УДК: 616.345-007.272 053.2-07-08

TACTICS FOR TREATMENT OF PAYRE'S SYNDROME IN CHILDREN

Rakhmatullaev A.A.[1], Terebaev B.A.[1], Mazhidov T.Kh.[2]

[1] Tashkent Pediatric Medical Institute, Tashkent, Uzbeistan
[2] Clinic of Tashkent Pediatric Medical Institute, Tashkent, Uzbeistan

ТАКТИКА ЛЕЧЕНИЯ СИНДРОМА ПАЙРА У ДЕТЕЙ

А.А. Рахматуллаев[1], Б.А. Теребаев[1], Т.Х. Мажидов[2]

[1] Ташкентский педиатрический медицинский институт, Ташкент, Узбекистан
[2] Клиника Ташкентского педиатрического медицинского института, Ташкент, Узбекистан

Summary. There are several views on surgical methods of treating Payre's syndrome in the literature, including Tsuman V.G. (2015) proposed to eliminate the high position and acute angle of the splenic angle by crossing the phrenic-colic and splenocolic ligaments of the colon. According to the recommendations (European Society of Coloproctology 2022), conservative treatment is recommended for this disease, and in unsuccessful cases, i.e. in the presence of persistent constipation and pain syndrome, it is recommended to reduce the splenic angle by surgical treatment. Thus, in children, the short length of the phrenic-colic ligament (Lig. phrenicocolicum sinistrum) leads to the formation of an acute angle in this branch of the colon, and when the colon is filled, it pulls the diaphragm down and causes pain under the left rib. There is very little information in the literature about Payr's syndrome in children, which means that it is necessary to conduct scientific research in this direction.

In the clinic of TashPMI. In 2018-2024, the treatment results of 83 patients with Payre syndrome aged 4 to 18 years were analyzed. Of these, 52 were girls and 31 were boys.

When analyzing the immediate and remote results of 42 patients who underwent surgical treatment, good and satisfactory results were found in 37 (88.1%) cases, unsatisfactory - in 5 (11.9%) cases. Patients with unsatisfactory results had constipation, and sometimes abdominal pain. In these 5 patients, due to concomitant dolichosigma, a conclusion was made about the unsatisfactory result and resection of the sigmoid colon was performed by minilaparotomy. After the rehabilitation measures, a good result was obtained.

Key words: *payr's syndrome, dolichosigma, diagnosis, treatment, children.*

Резюме. В литературе существует несколько взглядов на хирургические методы лечения синдрома Пайра, в том числе Цуман В.Г. (2015) предложил устранять высокое положение и острый угол селезеночного угла путем пересечения диафрагмально-толстокишечных и селезеночно-толстокишечных связок толстой кишки. Согласно рекомендациям (European Society of Coloproctology 2022), при этом заболевании рекомендуется консервативное лечение, а в безуспешных случаях, т.е. при наличии стойких запоров и болевого синдрома, рекомендуется уменьшение селезеночного угла путем хирургического лечения. Так, у детей короткая длина диафрагмально-толстокишечной связки (Lig. phrenicocolicum sinistrum) приводит к образованию острого угла в этой ветви толстой кишки, а при заполнении толстой кишки она тянет диафрагму вниз и вызывает боль под левым ребром. В литературе крайне мало сведений о синдроме Пайера у детей, что говорит о необходимости проведения научных исследований в этом направлении.

В клинике ТашПМИ в 2018-2024 гг. проанализированы результаты лечения 83 пациентов с синдромом Пайра в возрасте от 4 до 18 лет. Из них 52 девочки и 31 мальчик.

При анализе ближайших и отдаленных результатов 42 больных, перенесших оперативное лечение, хороший и удовлетворительный результат выявлен в 37 (88,1%) случаях, неудовлетво-рительный – в 5 (11,9%) случаях. У пациентов с неудовлетворительными результатами наблюдались запоры, а иногда и боли в животе. Из-за сопутствующей долихосигмы у этих 5 больных был сделан вывод о неудовлетворительном результате и выполнена резекция сигмовидной кишки путем мини-лапаротомии. После проведенных реабилитационных мероприятий получен хороший результат.

