding and contributes to its inflammation. On this ground there are occurred sharp catarrhal symptoms, ulcer on the mucosa, thickening of a bowel wall, peristalsis reduing with signs of atonia and formation of in the further of feces stones.
The constipations in 59 (55.1%) patients were accompanied by pains in the abdomen. The pains in the abdomen were persistent or spasmodic, from mild to moderate intensity. They increased at change of a mode of feeding and physical loading. Their localization was changeable, more often in the left ilioumbelical side - in 32 (54.2%), in the epigastral area - in 9 (15.2 %), in the left and in the right hypochondrium - in 7 (11.9%), in the bottom of the abdomen - in 11 (18.6%) cases. In 48 (44.9%) cases the sigmoid bowel, overflowed with feces was palpated.

The symptoms of chronic bowel obstruction were revealed in 23 (21.5 %) patients. From anamnesis there was revealed, that sometimes additionally to constipation and aching pains in the abdomen unexpectedly there is occurred increased peristalsis, difficulties in gases evacuating, andominal distension, increased pains in the abdomen, nausea, sometimes womiting. These symptoms were observed often, but were expressed mildly and became habitual for the patients.

At dolichocolon in 64 (59.8%) patients the deficiency of nody weight, hypochromiv anemia - in 53 (49.5%) patients was noted. Often the intestinal symptoms mask by dyspepsia complaints (in 34-31.8%) (heartburn, eructation, pain in the pit of the stomach), and also because of feces intoxication there are observed (at 36-33.6%) depressed mood, apathy, somnolence, headaches, dizziness, which direct an idea of the doctor in direction of the gastric pathology and psychological sphere. In 9 patients (8.4%) children chronic constipation was accompanied by encopresis of various degrees.

The main method of diagnosis of dolichocolonh is contrast irrigoscopy and irrigography in retrograde filling with barium and the colon emptying. The quantity of contrast substance manifested depends on age of the child and colon functional condition: to children from one year till 3 years - 150-200 мл, from 3 till 7 years - 250-350 мл, and children of the senior age - up to 500 мл. Thus, the attention was paid to length and form of various segments of the colon, speed of filling and emptying, unrolling and tonicity of walls, presence of spasms. Roentgenological investigation showed lengthening of the sigmoid bowel (dolichsigma) in 83 (77.6%) patients, lengthening of two and more segments (subtotal dolichocolon) - in 14 (13.1%), lengthening of all departments of the colon (total dolichocolon) - in 10 (9.3%) patients. In 23 (21.5%) patients alongside with lengthening there was noted distension of the colon (megadolichocolon and megarectum). In 9 (8.4%) cases dolichosigma was combined with right-side colonoptosis and in 5 (4.7%) cases - with Pauψ syndrome.

Roentgenological picture of changes in the colon in the patients reflected not only lengthening and distension of its departments, but also, as a rule, bad emptying from a contrast suspension after the defecation act, mucosa swelling relief in the places of lengthening. The performance of irrigoscopy in position of laying and standing allows more detailed study mobility of the sigmoid bowel, ptosis and arrangement of physiological bends of the colon, as the development of soloichocolon depends not only on the lengthening of sigmoid bowel, but on the pathological mobility of cecal and ascending bowels, colonoprosis and absence of ligament apparatus of the colon, which in norm are points of fixing at gradual peristalsis and passing of nutritional masses from one point of fixing to another.

