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Children's Life Quality With Hirsprung's Disease After Operation De La Torre-Ortega

Mirzakarimov B.Kh., Mamajonov U.SH., Yuldashev M.A., Djumabaev J.U.
Andijan State Medical Institute, Andijan city, Uzbekistan.
Mirzakarimov Bakhromjon Khalimzhonovich https://orcid.org/0000-0003-4597-2738,bahrommirzo73@gmail.com
Mamajonov Umidjon Shokirovich https://orcid.org/0000-0002-0407-3616 ,5umidjon435588@gmail.com.
Yuldashev Muzaffar Abduvaxabovich https://orcid.org/0000-0001-9121-9611,muzaffaryuldashev44@gmail.com
Djumabaev Jurakul Usmanovich https://orcid.org/0009-0008-3744-2626,GjumaevJ@mail.ru

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Annotation.

Hirschsprung's disease (or aganglionosis) is a congenital disease in which intestinal ganglia (nerve bundles) are absent in the rectum, sigmoid and colon.

Purpose of the study: *To investigate the functional activity and well-being indicators in children diagnosed with Hirschsprung's disease who have undergone the De La Torre-Ortega procedure.*

Materials and methods: *The research comprised 89 children diagnosed with Hirschsprung's disease who underwent surgical intervention at the pediatric surgery clinic of Andizhan state medical institute between 2015 and 2022. The patients' ages ranged from 2 to 18 years;17 (19.1%) children were in the decompensated stage, 51 (57.3%) were in the subcompensated stage and 21 (23.5%) were in the compensated stage; 55 (61.8%) children were operated on using the modified De La Torre-Ortega technique, 34 (38.2%) - using the classic Soave-Lenyushkin transanal colectomy technique. Living conditions and experiences of the operated children was assessed using the PedsQL™ 4.0 scale, consisting of 21 questions evaluating physical, emotional, social, and role performance.*

Results: *Analysis of the quality of life of children with Hirschsprung's disease showed a significant increase in indicators on all scales one year after surgical treatment, while the degree of improvement was higher ($p<0.05$) when performing the improved technique of the De La Torre-Ortega operation (from an average of 62.0 -70.8% on all scales before surgery to 84.1-92.3% in relation to the group of healthy children) with a change in the total score from 52.8±10.0 to 70.4±6.4 ($t= 6.79$; $p<0.001$), in turn, transanal resection of the colon using the classical Soave-Lenyushkin method allowed achieving compliance with the group of healthy children at an average level of 76.7-84.7% with an increase in the total score only to 64 .3±4.4.*

Conclusion: *Improving the tactical and technical aspects of the surgical management of Hirschsprung's disease in children in combination with a complex postoperative rehabilitation program can improve the functional prognosis.*

Key words: *Hirschsprung's cardia, quality of life, functional activity, children*

Relevance: According to Bradnock [1], the incidence of Hirschsprung's disease in recent decades ranges from 1 in 3000 to 1 in 2000 of the total number of newborns. Average estimates put the global incidence of Hirschsprung disease at about 1 in 5,000 births. In most cases, symptoms appear in the neonatal period or early infancy [1]. Early radical intervention by removing the innervated ganglion is the best treatment for HD.

Today, one-stage radical surgical treatment using minimally invasive surgical technologies is becoming increasingly common for Hirschsprung's disease. Contemporary surgical approaches for treating Hirschsprung's disease in infants and children have the potential to decrease mortality rates and enhance long-term outcomes.

Transanal resection is currently one of the most commonly used surgical procedures for managing Hirschsprung's disease worldwide. Over the past decades, this technique has undergone a number of technological changes. However, there are still some potential complications associated with these surgeries, which can lead to irreversible functional decline and severe social limitations [8]. The most popular procedure is transanal surgery according to the Soave and Swenson method [7]. The introduction of this type of surgery has reduced the the duration of hospitalization and reduced the number of postoperative complications. However, even with this type of surgical intervention, the risk of developing various complications remains [2].

In 1998 De la Torre used one-stage transanal endorectal colectomy to treat Hirschsprung's panniculosis, avoiding open surgery [3]. However, this surgical approach is not applicable for ectopic and subplanar forms of Hirschsprung's panniculosis.

In recent years, laparoscopic surgery for HD has become increasingly popular. However, like conventional surgeries, these procedures are characterized by the possibility of postoperative complications such as constipation (7.1%-22.2%) and infection (8.9%-14.8%) [6].

In recent years, researchers have begun to investigate the treatment outcomes, rehabilitation progress, and quality of life of patients with various diseases [4]. When assessing the outcomes of surgical intervention for Hirschsprung's disease gastroenteropathy, the study of the vital volume index (VVI), which is one of the aspects of quality of life, is of particular importance.

