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**ТИББИЁТДА ЯНГИ КУН
НОВЫЙ ДЕНЬ В МЕДИЦИНЕ
NEW DAY IN MEDICINE**

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THE MORBIDITY OF THE CHILD POPULATION WITH PECTUS EXCAVATUM OF THE REPUBLIC OF UZBEKISTAN

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✓ *Resume*

This article is devoted to the analysis of the morbidity of the child population of the Republic of Uzbekistan on funnel-shaped deformity of the chest of congenital etiology of origin. A variation series was created among 454 children with funnel-shaped deformity pathology and an analysis was carried out for each region and region of the Republic.

Key words: pectus excavatum, children, region.

ЗАБОЛЕВАЕМОСТЬ НАСЕЛЕНИЯ ДЕТСКОГО ВОЗРАСТА С ВОРОНКООБРАЗНОЙ ГРУДЬЮ РЕСПУБЛИКИ УЗБЕКИСТАН

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✓ *Резюме*

Данная статья посвящена анализу заболеваемости населения детского возраста Республики Узбекистан по воронкообразной деформации грудной клетки врожденной этиологии происхождения. Был составлен вариационный ряд среди 454 детей с патологией воронкообразной деформации и проводили анализ по каждому региону и области Республики.

Ключевые слова: воронкообразная деформация, дети, область.

ЎЗБЕКИСТОН РЕСПУБЛИКАСИ БОЛАЛИК ЁШИ АХОЛИСИНИНГ ГИРДОБСИМОН КЎКРАК БИЛАН КАСАЛЛАНИШИ

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✓ *Резюме*

Ушбу мақола Ўзбекистон Республикаси аҳолисининг туғма этиологияли кўкрак қафасининг гирдобсимон деформацияси билан касалланишини таҳлил қилишга бағишланган. Кўкрак қафаси гирдобсимон деформацияси патологияси бўлган 454 болалар ўртасида вариацион қатор яратилди ва Республиканинг ҳар бир минтақаси ва вилоятлари бўйича таҳлил ўтказилди.

Калит сўзлар: гирдобсимон деформация, болалар, вилоят.

Relevance

Pectus excavatum (PE) or funnel shaped deformity of the chest according to the international classification of diseases of the 10th revision (ICD-10) (2007) is numbered with the code Q 67.6 and is called "sunken chest" [1, 3, 5].

PE is a congenital malformation, it is a curvature of the sternum and the ribs attached to it that differ in shape and depth, leading to a decrease in the volume of the chest, compression and displacement of the mediastinal organs, causing functional disorders of the cardiovascular and respiratory systems [1, 2, 4].

According to the literature, the frequency of PE among the population ranges from 0.06 to 2.3% and this pathology accounts for 91% of all congenital deformities of the chest [3, 5]. According to international statistics, PE occurs in one case for every 400 newborns. This congenital malformation manifests itself in 30% of cases from the moment of birth, and in the remaining 70% of cases it can develop as the child grows [1, 6, 7].

A characteristic feature of PE is its progression with the growth of the child and can lead to more pronounced functional disorders on the part of the mediastinal organs, which ultimately manifests itself by decompensation of their function with possible early disability of children [8, 9].

All of the above are literary data, however, there is only undivided information from doctors dealing with pediatric orthopedic pathologies of different schools and there are no specific figures on the incidence of the population with the pathology of PE in the regional section of our Republic.

Recently, the head of our State has been carrying out large-scale and targeted measures to radically reform the healthcare system in terms of expanding the range of high-tech specialized care provided to sick children with congenital pathologies of the musculoskeletal system, which in turn will set a number of tasks for doctors to analyze the incidence rate of the population of our country with pediatric orthopedic diseases and requires improving methods diagnosis and treatment of these pathologies, including of PE.

The purpose: to determine the level of morbidity and solvability of the population with the problem of PE in the region of the Republic.

