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THE DISEASE GIRSHPRUNGA BESIDE ADULT: MODERN APPROACHES TO DIAGNOSTICS & TREATMENT

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ABSTRACT

The Material of the study has formed 78 sick, found on stationary treatment in Republican Scientific Centre Coloproktologii since 1992 on 2010. As it is seen, from table, from 78 sick mans was 58(74,3%), womans 20(25,7%). 19(24,3), sick were at age from 15 before 20 years, 49(60,2%) at age from 21 before 40 years and 13 (16,6%) sick from 41 before 60 years.

The Main complaint sick at arrival were a stubborn constipations, which noted beside 70 (89,7%) sick, including absence of the independent chair existed - beside 55(70,5%), but beside 54(69,2%) sick were noted periodic stomachache, growing on measure of the absence of the chair. The Ballooned belly existed beside all 78 (100%) sick moreover beside 20(25,6%) of them flatulence was constant. The Sickness and retching existed beside 24(30,7%), weakness, reduction to capacity to work beside 52(66,6%), increasing of the temperature of the body beside 10(12,8%), paradoxical diarrhoeas beside 6(7,6%) sick. Endoskopicheskiy method (rectoromonoscopy, colonoscopy) turned out to be else less informations - 51,8% coincidences of the diagnosis. So we biopsy on Svensonu executed beside all sick, entered with suspicion on disease Girshprunga. In our observations from 78 sick beside 42(53,8,1%) were aboveanalni, beside 20(25,6%) sick - rectalis, beside 13(16,6%)- rectosigmoideys , beside 2(2,5%) leftside and beside 1(1,2%) sick - subtotalis form hipoganglios. At biopsies on Svensonu on observations, from 78 sick, beside 44(56,4%) is revealled hipoganglios, but beside 35(44,8%) - aganglios rectum. As can be seen from presented tables, from 78 sick beside 68(87,2%) us is executed onemoments radical operation, 10(12,8%) sick is as far as possible made resection hipo- or aganglionarnaya of the zone, decompensate part of the large intestine and is formed colostomy. In all events at operations. The Remote results executed radical operation on cause disease Girshprunga traced from 1 before 10 years beside 57(73%) sick. The Results of the surgical treatment were valued on scale Vezika: good, satisfactory and unsatisfactory. In our observations beside 46(80,7%) sick results came in well, beside 10(17,5%) - satisfactory and beside 1(1,8%) sick was an unsatisfactory result.

UDC Code & KEYWORDS

■ 616.348-007.61 ■ HIPOGANGLIOS ■ AGANGLIOS ■ MEGARECTUM ■ DISEASE GIRSHPRUNGA ■

INTRODUCTION

The Hirschsprung's disease in adults is one of most vice malformation of colon. In spite of modern development of coloproctology the problem of the diagnostics and treatment of Hirschsprung's disease in adults remains actual and not solved in full.

Many of such patients get well-timed and adequate help by children surgeons but certain forms of this disease get a full develop in adult only. [1, 2, 3, 4].

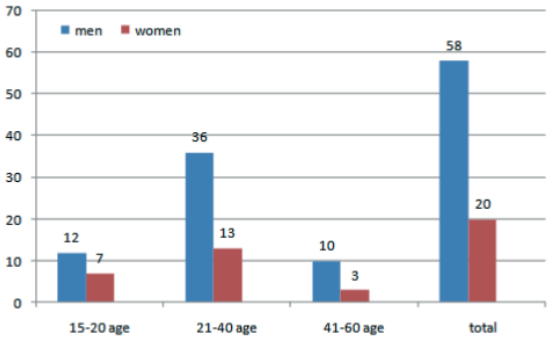
Aganglionary and hypoganglionary zones are most often located in a rectum (86%) and more than half times are only in infrorectal division. The affection of other parts of large intestine exist in 14% sick [2,3].

Experience shows that in adult surgery still make resections of dilated parts of large intestine and other palliative operations.

Suppose of our study is an improvement of the diagnostics and detection of the most effective methods operations at Hirschsprung's disease.

Material of our study is 78 patients that where on the stationary treatment in Republic Scientific Center of Coloproctology of Republic Uzbekistan.

Graph 1: Studied patients



Apparently from the table of 78 patients men where 58 (74,3 %), women 20 (25,7 %). 19 (24,3%), patients were at the age from 15 till 20 years, at the age 49 (60,2 %).