The aim of the study: To examine the functional activity index and quality of life among pediatric patients with Hirschsprung-associated gastroenteropathy who underwent surgical intervention employing the De La Torre-Ortega procedure.

Materials and methods: The research comprised 89 children diagnosed with Hirschsprung's disease who underwent surgical intervention at the pediatric surgery clinic of Andizhan state medical institute between 2015 and 2022. The patients' ages ranged from 2 to 18 years; 17 (19.1%) children were in the decompensated stage, 51 (57.3%) were in the subcompensated stage and 21 (23.5%) were in the compensated stage; 55 (61.8%) children underwent the modified De La Torre-Ortega technique, 34 (38.2%) - the classic De La Torre-Ortega technique. Ortega. Transanal colostomy was performed using the Soave-Lenyushkin method.

To study the quality of life, children with Hirschsprung's disease and their parents completed the PedsQL™ 4.0 questionnaire (form for children and parents), translated into Uzbek, before and one year after surgical treatment [4, 5]. Due to the small number of subjects, the results of the survey of children of different ages were summarized, and the responses of parents - accordingly. In addition, 40 healthy children were interviewed for comparative evaluation.

The survey comprised 21 questions, categorized into the following domains:

- Physical functioning (PF) - 8 questions,
- Emotional functioning (EF) - 5 questions,
- Social functioning (SF) - 5 questions,
- Role functioning (RF) - Functioning in kindergarten (KF) or functioning at school (SF) - 3 questions (depending on child's age).

1. Physical Functioning (PF): This domain reflects the degree to which a physical condition restricts the ability to engage in physical activities, such as self-care, walking, climbing stairs, and lifting heavy objects.

2. Emotional functioning (EF): This domain assesses how emotional states impact work and other daily activities, such as increased time pressure, reduced workload, and decreased quality of work.

3. Social functioning (SF): This domain assesses the degree to which physical or emotional conditions restrict social activities, particularly communication.

4. Role and physical functioning (RP): This domain evaluates how physical condition affects daily role activities, including work and daily tasks. The questionnaire is segmented into age groups (5-7, 8-12, and 13-18 years), with responses provided by both the child and parent. For the age group 2-4 years, responses are solely provided by the parent. Children under 5 years old have their questionnaire completed by a parent, while those over 5 years old fill it out independently. In the corresponding questionnaires (for children and parents), children and parents are asked to choose one of the proposed answer options for each question. Total scores for all modules were calculated on a 100-point scale.

The statistical significance of the obtained measurements when comparing average values was assessed using the Student's t-test. This test calculates the probability of error (P) by verifying the normality of the distribution based on the kurtosis criterion and the equality of total variance using Fisher's test (F-test).A statistically significant change was considered to be at a significance level of $P<0.05$.

Results of the study and their discussion: When comparing quality of life indicators in the healthy group and in the preoperative critical condition group (Table 1), the overall score (OS) in the preoperative critical condition group (p/o) was (52.8±10.0) ($t=9.61$, $p<0.001$), whereas in healthy children - (79.6±6.7).

Table-1.

Quality of life indicators in the group of healthy children and in the main group before surgery

Scale	Standard values (n=40)	Main group - p/o (n=89)	t	
			Values	P
Physical functioning (PF)	84,3±9,2	52,9±11,9	8,93	<0,001
Emotional functioning (EF)	76,7±10,1	54,3±11,0	6,31	<0,001
Social functioning (SF)	83,3±7,2	51,7±10,6	10,62	<0,001
Role functioning (RF)	74,0±8,3	52,4±8,7	7,54	<0,001
Overall score (average) (OS)	79,6±6,7	52,8±10,0	9,61	<0,001

When comparing quality of life indicators in the study groups 12 months after surgery, the results changed with positive dynamics (Table 2).