Materials and methods

We conducted a study based on the analysis of data from 454 children and adolescents with PE treated in the period from 2010 to 2018. Patient data were collected selectively based on materials from different clinics, depending on the geographical division of the Republic, such as the Ferghana Valley, Jizzakh, Navai, Bukhara regions, Khorezm and Karakalpak oases (Figure 1).

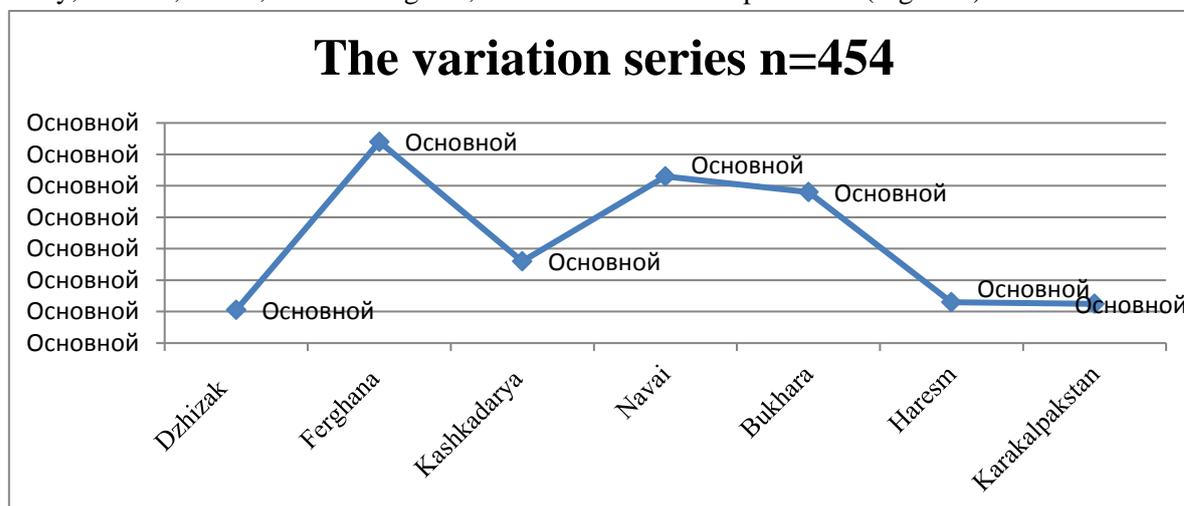


Figure 1

Diagram of the distribution of patients registered in different regions of the Republic.

Data collection was carried out using clinical materials from leading clinics of our Republic, both from the clinic of the Republican Specialized Scientific and Practical Medical Center of Traumatology and Orthopedics of the Ministry of Health of the Republic of Uzbekistan, the clinic of the Tashkent Pediatric Medical Institute, the clinic of the Andijan State Medical Institute and the clinic of the Bukhara branch of the Republican Scientific Center for Emergency Medical Care. Based on the collected materials, a variation series on the frequency of occurrence of PE has been compiled. According to Diagram 1, it can be seen that PE was registered with the highest degree in Ferghana

(n=128), Navai (n=106) and Bukhara (n=96) regions, and in small numbers it was found in Jizzakh (n=21), Kashkadarya (n=52), Khorezm (n=26) and the Karakalpak (n=25) regions, respectively.

The collected data were analyzed taking into account the following criteria, such as the age and gender of patients, the severity of PE according to the I Gyzicka (1962), the presence of syndromic pathologies in them and by type of treatment.

Results and discussions

By age, the children were distributed according to the classification of L.A. Isaeva (1987), the distribution of patients was carried out by regional section.