Ключевые слова: *синдром Пайра, долихосигма, диагностика, лечение, дети.*

For citation: Rakhmatullaev A.A., Terebaev B.A., Mazhidov T.Kh. TACTICS FOR TREATMENT OF PAYRE'S SYNDROME IN CHILDREN. INNOVATIVE SURGERY ON THE SILK ROAD. 2024; 2(2):21-26.

Для цитирования: Рахматуллаев А.А., Теребаев Б.А., Мажидов Т.Х. ТАКТИКА ЛЕЧЕНИЯ СИНДРОМА ПАЙРА У ДЕТЕЙ. INNOVATIVE SURGERY ON THE SILK ROAD. 2024; 2(2):21-26.

Relevance. Payre's syndrome is an increase in the length of the transverse colon and an acute angle of the splenic angle, i.e. "double-barreled shotgun", which in turn is accompanied by chronic constipation and abdominal pain. This is a congenital disease characterized by a short length of the diaphragmatic-colic ligament, the appearance of pathological adhesions, as a result of which the splenic angle increases and colodynamics is disrupted due to the acute angle. This disease first appeared in 1910. It was introduced into medicine by I. Payre [1, 3, 4, 5, 7, 8, 11,13].

There are several views on surgical methods of treating Payre's syndrome in the literature, including Tsuman V.G. (2015) proposed to eliminate the high position and acute angle of the splenic angle by crossing the phrenic-colic and splenocolic ligaments of the colon. According to the recommendations (European Society of Coloproctology 2022), conservative treatment is recommended for this disease, and in unsuccessful cases, i.e. in the presence of persistent constipation and pain syndrome, it is recommended to reduce the splenic angle by surgical treatment. Beilin N.I. (2018) according to scientific results in Payre's syndrome, pain syndrome increases with age. For this reason, it is recommended to operate on the ascending and transverse colon, without increasing the expansion and without waiting for reflux ileitis [2, 6, 9, 10, 12, 14, 15].

Thus, in children, the short length of the phrenic-colic ligament (Lig. phrenicocolicum sinistrum) leads to the formation of an acute angle in this branch of the colon, and when the colon is filled, it pulls the diaphragm down and causes pain under the left rib. There is very little information in the literature about Payr's syndrome in children, which means that it is necessary to conduct scientific research in this direction.

Objective. To improve treatment outcomes for Peyre's syndrome in children.

Material and methods. In the clinic of TashPMI In 2018-2024, the treatment results of 83 patients with Payre's syndrome aged 4 to 18 years were analyzed. Of these, 52 were girls and 31 were boys. The main contingent consisted of 42 patients aged 13-18 years. In 41 (49.4%) of these patients, the compensatory stage of the disease was determined and conservative treatment was carried out according to the "Methodological recommendations for the diagnosis and treatment of Payre's syndrome". Of the remaining 42 (50.6%) patients, 34 underwent laparoscopic correction of the acute splenic angle of the colon, and in 8 patients, the length of the transverse colon was shortened by laparotomy and an end-to-end anastomosis was performed (Table 1).

Table 1.

Distribution of patients by age, gender and type of treatment

No	Treatment method	Age of the patient				Floor		General
		0–3	4–7	8–12	13–18	M	AND	
1	Conservative	–	8	15	18	17	24	41
2	Laparoscopic	–	2	11	21	11	23	34
3	Laparotomy with resection of the transverse colon	–	–	2	6	3	5	8
General		–	10	28	45	31	52	83

Analyzing the reasons for patients' visits to the clinic, it was found that 33 (39.8%) of them had constipation as their main complaint, 36 (43.4%) had abdominal pain and constipation, and 14 (16.9%) had only abdominal pain (Table 2).