Last years in diagnosis of dolichocolon the multisliced computed tomography (MSCT) and virtual colonoscopy (VCS) began to use. Computes-tomohraphic (CT) investigations were performed on the computed tomograph Brilliance-64 of firm Phillips. VCD differs by the greatest comfort of investigation for the children in comparison with the fibrocolonoscopy (FCS) and irrigoscopy (IS), with sufficiently high capacity and lower (in comparison with IS) radiation loading. The reinvestigation showed 100% sensitivity of VCS in revealing anomalies of colon development. The advantages of VCS included the large spatial realization, opportunity of the assessment of interrelation of abnormal colon with other organs of adominal cavity, opportunity of study of the bowel wall and perintestinal changes of the of volumetric representation of pathologically changed colon, that is important for planning operative intervention.
62 (57.9%) patient underwent ultrasound investigation of the colon. The main stage of investigation of the distal segments of the colon is its evaluation in filling with liquid (1% solution of sodium chloride, at temperature +30ºС). The volume of liquid administered to children at the age from 1 year till 7 years - 100 ml for year of life, from 8 till 15 years - 800-1000 ml. In all patients the ultrasonic picture of dolichocolon and dolichosigma is revealed. At filling of the bowel with the liquid there are clearly defined loops of sigmoid bowel, especially precisely visualized at the moment of bowel filling, when the liquid passed from one loop into another. The greatest changes in dolichosigma the muscular layer underwent. In norm its wideness in spite of age does not exceed 2 mm. At long constipation the thickness of this layer may increase to 10 mm, its regularity damaged, that is, hypertrophy development. Ultrasonography is perspective method of diagnosis of the congenital lengthening of the colon, accompanying by chronic constilation.

Treatment at inherent lengthenings of the colon began from complex conservative measures directed to increase in bowel peristalsis, softening and simplification of feces evacuation from the colon. The conservative treatment includes:

a. diet therapy: at constipation the diets No.3, No.4 (B) by Pevzner are prescribed. The diet No.3 is nominated at constipation in the period of mild exacerbation and remission, and also at a combination with lesion of stomach, liver, biliary tract, pancreas, functions, promoting restoration of damaged functions of the bowel and other organs of digestive system, decrease in dystrophic processes in the organs. Diet No.4 (B) by Pevsner is a physiologically full value with normal contents of all nutrient ingredients and includes products rich in rude cellulose, stimulating bowel peristalsis. The products and meals increasing the processes of fermentation and putrefaction. The food prepared on the steam, is boiled in the water; vegetables and fruits in uncooked and boiled type - dry-eating is excluded;

b. medicamentous therapy: preparations stimulating peristalsis (proserpin, ubretid and others), vitamins of group B, ATF, glicin, motilium; at the early period of treatment - laxatives (not saline purgatives): dufalak, guttalax, microlax, Vaseline oil and others;

c. medical gymnastics strengthening muscles of the abdominal press; massage;

d. siphon enema to the full bowel clearing;

e. disbiosis correction - pro- and prebiotics (nactisubtyl, bifidumbacterin, lactobactobacterin); about and препараты (botektобактерии, лактобактерии);

f. physical therapy: electrophoresis, bowel electrostimulation, thermal procedures, diathermia etc.

After one- and two-fold course of conservative therapy in 79 (73.8%) from 107 children the general health state was considerably improved, the abdominal pains stopped or decreased, and constipation intensity decreased too.

In 29 (27.1%) children (from them 16 with dolichosigma, 7 - with subtotal dolichocolon, 6 - with total dolichocolon) the repeated course of complex conservative therapy has appeared inefficient, and they were оперированы. The indications to surgical treatment of dolichocolon were the increasing symptoms of disturbance of function of bowel emptying, pain syndrome, presence of the extreme pathological forms of the colon long loops - “double-barrelled gun”, omega and damage and absence of fixation.

The method of surgical correction and volume of intervention at the developed indications to operation has not been determined. Thus, the problem of chronic, colon stasis is urgent, and the resolving of it can be supplied by development of adequate tactics of treatment, including surgical. Резекция of excessive segment of the colon with fixing of the remained segment, in our opinion, should be used for surgical correction of chronic colostasis, caused by dolichocolon. The operative treatment should be preceded by high-grade investigation for exception of other reasons of chronic colostasis.