The data is shown in table 1

Region	Jizzakh	Ferghana	Kashkadarya	Navai	Bukhara	Khorezm	Karakalpak	Total
Age								
Till 3 y.o.	1	10	5	9	7	3	3	38 (8,4%)
3-7 y.o.	2	28	14	20	6	4	6	80 (17,6%)
8-11 y.o.	15	73	20	49	56	12	6	231 (50,9%)
12-18 y.o.	3	17	13	28	27	7	10	105 (23,1%)
Total	21 (4,6%)	128 (28,2%)	52 (11,5%)	106 (23,4%)	96 (21,1%)	26 (5,7%)	25 (5,5%)	454

Table 1 shows that, according to the severity of PE, it was recorded to a greater extent in children of the 3rd and 4th age groups (from 8 to 18 years old) in 336 (74%) cases, which are the most optimal age for thoracoplastic surgery. Before the age of 3, children with PE accounted for 38 (8.4%) of cases, which in our opinion, these patients are not recommended to undergo surgery, they should be under the supervision of doctors. At the age of 3 to 7 years, PE was observed in 80 (17.6%) cases and they were rarely prescribed surgery, only with II, III and IV degrees of severity of deformity. Regional analysis showed that in the Ferghana, Navoi and Bukhara regions, PE was higher in children aged 8-11 years. It should be noted that, by gender, PE was predominantly higher among boys in 360 (79%) cases versus 94 (21%) for girls, with a ratio of 4:1. This ratio of the occurrence of PE was once again proved by the literature data.

The severity of PE is established by the Gyzicka index (1962), determined by a lateral radiograph with the calculation of the largest retrosternal distance to the smallest, which at the same time corresponds to the I degree of this index 0.7-0.9; II degree – from 0.7 to 0.5; III degree from 0.5 to "0"; and some authors even distinguish the IV degree, which is below "0" to "-0.5", but, we used the three-power division of gravity of the PE. The data is shown in table 2.

Region	Jizzakh	Ferghana	Kashkadarya	Navai	Bukhara	Khorezm	Karakalpak	Total
Severity								
I d.	2	12	4	8	4	2	2	34 (7,5%)
II d.	11	78	30	60	50	14	17	260 (57,3%)
III d.	8	38	18	38	42	10	6	160 (35,2%)
Total	21 (4,6%)	128 (28,2%)	52 (11,5%)	106 (23,4%)	96 (21,1%)	26 (5,7%)	25 (5,5%)	454

From table 2, it should be noted that according to the severity of PE, we established the predominance of the II degree of PE in 260 (57.3%) cases, against the III degree in 160 (35.2%) cases, which were a direct indication for corrective thoracoplasty. Regional data analysis showed the prevalence of children with II and III degrees of PE in Ferghana (n=116), Navoi (n=98) and Bukhara (n=92) regions.

There were also children with grade I degree of PE - 34 (7.5%) cases, due to the small degree of deepening of the area of the birth defect, these patients were recommended to undergo a course of physical therapy against the background of active respiratory gymnastics. Younger children were under strict supervision, due to the fact that they have a period of active growth ahead and a high risk of deformity progression, although their deformity corresponded to a minor degree.

We consider it important to determine the relationship between the degree of PE and the age of the child, which makes it possible to determine the risk of progression of PE. The data is shown in figure.

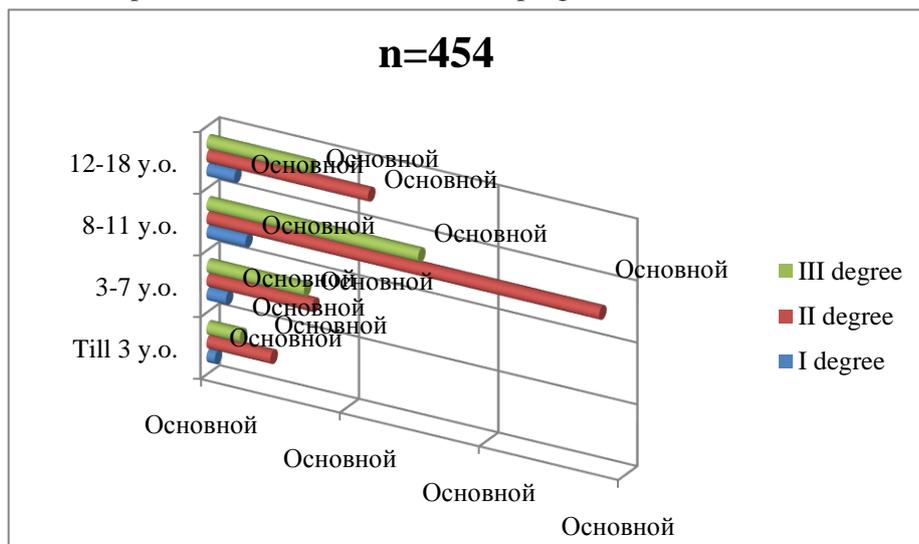


Figure 2. Diagram of the distribution of patients, taking into account the degree of PE in age gradations.