Table 2

Reasons for the initial referral of patients to hospital

Age	Number of patients	Complaints		
		Constipation	Constipation + pain	Stomach ache
0–3	–	-	–	–
4–7	10	4	4	2
8–12	28	14	8	6
13–18	45	15	24	6
Total	83 (100%)	33 (39,8%)	36 (43,4%)	14 (16,9%)

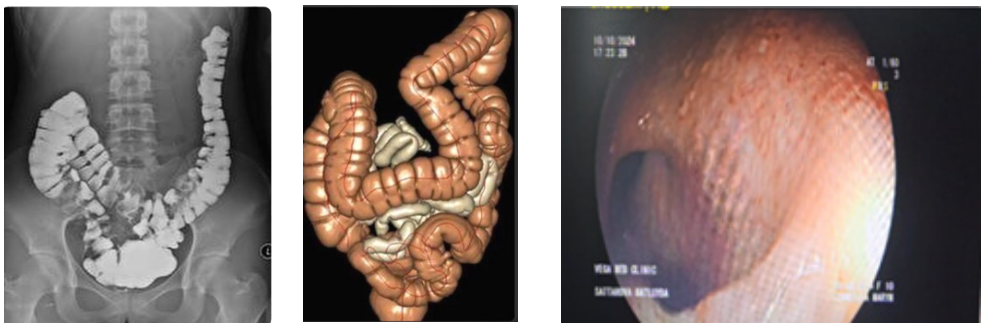

Figure 1. A – irrigation, B – virtual colonoscopy, IN – colonoscopic description of research methods

For differential diagnosis of these clinical signs observed in patients, they used special examination methods: **Dopplerography** - such parameters as the presence or absence of congestion in the mesenteric veins of the transverse colon, blood flow velocity and vascular resistance were assessed; **irrigography** - a radiopaque examination method focused on the architecture of the colon, the coefficient of change in its length after filling with contrast and after emptying the contrast, the acuteness of the angle of the spleen, changes in the location of the large intestine in a horizontal and standing position; **virtual colonoscopy** - anatomical topography of the colon was assessed by inspection; The degree of inflammation in the mucous membrane of the colon was assessed by **colonoscopy** , passing the colonoscope tube along the splenic angle, the degree of severity of the acute angle of the colon was assessed (Fig. 1).

In order to determine the method of operation, the coefficients evaluating the evacuation properties of the colon in patients with Payr's syndrome were analyzed. That is, during irrigorrhaphy, the length of the ascending part of the colon (a), the length of the transverse part (b) and the length of the descending part (c) were measured. In turn, the sum of these three lengths (d) is the entire length of the colon. The coefficient of the relative length of all three sections of the colon was determined separately (L1 (ascending part) = a/d; L1 (transverse part) = b/d; L1 (descending part) = c/d;).

The same parameters were measured after emptying the colon of contrast. That is, the length of the ascending part of the colon (a), the length of the transverse part (b) and the length of the descending part (c) were measured. In turn, the sum of these three lengths (d) is the entire length of the colon. The coefficient of the relative length of all three sections of the colon was determined separately (L 2 (ascending part) = a/d; L 2 (transverse part) = b/d; L 2 (descending part) = c/d;).

By dividing the average L1 index obtained with the colon filled with contrast by the average L2 index after emptying the colon, we determined the coefficient of increase in the length of the colon. In this way, we assessed the evacuation nature of the colon and the degree of elongation of the transverse colon, as well as the degree of " double-barreledness" (Table 3).

Table 3

Colon length coefficient values in irrigography in Payr's syndrome

Indicator	Parts of the large intestine	Result	
		Norm***	Patients
L 1 (with contrasting filling)	Ascending part**	0,21±0,03	0,22±0,04
	Transverse part*	0,41±0,04	0,48±0,02
	Descending part *	0,37±0,02	0,35±0,06
L2 (state without contrast)	Ascending part**	0,21±0,04	0,21±0,24
	Transverse part*	0,43±0,05	0,57±0,04
	Descending part *	0,35±0,04	0,26±0,05
Length coefficient	Ascending part**	1,11±0,29	1,29±0,32
	Transverse part*	1,04±0,11	0,88±0,12
	Descending part *	1,11±0,10	1,59±0,40

(* – P <0,01; ** – P >0,01. *** – Payra's disease in children (clinic, diagnostics, surgical treatment and rehabilitation) – Dorofeeva Elena Igorevna - 14.00.35 - Disease)

This allowed us to determine the method of surgical treatment of patients with Payre's syndrome by determining the colon length ratio. A length ratio below "0.8", especially in the transverse colon,