From 29 radical operations 24 (82.7%) were performed by intraabdominal technique by Rehnein (intra-abdominal resection of a superfluous part of the colon with formation of
colorectal anastomosis “end-to-end” with application of anastomosis outside of peritoneum. The method of Rehbein with fixing of sigmoid bowel in a physiological position is an effective method of surgical correction of congenital lengthenings of the colon in children. Its advantages consist of smaller traumatism, safety of obtrusive apparatus of the rectum and reflexogenic zones of the small pelvis, as well as in an opportunity of application at any forms of dolichocolon with the exception of megaureter. The volume of the operations performed: 19 - segmentary colectomy; 10 - left-side hemocolectomy. Total colectomy is considered not rational in children because of not indifferent of this intervention for growing and developing children's organism.

At lengthenings and distension of the colon with involving of the rectum as megaureter F.Rehbein offered to resect from a back wall of the rectum of triangular flape with suture of the formed defect with interrupted sutures, thus resulting in diameter of rectum correction accordingly to the diameter of sigmoid bowel. Then the “end-to-end” colorectal anastomosis is formed. The similar technique was offered also for a situation, when the diameter of sigmoid bowel is considerably exceeds a diameter of rectum. In these cases the flape is resected from the margin of the sigmoid bowel against mesenterium with suture of the defect of bowel wall in the longitudinal direction with interrupted sutures.

The method of abdominal-perineal proctoplasty used in hirschsprung’s disease, Soave operation in various modifications (which essence consists in demucosation of distal part of the colon with subsequent intrarectal pulling through of the sigmoid to the perineum in the muscle flap of the rectum) can not be recommended at dolichocolon because in this case besides pathologically changed colon (sigmoid bowel, ascending colon etc.) the rectum is removed too, anal internal sphincter is injured, the anorectal angel changed, having important value in the mechanism of anorectal retraction. We consider, that abdominal-perineal proctoplasty by Soave in modification by Lenushkin and Boley in lengthening of the colon with signs of megaureter seems more radical and effective technique of surgical correction. In our observations 5 (17.2%) patients at dolichocolon with signs of megaureter were operated by method of Soave-Lenushkin-3, Soave-Boley-2.

The results of histological investigations of resected segments of the colon also show rationality of the operative intervention because in this case there were revealed atrophy of the mucosa, marked sclerosis of the unmucosa layer with presence of multiple lymphoid follicules; degenerative changes of the muscular fibres: focal dystrophy of the myocytes and presence of great number of collagenous fibres; decrease in numbirt of nerve ganglia; dystrophic and atrophic changes of the gangleous cell nodes of Meissner and myenteric plehuses; serous membrane thickening with sclerotic mesenteric changes.

In the postoperative period the early complications connected directly to an operative technique, are notfound. The lethal outcomes were not.

The long-term results of the operative treatment were observed in 26 (89.6%) from 29 operated patients. The follow-up period was deom 1 to 8 years. The final evaluation of the functional result of the surgical treatment of dolichocolon was performed not earlier than 12 months later after operation. In 22 (84.6%) cases the good functional results were achived, in 4 (15.4%) - satisfactory results. The complaints from them were absent; the children are developing according to the age. A stomach is soft, the signs of coprostasis were absent. At control contrast irrigoscopy the sizes and position of the colon corresponds to age parameters. The unsatisfactory functional outcome in our observations is not fixed.

**Conclusion**

The local forms of congenital lengthening of the colon My be considered as varial of norm. Subtotal and total lengthening with distention proceeds more persistently with signs of chronic colostasis, with great sensitivity to development of inflammatory changes, aggravating their course.
Diagnosis of congenital lengthening of the colon should be based on the complex evaluation both of its structure and its function, as well as assessment of the efficacy of therapy performed.

A wide range of clinical symptoms and efficiency of conservative therapy of congenital lengthenings and distension of the colon are connected to the form and duration of disease, with a kind of complications appeared, individual compensatory possibilities of the organism, as well as depends on the presence or absence of the disorders of the colon fixation.

At dolichocolon intrabdominal resection of the excessive part of the colon with formation of colorectal anastomosis by Rehbein with fixation of the sigmoid bowel provides good immediate and long-term results. In dolichocolon with megarectum the preference should be given to the operation by Soave and its modifications.

References

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