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CHOOSING A TREATMENT METHOD FOR PAYRE'S SYNDROME IN CHILDREN

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Abstract. Payr syndrome is characterized by elongation of the transverse colon and a high position of the splenic flexure forming an acute angle, known as the “double-barrel Payr phenomenon,” which leads to chronic constipation and abdominal pain. To improve the outcomes of treatment of Payr syndrome in children. The results of treatment of 83 patients aged 4 to 18 years with Payr syndrome who were treated at the clinic of the Tashkent Pediatric Medical Institute during 2018–2024 were analyzed. Among them, 52 were girls and 31 were boys. When analyzing the early and long-term outcomes of 42 surgically treated patients, good and satisfactory results were observed in 37 cases (88.1%), while unsatisfactory results were noted in 5 cases (11.9%). Therefore, sigmoid colon resection via mini-laparotomy was performed. After rehabilitation measures, satisfactory results were achieved. Indications for surgical treatment in children with Payr syndrome include failure of conservative therapy, increased frequency of abdominal pain, and the development of reflux ileitis. In cases where elongation of the transverse colon is not pronounced, laparoscopic descent (mobilization) of the splenic flexure is considered the preferred surgical approach.

Keywords: Payr syndrome, diagnosis, treatment, children, laparoskopiya, refluyuks.

БАЛАЛАРДАҒЫ ПАЙР СИНДРОМЫН ЕМДЕУ ӘДІСІН ТАҢДАУ

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Аңдатпа. Пайр синдромы көлденең тоқ ішектің ұзаруымен және көкбауыр иінінің жоғары орналасып, өткір бұрыш түзуімен сипатталады. Бұл жағдай «Пайрдың қос дінгекті феномені» деп аталады және созылмалы іш қату мен абдоминальды ауырсыну синдромының дамуына әкеледі. Балалардағы Пайр синдромын емдеу нәтижелерін жақсарту. 2018–2024 жылдар аралығында Ташкент педиатриялық медицина институтының клиникасында Пайр синдромымен емделген 4–18 жас аралығындағы 83 науқастың ем нәтижелері талданды. Зерттелген балалардың ішінде 52 қыз және 31 ұл болды.

Хирургиялық ем қабылдаған 42 науқастың жақын және ұзақ мерзімді нәтижелерін талдау барысында 37 жағдайда (88,1%) жақсы және қанағаттанарлық нәтижелер анықталды, ал 5 жағдайда (11,9%) қанағаттанарлықсыз нәтижелер тіркелді. Қанағаттанарлықсыз нәтижелері бар науқастарда тұрақты іш қату мен мерзімді іштің ауыруы сақталды. Реабилитациялық шаралар жүргізілгеннен кейін емнің қанағаттанарлық нәтижелеріне қол жеткізілді. Балалардағы Пайр синдромын хирургиялық емдеуге көрсеткіштерге консервативті терапияның тиімсіздігі, абдоминальды ауырсыну синдромының жиілеуі және рефлюкстік илеиттің дамуы жатады. Көлденең тоқ ішектің ұзаруы айқын болмаған жағдайларда хирургиялық емдеудің басым әдісі ретінде көкбауыр иінін лапароскопиялық төмендету (мобилизациялау) ұсынылады.

Түйін сөздер. Пайр синдромы, диагностика, емдеу, балалар, лапароскопия, рефлюкс.