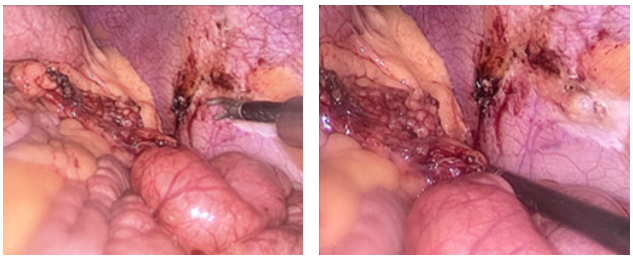

Figure 2. Laparoscopic excision of longitudinal and pathological adhesions of the ileosplenic angle of the colon in Peyre's syndrome

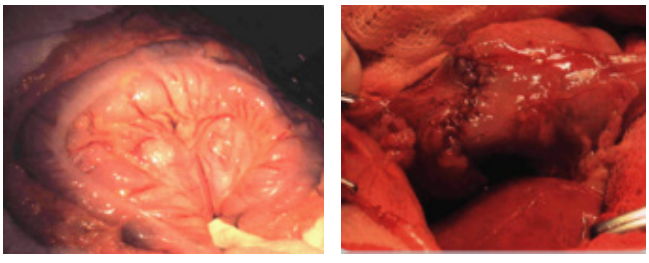

Figure 3. Transverse colon resection and end-to-end anastomosis in Peyre's syndrome

was considered an indication for direct transverse resection of the colon and end-to-end anastomosis. On the other hand, if this indicator is within "0.8-1.0", this is considered an indication for a minimally invasive laparoscopic treatment method, in which pathological adhesions of the splenic angle are excised.

In 34 patients, a high position of the splenic angle of the colon was diagnosed, a violation of the passage from this part, as a consequence, expansion of the afferent parts of the colon and reflux ileitis. Taking these changes into account, laparoscopic separation of the splenic angle of the colon from pathological scars was performed (Fig. 2).

On the other hand, in 8 patients with Payre's syndrome, the transverse colon was excessively long; the colonic arch was restored to normal by resection of the transverse colon and end-to-end anastomosis (Fig. 3).

The immediate and remote results of patients were analyzed based on the "Scale for the Analysis of the Results of Surgical Interventions for Peyre's Syndrome in Children" developed by us (UzR IMA DGU 22738 dated 02.03.2023). The following factors were taken into account: i.e. the Bristol scale by type 1 small ball-shaped hard stool, type 2 sausage-shaped hard stool, type 3 sausage-elastic stool, type 4 sausage-shaped stool with a smooth surface, intensity of abdominal pain, radiography of the colon using a contrast study method, intestinal clearance from the contrast agent as a percentage during irrigography, frequency of constipation. Depending on the presence of these factors, the results were assessed as good, satisfactory, and unsatisfactory. «-» means the absence of a factor, «+» means average frequency of observation, and «++» means constant observation. According to the sum of points of factors on this scale: 15-21 points mean a good result, 8–14 points satisfactory, and 1–7 points unsatisfactory (Table 4).

Table 4

Pediatric Peyre's Syndrome Surgical Outcome Analysis Scale

Factors		Results					
		good	**score**	**Satis-factory**	**score**	**Unsatis-factory**	**Score**
Bristol Stool Chart	Type 1 	–	3	–	2	+	**1**
	Type 2 	–	3	+	2	+	**1**
	Type 3 	++	3	+	2	–	**1**
	Type 4 	++	3	+	2	–	**1**
Stomach ache		–	3	+	2	++	**1**
Irrigography with bowel emptying from contrast		above 80%	3	60-75%	2	less than 50%	**1**
Frequency of bowel movements		Every day	3	1 time in 2 days	2	1 time in 4-5 days	**1**
General		**21**		**14**		**7**	

When analyzing the immediate and remote results of 42 patients who underwent surgical treatment, good and satisfactory results were found in 37 (88.1%) cases, and unsatisfactory results were found in 5 (11.9%) cases. Patients with unsatisfactory results had constipation and sometimes abdominal pain. In these 5 patients, due to the addition of dolichosigmoid, a conclusion was made about the unsatisfactory result and resection of the sigmoid colon was performed by minilaparotomy. After the rehabilitation measures, a good result was obtained.