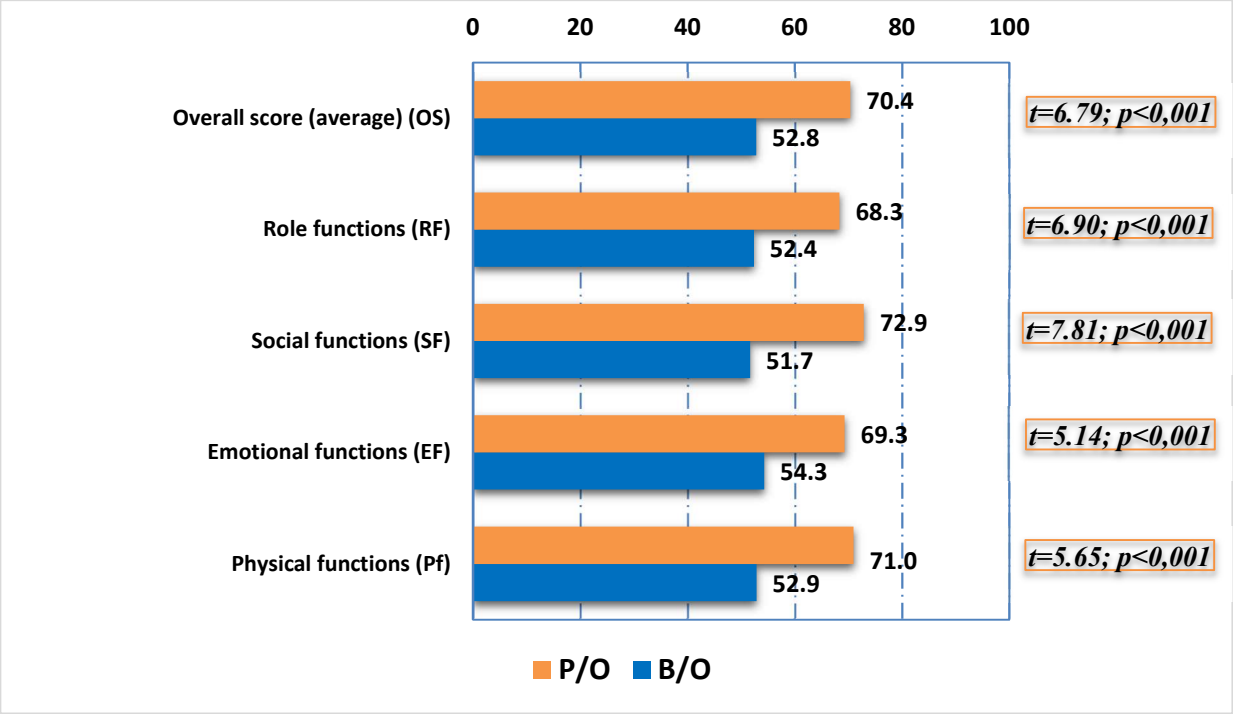
Table 2.

Quality of life indicators in the compared groups 12 months after surgery

Scale	Main group - p/o (n=89)	t к норме		Comparison group - p/o (n=40)	t compared to normal		t between groups	
		Значение	P		Value	P	Value	P
Physical functioning (PF)	71,0±8,6	4,41	<0,001	64,7±7,2	6,51	<0,001	-2,38	<0,001
Emotional functioning (EF)	69,3±7,6	2,38	<0,05	64,3±5,9	4,07	<0,001	-2,19	<0,05
Social functioning (SF)	72,9±6,4	4,48	<0,001	65,7±8,6	6,07	<0,001	-2,73	<0,01
Role functioning (RF)	68,3±6,0	2,26	<0,05	62,7±5,0	4,55	<0,001	-3,10	<0,001
Overall score (average) (OS)	70,4±6,4	4,15	<0,001	64,3±4,4	7,39	<0,001	-3,37	<0,001

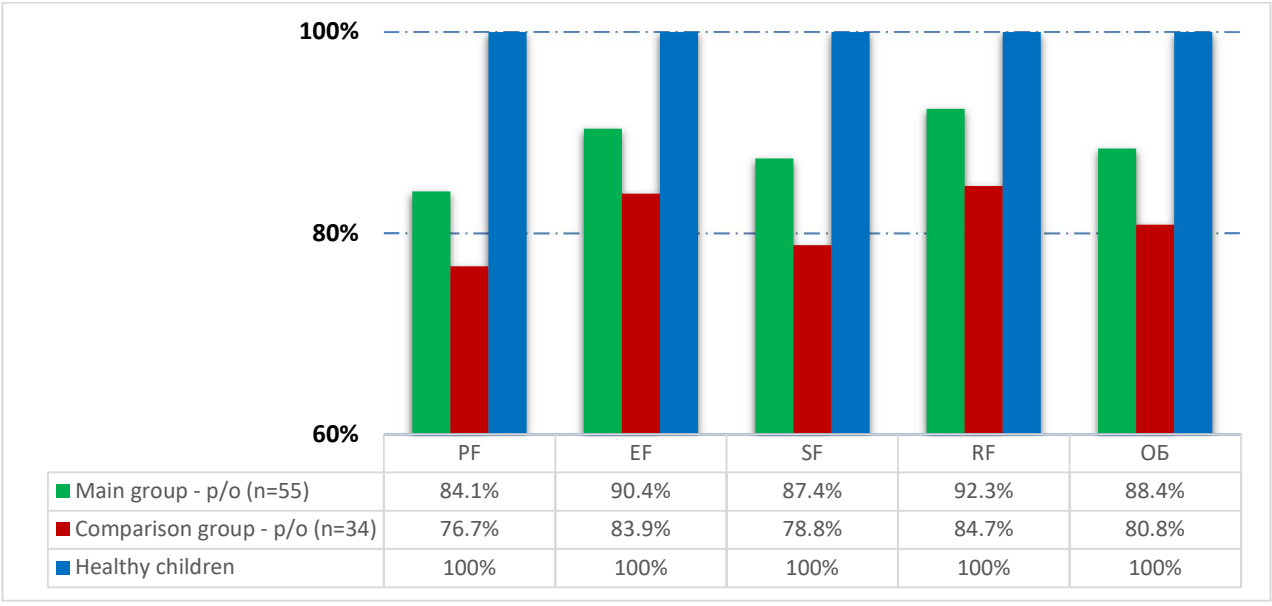
Thus, the indicators of PF, EF, SF, RF and OS in all patients of the main group improved significantly. In particular, FF in the main group after surgery was 71.0±8.6 (t=4.41; P <0.001), in the comparison group - 64.7±7.2 (t=6.51; P <0.001).