ВЫБОР МЕТОДА ЛЕЧЕНИЯ СИНДРОМА ПАЙРА У ДЕТЕЙ

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Аннотация. Синдром Пайра характеризуется удлинением поперечной ободочной кишки и высоким расположением селезёночного изгиба с образованием острого угла, известного как «двуствольный феномен Пайра», что приводит к развитию хронических запоров и абдоминального болевого синдрома. Улучшить результаты лечения синдрома Пайра у детей. Проанализированы результаты лечения 83 пациентов в возрасте от 4 до 18 лет с синдромом Пайра, находившихся на лечении в клинике Ташкентского педиатрического медицинского института в период с 2018 по 2024 годы. Среди обследованных было 52 девочки и 31 мальчик. При анализе ближайших и отдалённых результатов лечения 42 пациентов, перенёсших хирургическое вмешательство, хорошие и удовлетворительные результаты были отмечены в 37 случаях (88,1%), неудовлетворительные - в 5 случаях (11,9%). У пациентов с неудовлетворительными результатами сохранялись стойкие запоры и периодические боли в животе. После проведения реабилитационных мероприятий были достигнуты удовлетворительные результаты лечения. Показаниями к хирургическому лечению синдрома Пайра у детей являются неэффективность консервативной терапии, учащение абдоминального болевого синдрома и развитие рефлюкс-илеита. В случаях, когда удлинение поперечной ободочной кишки выражено незначительно, предпочтительным методом хирургического лечения является лапароскопическое низведение (мобилизация) селезёночного изгиба ободочной кишки.

Ключевые слова. синдром Пайра, диагностика, лечение, дети, лапароскопия, рефлюкс.

Introduction. Payr syndrome is characterized by elongation of the transverse colon and a high position of the splenic flexure forming an acute angle, known as the “double-barrel Payr phenomenon,” which leads to chronic constipation and abdominal pain. This is a congenital condition associated with shortening of the phrenicocolic ligament, formation of pathological adhesions, and high positioning of the splenic flexure, resulting in impaired colonic dynamics. The disease was first described by I. Payr in 1910 [1,3,4,5,7,8,11].

The literature presents various approaches to the surgical treatment of Payr syndrome. Tsumann V.G. (2015) proposed transection of the phrenicocolic and splenicocolic ligaments to eliminate the high position and acute angulation of the splenic flexure. According to the Guidelines of the European Society of Coloproctology (2022), conservative treatment is primarily recommended, while surgical treatment with lowering of the splenic flexure is advised in cases of persistent constipation and pain unresponsive to conservative therapy. Beilin N.I. (2018) reported that pain intensity increases with age in patients with Payr syndrome and recommended timely surgical intervention before the development of significant colonic dilatation and reflux ileitis [2,6,9,10,12,13].

Thus, in children with Payr syndrome, shortening of the left phrenicocolic ligament leads to the formation of an acute angle in this region of the colon. When the colon is filled, it pulls the diaphragm downward, causing pain in the left hypochondrium. Data on Payr syndrome in children are scarce in the literature, indicating the need for further research in this field.

Objective. To improve the treatment outcomes of Payr syndrome in children.

Materials and Methods. The treatment outcomes of 83 patients aged 4–18 years with Payr syndrome who were treated at the clinic of TashPMI between 2018 and 2024 were analyzed. Of these patients, 52 were girls and 31 were boys. The main cohort consisted of adolescents aged 13–18 years, comprising 45 patients. In 41 patients (49.4%), the disease was identified at the compensated stage, and conservative treatment was carried out in accordance with the *Guidelines for the Diagnosis and Treatment of Payr Syndrome*. The remaining 42 patients (50.6%) underwent surgical treatment: 34 patients had laparoscopic correction of the acute splenic flexure of the colon, while in 8 patients laparotomy was performed with shortening (resection) of the transverse colon and creation of an end-to-end anastomosis (Table 1).

Table 1. Distribution of patients according to age, sex, and type of treatment performed

№	Treatment method	to age				sex		Total
		0-3	4-7	8-12	13 -18	boys	girls	
1.	Conservative	-	8	15	18	17	24	41
2.	Laparoscopic surgery	-	2	11	21	11	23	34
3	Laparotomy with transverse colon resection	-	-	2	6	3	5	8
Total		-	10	28	45	31	52	83

Analysis of the reasons for patients' visits to the clinic showed that 33 patients (39.8%) had constipation as the main complaint, 36 patients (43.4%) presented with a combination of abdominal pain and constipation, and 14 patients (16.9%) complained only of abdominal pain ().