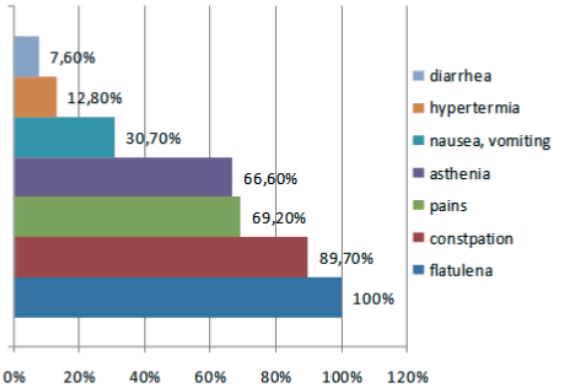
Investigation included: the endoscopy; the radiological; physiological (spincterometris, electrocolografy), morphological (biopsy a material and operational preparations) methods.

The basic complaints of patients at entering were persistent constipations - with 70 (89,7 %) patients, including absence of an independent chair was observed - at 55 (70,5 %), and at 54 (69,2 %) patients became perceptible the periodic abdominal pains accruing in process of absence of a chair. The abdominal distention was observed at all 78 (100 %) patients, and at 20 (25,6 %) from them the meteorism was to constants. The nausea and vomiting were observed at 24 (30,7 %), delicacy, depression of working capacity at 52 (66,6 %), a fervescence at 10 (12,8 %), paradoxical diarrheas at 6 (7,6 %) patients.

More than half of the patients 58 (74,3 %) noted the disease beginning from children's age

Important diagnostic researches are manual research of a rectum and the rectoromanoscopy. At manual research paid attention to a condition of a sphincter of an anus, presence of strictures, cicatrixes, etc. At the rectoromanoscopy transition from slightly narrowed a distal part of a rectum in sharply expanded proximal departments is characteristic. In

Graph 2: Clinical symptoms (n=78)



this zone often are fecal masses or fecal stones, despite thorough preparing of an intestine for research.

One of leading methods of diagnostics of Hirschsprung's disease is the colon X-ray inspection. Research of a colon by means of an opaque enema allows to define a place aganglyonary, a hypoganglionic part, they sizes and extent of expanded parts of a colon. One of the basic radiological signs of Hirschsprung's disease in adults is presence of enough sharp, accurate transitive zone from the narrowed distal departments to dilated which occupy often all abdominal cavity. Diameter of these departments fluctuates from 8-10 to 12-15 sm and more.

In diagnostics from functional methods of research the anorectal manometry defining a condition of a rectoanal reflex has great value. The Negative rectoanal reflex from an internal sphincter testifies to presence of Hirschsprung's disease.

The most difficult for diagnostics are cases of a finding short aganglyonary zones (proctal, overproctal) of rectum, and hypoganglionic zones of a colon. Therefore exact revealing aganglyonary zones probably not in all cases.

For differential diagnostics Hirschsprung's disease with other forms the megacolon is used a transproctal biopsy of a wall of the rectum, the offered O.Svensonom in 1955.

Necessity of application of the transproctal biopsy is caused by frequent contradiction of the information received by other methods.

By means of radiological inspection of a colon Hirschsprung's disease has been defended by 61,2 % of patients. The endoscopic method (a proctosigmoidoscope, a colonoscopy) has appeared even less informative 51,8 % of coincidence of the diagnosis.

Therefore a Svenson's biopsy has been executed at all patients arriving with suspicion on Hirschsprung's disease.

On prevalence hypo- or aganglyonary zones allocate following variants of Hirschsprung's disease: overanaly, rectal, rectosigmoid, link sided, subtotal, total.

In our observations from 78 patients at 42 (53,8,1 %) were overanal, at 20 (25,6 %) patients - rectal, at 13 (16,6 %) - rectosigmoid, at 2 (2,5 %) link sided and at 1 (1,2 %) the patient - the subtotal form hypoganglyosis.

The Svenson's biopsy has showed that hypogangliosis was revealed in 44 (56,4%) patients and agangliosis was in 35 (44,8%) from 78 patients.

But not only length of process or it's localization enough for successful treatment of disease.

Prevalence hypo- or aganglyonary zones:

№	Prevalence	Patients	%
1	Overanal	42	53.8
2	Rectal	20	25.6
3	Rectosigmoid	13	16.6
4	Link sided	2	2.5
5	Subtotal	1	1.2
6	Total	78	100

The estimation definition of this data is reached, as we have specified above, by means of radiological and endoscopic inspections, it is obligatory with research motor-evacuation function of all gastroenteric tract. The diagnosis at Hirschsprung's disease should include extent aganglionar or hypoganglionic zones and also an expanded zone.

At all 78 patients aren'ted expansion of various parts of a colon from moderately expressed (diameter of an intestine made from 7 to 10 cm) to appreciable (15-25 cm). Degree prevalence of a megacolon was various, but obvious prevalence of expansion of distal parts became perceptible. Expansion only a rectum is established by us at 16 (20,5 %), a direct and sigmoid intestine at 51 (65,3 %) patients, the left parts of colon have been dilated at 8 (10,2 %), the subtotal megacolon is taped at 1 (1,2 %) the patient.