According to Figure 2, it should be noted that among patients, children with grade II of PE were predominantly more in comparison with grades I and III of this deformity. In the age groups, this congenital malformation was often observed with grade II in 141 (31%) cases aged 8-11 years, also at this age deformities with grade III were noted in 76 (16.7%) cases, and at the age of 12-18 years 58 (12.8%) cases with grade II and 37 (8.1%) cases with III degrees of PE. The risk group is under the age of 3 years, especially children with grade II in 23 (5.1%) and grade III PE in 12 (2.6%) cases.

As we know, the PE is one of the phenotypic external manifestations of syndromic pathologies, such as Marfan syndrome, Ehlers Danlo, Recklinghausen, etc. PE can be manifested in isolation or in combination with other phenotypic manifestations at the basis of syndromic pathologies. However, very few scientific papers related to the diagnosis and treatment of children with PE against the background of connective tissue dysplastic syndrome are given in the literature available to us. To this end, a phenotypic analysis was performed in 320 (70.5%) children with PE, in whom other phenotypic manifestations of connective tissue dysplasia were determined during external examination with the severity of connective tissue dysplasia.

Clinical manifestations are the state of the physique and posture of children, the correspondence of height and weight, the condition of the skin, the presence of joint hypermobility, flat feet, Gothic palate, and the intellectual development of children was assessed according to the classification of T. Milkovska-Dmitrov and A. Karakashov (1963). The following main signs were most common in children: flat feet in 125 (39%) cases, Gothic palate in 83 (26%) children, hypermobility of joints in 231 (72%) patients, the presence of thin, thin and elongated limbs (arachnodactyly) in 228 (71%) patients, pathology of the visual organs in 8 (2.5%) cases, increased extensibility and sagging of the skin in 128 (40%) cases.

Of the secondary signs, anomalies of the auricles and teeth were detected the most - in 83 (26%) cases, transient joint pain in 22 (6.9%) children, pterygodactyly in 14 (4.4%) cases. Taking into account the severity of clinical manifestations, the severity of connective tissue dysplasia was determined depending on the age groups of the distribution. Among 320 patients, the average degree of connective tissue dysplasia

was more common - in 197 (61.5%) cases, compared with mild - in 76 (23.8%) and severe - in 47 (14.7%) cases. According to the literature and our own opinion, a mild degree of dysplasia in patients with PE is considered as a separate nosology. Moderate and severe connective tissue dysplasia degrees present difficulties in choosing surgical treatment tactics in patients with PE, especially in children under 7 years of age and over 14 years of age, due to the fact that the elasticity of the thoracic frame does not correspond to age, which is confirmed by the point of view of N.I. Kondrashin (1963).

The issues of surgical correction of PE in our country have found their solution much more, that the elimination of this congenital defect is carried out by different methods of thoracoplasty in different schools. In the clinics of the Republican Specialized Scientific and Practical Medical Center of Traumatology and Orthopedics, the regional children's multidisciplinary center of the Samarkand region, the Bukhara Regional Children's Multidisciplinary Medical Center, surgical correction of PE is carried out by the D. Nuss method with its modifications, and in the clinics of TashPMI, the regional children's multidisciplinary center of the Andijan region, this congenital malformation is eliminated by the methods of Ravich Grosso and Bairov using Marshev splints and external traction device.

Of all 454 children with PE, surgical intervention was performed in 420 (92.5%) patients in whom this congenital defect corresponded to grade II and III according to the Gyzicka index. Of these, in 80 (19%) cases (comparative group), the Ravich Grosso and Bairov method was used using an external traction device (Fig. 4) and in the remaining 340 (81%) cases (main group), the D. Nuss method with its modifications (Fig. 3).