Conclusion

Indications for surgical treatment of children with Payr's syndrome are considered to be the ineffectiveness of conservative treatment, as well as an increase in the frequency of abdominal pain, the development of reflux ileitis. When the patient has an acute angle in the splenic colon, but without pronounced elongation of the colon, the choice of method is laparoscopic excision of ligaments and pathological adhesions of this section. In case of excessive length of the transverse colon and the presence of congestion of the mesenteric veins, resection of the transverse colon by laparotomy and imposition of an end-to-end anastomosis is advisable.

Литература/References

1. Akilov H.A., Saidov F.Kh. Complicated forms of chronic colostasis in children in urgent surgery. // Orenburg Medical Journal. 2015. Vol. III. No. 3 (11). P. 6–7.

2. Beylina N.I., Pozdnyak A.O., Malysheva E.B. Payra's syndrome (disease). Practical medicine. 2018;16(9): 152–154.

3. Berezhnoy V.V., Kozachuk V.G., Rybalchenko V.F. Differentiated approach and treatment of chronic constipation in children with congenital intestinal malformations. // Modern pediatrics. 2016. No. 8 (80). P. 103–110.

4. Bodnar O.B., Bodnar B.M., Vatamanescu L.I., Bodnar G.B., Khashchuk V.S. Payra's disease and children: local immunity disorder and surgical treatment. // Pediatric Surgery. 2017. No. 4 (57). P. 88–91.

5. Isakova V.P., Kugasova E.A. // Electronic resource ICRON (Innovation Center for the Development of Education and Science) – 2016.

6. Galitsa V.V., Nazarova I.M., Levochkin S.P., Munasipov R.N., Frolov E.A. Experience in treating children with Payra's disease. // Healthcare of Yugra: experience and innovation. 2020. No. 3 (24). P. 18–22.

7. Druchkova S.L., Gaimolenko S.G., Mazin A.S., Sushchenko R.A. Diagnostics of chronic diseases of organic origin in children in the practice of a pediatrician. // In the collection: Actual issues of primary health care for children and adolescents. Materials of the scientific and practical conference with international participation. 2018. Pp. 36–43.

8. Muminov F.B., Kakharov M.A., Dzhuraev H.A. Comprehensive diagnostics and surgical treatment of patients with developmental and fixation anomalies in adult men and women. Coloproctology. 2019;18 (2):49-54.

9. Kamilova A.T., Ergashev N.Sh., Nurmatova N.S., Geller S.I. The cause of Payre's syndrome is chronic, and these are children. // Pediatrics. Consilium Medicum 2020. No. 2. P. 21–24.

10. Konovalova A.M., Markova M.N., Yamaliev A.T., Kulikova N.I., Kavkaev A.V., Skripitsyn N.A. Evaluation of the results of surgical treatment of Payra's diseases and childhood. // In the collection: Current issues in pediatrics: 100th anniversary of pediatric education in the Samara region. Collection of scientific papers dedicated to the 100th anniversary of the Department of Pediatric Disease. Samara, 2021. P. 57–59.

11. Makarov I.V., Zaitsev V.E., Korolev D.A., Tyumin I.A., Petrov A.V., Panarin E.A., Kudashova A.A. Clinical observations of surgical treatment of patients with Payre's syndrome. Bulletin of the Medical Institute "REAVIZ". Rehabilitation, Doctor and Health. 2023;13(4):121–126.

12. Khamraev A.J. Surgical treatment of children with acquired chronic diseases // Pediatric surgery. – 2008. – No. 1. – P. 7–9.

13. Khamraev A.J. Diagnostics and comprehensive treatment of sub and decompensated forms of chronic colostasis in children // Journal of hepato-gastroenterological research. – 2022. – no. SI-3.

14. Aliyev M.M. et al. Surgical treatment of postoperative anal incontinence in children // Central Asian Pediatric Journal. – 2019. – V. 2. – no. 1. – P. 179–184.

15. Aldamuratovich T.B., Kattaevich A.A., Ogli M.M. Analysis of the causes of postoperative anal incontinence in children // European Scientific Review. – 2018. – no. 5–6. – P. 196–199.