The diagnosis of Hirschsprung's disease is the indication for surgical treatment. Application of conservative treatment is surveyed only as preoperative preparation. Overall objective of surgical treatment of Hirschsprung's disease is excision hypo- or aganglionary the zones, an adequate resection of expanded parts in decompensation and saving of a functioning part of a colon.

Sometimes we have very desolate conditions as this disease when many zones or whole colon out to be on galley proof of decompensation. In such cases discussion of questions of surgical tactics, expediency operation separation into some stages with preliminary formation of colostomy is required.

The time of performance of radical operation after colostomy defined depending on the reasons induced to use several-stages treatment.

The radical treatment is possible through 2 -3 mounts if the reason is intestine impossibility or fecal stones present. But if the reason of several-stages treatment is a chronic intoxication than the main stage of treatment should be postponed for 9 – 12 month. But if colostomy was made for compensation of proximal parts of colon then the radical operation should be postponed for 12 – 18 month. The optimum term of the second stage of surgical treatment is defined by a dispensary observation for patients.

As shows our experience the most adequate and radical for treatment of Hirschsprung's disease in adults is the operation offered in 1956 by French surgeon Duamel in

Character of the surgical operations applied at Hirschsprung's disease in our clinic, was the following:

№	Name of operations	Operations	%
1	Dumael's operation in modification	46	58.9
2	Abdomen-anal resection of rectum with bringing down of functionable parts os the large intestine	22	28.2
3	Extended left colon resection	5	6.4
4	Anterior resection of rectum and sigmoid ectomy with descendostomy	3	3.8
5	Subtotal colectomy with ascendostomy and anterior resection of rectum.	2	2.5
	Total	78	100

modification and a abdominal-proctal rectectomy with bringing down in the proctal channel of functioning parts of a colon.

Apparently from the presented table, of 78 patients at 68 (87,2 %) we execute single-step radical operation, 10 (12,8 %) the patient whenever possible makes a resection hypo- or an aganglionic zone, decompensation parts of a colon and was made colostoma. In all cases at Dumael's operation the rectum stump was formed on distance by of 6-7 sm from external edge of the proctal channel by means of apparatus UO-40 or UO-60 and then in addition shortened separate seams for 1,5-2 cm, reached formations of a short stump of a rectum and prevented formation of the big blind pocket in which fecal masses could accumulate.

In the postoperative period following early complications were observed:

№	Complications	Pat.	%
1	Pyesis of a postoperative wound	6	7.6
2	Stricture of the reduced intestine	3	3.8
3	Fistula of the reduced intestine	1	1.2
4	Stricture of transversostoma	1	1.2
	Total	11	14.1

Apparently from the table the pyesis of a postoperative wound was observed at 6 (7,6 %) patients, a stricture of the reduced intestine - at 3 (3,8 %), a fistula of the reduced intestine – at 1 (1,2 %) and a stricture transversostoma - at 1 (1,2 %) the patient. These complications after corresponding treatment have been eliminated. The lethal outcome wasn't observed.

The remote results of the executed radical operations concerning illness of Girshprunga are tracked from 1 till 10 years at 57 (73 %) patients. This patient, except clinic and laboratorial researches spent endoscopic, X-ray inspections and a sphincterometry and other functional methods of research.

Results of surgical treatment were estimated on a scale of Vezik: good, adequate and inadequate.

As the good considered results of observations in which after operation there were no complaints. The general condition and functional indicators of a colon were satisfactory. Relapse of disease isn't present. Working capacity is restored.

Results were considered adequate in cases when after operation periodic abdominal pains became perceptible, sometimes an irregular chair, insufficiency of a proctal constrictor of 1 degree, but working capacity isn't broken.

As inadequate results considered cases at which constipations with implications of chronic intestinal impassability in which occasion the repeated intervention is necessary were observed.

In our observations at 46 (80,7 %) patients results were good, at 10 (17,5 %) - adequate and at 1 (1,8 %) the patient was inadequate result.

CONCLUSION

1) The most frequent reason of development the megacolon in adults is Hirschsprung's disease is characterized congenital absence or reduction of ganglions in a intramural nervous plexus in a colon. For adults short zones agangliosis localized in a rectum are characteristic.

2) For definition of type a megacolon, except radiological, functional researches of a colon, the transproctal biopsy of a wall of a rectum which is the most informative method of diagnostics of Hirschsprung's disease in adults is necessary.

3) Hirschsprung's disease in adults necessarily is subject to surgical treatment. It is referred on excision hypo-aganglionary zones and parts of decompensation of the colon.

4) At proctal or overanal forms of Hirschsprung's disease choice operation is the abdomeno-proctal rectectomy with bringing down proximal part of a colon in the proctal channel. This method of a surgical intervention gives good results in 80 % of cases.

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