Table 2. Reasons for patients' initial hospital visit

Age	Number of patients	Complaint		
		Constipation	Constipation + pain	Abdominal pain
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%)

“In order to perform a differential diagnosis of the clinical signs observed in the patients, special diagnostic methods were applied. Doppler ultrasonography was used primarily to assess the presence or absence of venous congestion in the mesenteric veins of the transverse colon, as well as to evaluate parameters such as blood flow velocity and vascular resistance. Irrigography, as a contrast-enhanced radiographic examination, focused on the architectonics of the large intestine, the coefficient of length change before and after contrast evacuation, the acuteness of the splenic flexure angle, and changes in the position of the colon in horizontal and vertical body positions. Virtual colonoscopy was performed to assess the anatomical and topographic features of the large intestine. Colonoscopy was used to evaluate the degree of inflammatory changes in the colonic mucosa and, additionally, to assess the sharpness of the splenic flexure angle based on the passage of the endoscope tip through this region (Figure 1).

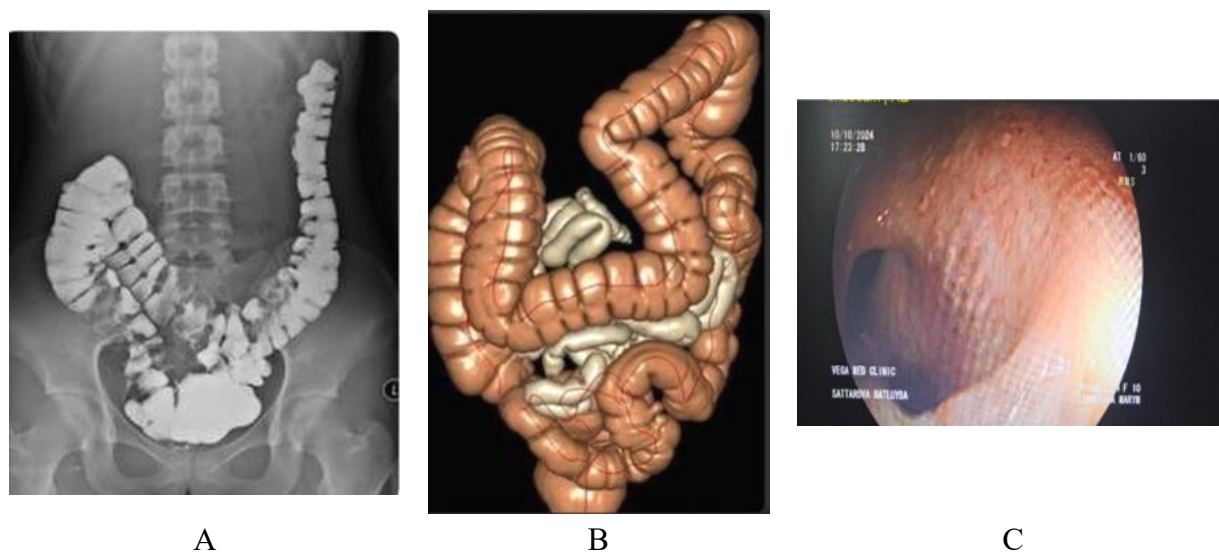
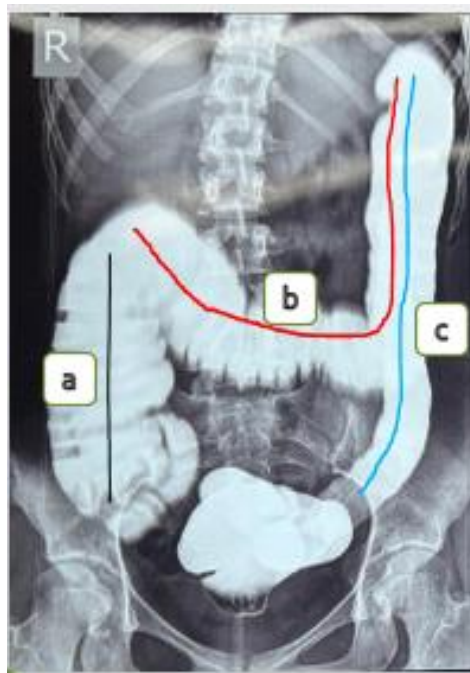