In Pic. 1. shows the dynamics of the quality of life indicator in the main group before and 12 months after surgery in children with HD. Quality of life indicators in the main group of children with HD improved: SF by 1.4 times (from 51.7% to 72.9%) (t=7.81; p <0.001), FF by 1.3 times (from 52, 9% to 71.0%) (t=5.65; p <0.001), EF (from 54.3% to 69.3%) (t=5.14; p <0.001) and RF (from 52.4% to 68 .3%) (t=6.90; p<0.001), and the OS rate increased from 52.8% to 70.4% (t=6.79; p<0.001).



Pic. 1. Dynamics of quality of life indicator in the main group before and 12 months after surgery

A more clear picture of the ratio of quality of life indicators in comparison groups 12 months after surgery to healthy children can be seen in the diagram in Pic. 2.



	PF	EF	SF	RF	ОБ
Main group - p/o (n=55)	84.1%	90.4%	87.4%	92.3%	88.4%
Comparison group - p/o (n=34)	76.7%	83.9%	78.8%	84.7%	80.8%
Healthy children	100%	100%	100%	100%	100%

Pic. 2. The ratio of quality of life indicators in comparison groups 12 months after surgery to healthy children

Thus, the ratio of quality of life indicators in children with HD in the postoperative period to healthy

children is minimized in terms of "role functioning" (92.3%), "emotional functioning" (90.4%) and "total score" (88.4%) respectively.

During follow-up periods of up to 12 months after pediatric surgical intervention with Hirschsprung's disease in the main group, there was a decrease in the frequency of functional-organic complications (anal stenosis, constipation, encopresis) from 40.4% to 18.4% ($\chi 2$=4.792; Df=1 ; P=0.029), which overall allowed an increase in the proportion of good results from 46.8% to 73.7% ($\chi 2$=7.046; Df=2; P=0.030).

An assessment of the quality of life among children diagnosed with Hirschsprung's disease revealed a substantial improvement across all metrics one year following surgical intervention, while the degree of improvement was higher ($p<0.05$) when performing the improved technique of the De La Torre-Ortega operation (from an average of 62.0 -70.8% on all scales before surgery to 84.1-92.3% in relation to the group of healthy children) with a change in the total score from 52.8±10.0 to 70.4±6.4 (t= 6.79; $p<0.001$), in turn, transanal resection of the colon using the classical Soave-Lenyushkin method made it possible to achieve compliance with the group of healthy children on average at the level of 76.7-84.7% with an increase in the total score only to 64 .3±4.4.

The use of a modified method of the De La Torre-Ortega operation in pediatric patients diagnosed with Hirschsprung's disease made it possible to improve all the main parameters for assessing the quality of the course of the immediate postoperative period ($p<0.05$ - when comparing indicators of the duration of anesthesia, restoration of peristalsis, the start of enteral nutrition and the timing of activation of patients), and also reduce the overall incidence of complications from 63.8% to 31.6% ($\chi 2$=8.743; Df=1; p=0.004) and, accordingly, the period of hospitalization from 22.1±3.2 to 15.1±5.2 days (t=7.23; $p<0.001$).

Conclusion: Improving the tactical and technical aspects of pediatric surgical treatment of Hirschsprung's disease in conjunction with a comprehensive program of postoperative rehabilitation made it possible to improve functional results, the value of which was more consistent with the normative indicators in healthy children.

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