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OUR EXPERIENCE IN THE TREATMENT OF HIRSCHSPRUNG'S DISEASE IN CHILDREN

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ABSTRACT

Hirschsprung's disease remains one of the most severe congenital malformations of abdominal organs in children requiring complex reconstructive surgical interventions. Despite the high incidence of the disease and dangerous complications of both the disease itself and the course of the postoperative period, there is no modern single algorithm for the diagnosis and treatment of children with Hirschsprung's disease. Technical features of the bowel reduction, the residual aganglionosis zone lead to the need for repeated operations. Despite the apparent study of Hirschsprung's disease, there are still problems of its diagnosis and treatment in children.

KEYWORDS: *Hirschsprung's Disease, Surgery, Colostomy, Children.*

INTRODUCTION

Hirschsprung's disease — congenital megacolon, intestinal aganglionosis. According to European studies of recent years, mortality in Hirschsprung's disease ranges from 0 to 3% [1]. A number of authors indicate that in the first year after surgery, the mortality rate is 9% [2]. At risk are patients with Down syndrome, recurrent hirschsprung-associated enterocolitis and total aganglionosis. Technical features of bowel reduction, residual aganglionosis zone lead to the need for repeated operations [3].

There are very few works devoted to Hirschsprung's disease with a super-short form [4]. Despite the successes achieved, unsatisfactory treatment results remain. Great importance is attached to the quality of life of children with this pathology [5, 6]. At the same time, there is practically no data of such studies in children with a super-short form of the disease.

Despite the apparent study of Hirschsprung's disease, there remain problems of its diagnosis and treatment in children.

The purpose of the research: diagnosis and determination of treatment tactics in various clinical forms of Hirschsprung's disease in children.

Material and methods of the research: This work is based on the analysis of the results of diagnosis and treatment of 245 children with various clinical forms of Hirschsprung's disease.

All patients were treated in the Department of Neonatal surgery and the Department of General Surgery No. 2 of the 2nd clinic of the Samarkand State Medical University for the period from 2008 to 2017, there were more boys - 173 (70.5%), girls - 72 (29.5%). Children under 6 months 97 (39.6%), under 1 year 39 (16%), 1-3 years 47 (19.2%), 3-5 years 30 (12.2%), 5-7 years 13 (5.3%), over 7 years 19 (7.7%). The examination complex included a targeted survey of parents, a thorough history collection, a finger examination of the rectum, an overview radiography of the abdominal cavity, passage of the gastrointestinal tract, ultrasound sonography, irrigation. After a complete clinical and laboratory, instrumental examination, the following anatomical forms of Hirschsprung's disease were identified (Table 1).

TABLE 1 DISTRIBUTION OF PATIENTS WITH HIRSCHSPRUNG'S DISEASE BY AGE AND ANATOMICAL FORMS OF THE DISEASE

Age	**Anatomical forms of the disease**				**Total**
	rectal	**recto-sigmoid**	**subtotal**	**total**	
up to 1 years old	102(41,6%)	27 (11,4%)	5 (2%)	3 (1,2%)	137 (56,0%)
1– 3 years old	35 (14,2%)	11 (4,4%)	1 (0,4%)		47 (19,1%)
3 – 5 years old	21 (8,6%)	7 (2,8%)	-	2 (0,8%)	30 (12,4%)
5 – 7 years old	12 (4,9%)	1 (0,4%)	-	-	13 (5,3%)
> 7 years old	17 (6,9%)	1 (0,4%)	-	-	18 (7,3%)
Total	**187 (76 %)**	**47 (19,2%)**	**6 (2,45%)**	**5 (2,0 %)**	**245 (100%)**

Table 1 shows that the rectal form was more common, it was diagnosed in 143 (58.3%) cases, rectosigmoid was noted in 89 (36.5%), subtotal form in 6 (2.4%), total form in 7 (2.8%) patients. Of 245 patients, 14 (5.7%) underwent colostomy during the newborn period.

Results and their discussion: All children complained of persistent constipation, lack of independent stool, decreased appetite, increased abdominal size, lack of effect from conservative therapy. Young children were admitted in a serious condition with fecal intoxication.

Treatment tactics depended on the length of the aganglionic zone and the severity of the course of the disease. In the rectal form, the compensated course of the newborn was carried out one or two-fold anal opening devulsion. With the rectosigmoidal form of decompensated flow, subtotal and total form, a colostomy was superimposed above the aganglionic zone. The basis of radical surgical treatment of Hirschsprung's disease is the etiological principle of removing the affected

part of the intestine and reducing the anatomically and functionally normal part of the intestine. Radical operations were performed from the age of one year. Surgical interventions were performed in an open way using the techniques of Duhamel-Bairov in rectal forms of u-78 (55.3%), and in rectosigmoid forms of u-31 (22%) and according to Soave-Lenyushkin in rectal forms of u-21 (14.9%), and in rectosigmoid forms of u-11 (7.8%) patients. After the Duhamel-Bairov operation, 15 patients had partial fecal incontinence, the cause of which was a posterior rectum spur (sail), which was eliminated by repeated application of the Bairov clamp. After the Soave-Lenyushkin operation, 3 patients had early falling off of the stump of the reduced intestine and intestinal retraction.

CONCLUSION

Thus, with an ultrashort segment of Hirschsprung's disease, a compensated course was conservative treatment, and with decompensated currents, colostomy was applied to young children as preparation for radical surgery, followed by radical surgical intervention according to Duhamel-Bairov or Soave-Lenyushkin.

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