Figure 2. Imaging findings: (A) irrigography, (B) virtual colonoscopy, (C) colonoscopy

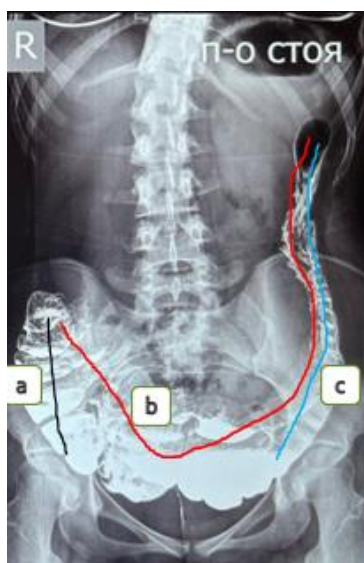
In order to determine the surgical approach, coefficients assessing the evacuatory function of the colon in Payr syndrome were analyzed. Specifically, during irrigographic examination with the colon filled with contrast medium, the length of the ascending colon (a), the transverse colon (b), and the descending colon (c) were measured. The sum of these three lengths (d) represented the total length of the colon. Subsequently, the relative length coefficients of all three segments of the colon were calculated separately ($L1$ [ascending colon] = a/d ; $L1$ [transverse colon] = b/d ; $L1$ [descending colon] = c/d) (Figure 2).



- a – length of the ascending colon (Asc. col. length)
- b – length of the transverse colon (Transv. col. length)
- c – length of the descending colon (Desc. col. length)
- d – total length of the colon
- $L1$ (ascending colon) = a/d
- $L1$ (transverse colon) = b/d
- $L1$ (descending colon) = c/d

Figure 2. Relative length coefficient of the colon with contrast-filled large intestine

Using the same parameters, measurements were also performed after evacuation of the contrast medium from the large intestine. Specifically, the length of the ascending colon (a), the transverse colon (b), and the descending colon (c) were measured. The sum of these three lengths (d) represented the total length of the colon. Subsequently, the relative length coefficients of all three segments of the colon were determined separately ($L2$ [ascending colon] = a/d ; $L2$ [transverse colon] = b/d ; $L2$ [descending colon] = c/d) (Figure 3).



- a – length of the ascending colon (Asc. col. length)
 b – length of the transverse colon (Transv. col. length)
 c – length of the descending colon (Desc. col. length)
 d – total length of the colon
 $L2$ (ascending colon) = a/d
 $L2$ (transverse colon) = b/d
 $L2$ (descending colon) = c/d

Figure 3. Relative length coefficient of the colon after evacuation of the contrast medium from the large intestine

By dividing the mean L1 value obtained with the large intestine filled with contrast medium by the mean L2 value measured after contrast evacuation, the coefficient of colonic length increase was determined. This approach allowed us to assess the evacuatory function of the large intestine, the degree of elongation of the transverse colon, and the extent of manifestation of the ‘double Payr fold’ (Table 3).

Table 3. Indices of the transverse colon length coefficient on irrigography in Payr syndrome

Indicator	Segments of the large intestine	Result	
		Normal value	Patients
L1 (contrast-filled state)	Ascending segment **	0,21±0,03	0,22±0,04
	Transverse segment *	0,41±0,04	0,48±0,02
	Descending segment *	0,37±0,02	0,35±0,06
L2 (after contrast evacuation)	Ascending segment **	0,21±0,04	0,21±0,24
	Transverse segment *	0,43±0,05	0,57±0,04
	Descending segment *	0,35±0,04	0,26±0,05
Length coefficient	Ascending segment **	1,11±0,29	1,29±0,32
	Transverse segment *	1,04±0,11	0,88±0,12
	Descending segment *	1,11±0,10	1,59±0,40

(*- $P < 0,01$; ** - $P > 0,01$)

“Determination of the colon length coefficient made it possible to select the appropriate surgical treatment strategy for patients with Payr syndrome. When the length coefficient was less than 0.8, particularly in the transverse colon, this was considered an indication for direct transverse colon resection with end-to-end anastomosis. Conversely, when this parameter ranged between 0.8 and 1.0, a minimally invasive laparoscopic procedure involving division of the splenic flexure ligament and pathological adhesions was indicated.