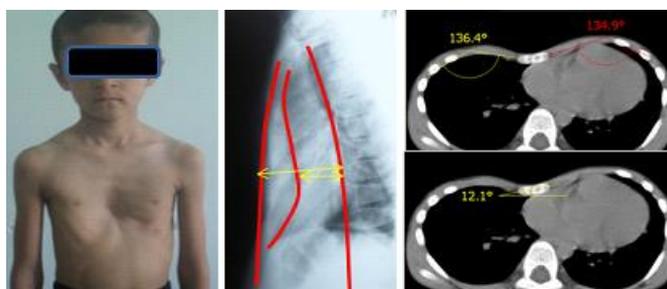


Fig. 3. a, b, c, G. Patient Z., age with PE, symmetrical appearance, grade II, appearance, X-ray and computer tomography.

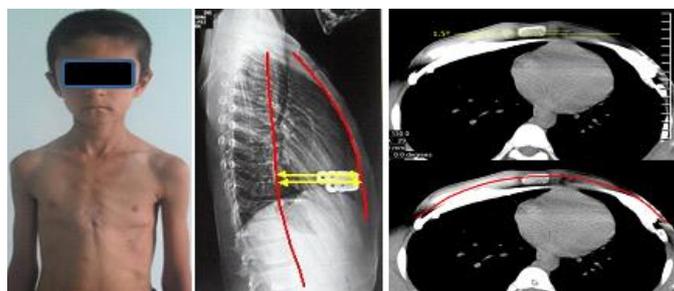


Fig. 3. d, e, w, Z. the same patient (photo after surgery – 2 years later). Condition after surgery with the best cosmetic result.



Fig. 4. a, b, patient H, 4 years old, photo before surgery, symmetrical PE, grade II, c, d, condition after surgery with unsatisfactory cosmetic result – recurrence of deformation corresponds to grade II.

The long-term results of using different thoracoplasty methods were analyzed, taking into account both cosmetic and spirometric parameters in equal numbers of children in 40 patients.

To analyze the cosmetic effect, the level of achievement of cosmetic restoration of the relief of the anterior chest wall was determined based on criteria such as the occurrence of a recurrence of the funnel requiring repeated surgery, the preservation of pathological rotation of the sternum, the appearance of a significant rough keloid scar with uneven areas (tuberosity) of the surface of the anterior chest wall.

In children of the main group using the D. Nuss method, good results were found in 34 (85%) children with the formation of an ideal anatomical relief of the anterior chest wall, satisfactory in 6 (15%) children with the development of keloid scars in the area of surgical wounds, however, cases with unsatisfactory results in children of the main group were not established. In the comparative group, children with good cosmetic results were noted in 28 (70%) cases ($p < 0.001$), with satisfactory results in 7 (17.5%) cases ($p < 0.05$) and with unsatisfactory results in the remaining 5 (12.5%) cases ($p < 0.01$), respectively.

Unsatisfactory results include cases associated with recurrence of a funnel of the corresponding II degree of severity. A comparative analysis of spirometric indicators with an assessment of the function of external respiration in children of both groups was also carried out. The recovery of the lung vital capacity index (SVC) in the main group was revealed, which was with a high degree of confidence ($p < 0.001$), which differed from the corresponding indicator in the comparative group, and the accelerated indicators with a high degree of confidence approached the norm - FVC – $70.8 \pm 2.7\%$ ($P < 0.001$).

Conclusion

It can be said that PE is one of the most frequent clinical manifestations of syndromic pathologies of the human body. A study conducted on the regional analysis of the occurrence of PE in our Republic showed that this congenital defect was registered to a greater extent in the Ferghana, Navai and Bukhara regions.

The most optimal age for thoracoplastic surgery is 8-14 years old. Based on the analysis of the long-term results of surgical treatment of PE by various thoracoplasty methods, it was found that the D. Nuss technique is the most optimal surgical method that allows ideal restoration of the anterior relief of the chest.

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