In 34 patients, taking into account the high position of the splenic flexure of the colon, impaired passage at this level, and the resulting dilation of the proximal segments of the colon with associated reflux ileitis, laparoscopic division of the splenic flexure of the colon from ligamentous structures and pathological adhesions was performed (Figure 4).



Figure 4. Laparoscopic division of the splenic flexure of the colon from ligamentous structures and pathological adhesions in Payr syndrome





In turn, in 8 patients with Payr syndrome presenting with excessive elongation of the transverse colon, normalization of the colonic arch was achieved by resection of the transverse colon followed by creation of an end-to-end anastomosis (Figure 5).



Figure 5. Resection of the transverse colon with end-to-end anastomosis in Payr syndrome

The short-term and long-term outcomes of the patients were analyzed using the ‘Scale for Analysis of Surgical Outcomes in Children with Payr Syndrome’ developed by our team (UzR IMA DGU No. 22738, dated 02.03.2023). The following factors were taken into account: stool characteristics according to the Bristol Stool Scale—type 1 (hard pellet-like stools), type 2 (hard sausage-shaped stools), type 3 (sausage-shaped elastic stools), and type 4 (smooth, soft sausage-shaped stools); intensity of abdominal pain; the percentage of contrast evacuation from the intestine on irrigographic examination; and bowel movement frequency. Based on the presence of these factors, outcomes were classified as good, satisfactory, or unsatisfactory. The symbol ‘-’ indicated the absence of a factor, ‘+’ indicated moderate frequency of the factor, and ‘++’ indicated constant presence. According to this scale, based on the total score of the evaluated factors, 15–21 points indicated a good outcome, 8–14 points a satisfactory outcome, and 1–7 points an unsatisfactory outcome (Table 4).

Table 4. Scale for Analysis of Surgical Outcomes in Children with Payr Syndrome

Factors		Results					
		good	Point	Satisfactory	Point	Unsatisfactory	Point
Stool form according to the Bristol Stool Scale.	type 1 	-	3	-	2	+	1
	type2 	-	3	+	2	+	1
	type 3 	++	3	+	2	-	1
	type 4 	++	3	+	2	-	1
Abdominal pain		-	3	+	2	++	1
Contrast evacuation of the intestine on irrigography		More than 80%	3	60-75%	2	Less than 50%	1
Bowel movement frequency		Every day	3	Once every 2 days	2	Once every 4–5 days	1
total		21		14		7	

Analysis of the short-term and long-term outcomes in 42 patients who underwent surgical treatment showed good and satisfactory results in 37 cases (88.1%) and unsatisfactory results in 5 cases (11.9%). In patients with unsatisfactory outcomes, constipation and occasional abdominal pain were observed. It was concluded that the unsatisfactory results in these 5 patients were due to the concomitant presence of dolichosigmoid, and sigmoid colon resection via minilaparotomy was performed. After the rehabilitation measures undertaken, satisfactory outcomes were achieved.

Conclusion. Indications for surgical treatment in children with Payr syndrome include failure of conservative therapy, increased frequency of abdominal pain, and the development of reflux ileitis. In cases where elongation of the transverse colon is not pronounced, laparoscopic descent (mobilization) of the splenic flexure is considered the preferred surgical approach. However, in cases of excessive elongation of the transverse colon accompanied by venous congestion in the mesenteric veins, transverse colon resection via laparotomy with end-to-end anastomosis is deemed appropriate.

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