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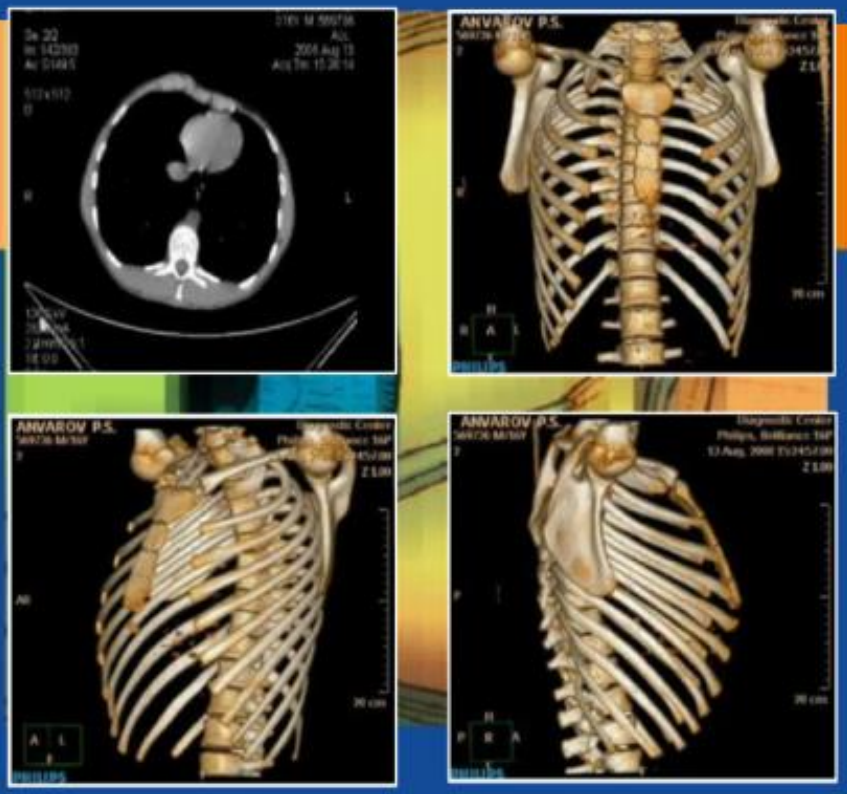
KEEL SHAPED DEFORMITY IN CHILDREN



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EDUCATIONAL MANUAL ON

KEEL SHAPED DEFORMITY IN CHILDREN

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The study manual is written based on the results of clinical research on the development and application of new, less invasive methods of operative treatment according to the clinical and anatomical form of pigeon chest deformity in children.

Pediatric surgeons, orthopedic traumatologists, as well as researchers and medical residents can use this manual. Also, the manual gives children's surgeons, orthopedic traumatologists an opportunity to learn practical skills in diseases and methods of emergency care.

The study manual was approved by the Council of the Andijan State Medical Institute.

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KEEL SHAPED DEFORMITY IN CHILDREN

C O N T E N T

Abbreviations

AP	-arterial pressure
ASA	-atrial septal aneurysm
DVI	-diastolic volume index
SI	-stroke index
SV	-stroke volume
TI	-thorax index
PCD	-pigeon chest deformity
FCD	-funnel chest deformity
CT	-computed tomography
MV	-minute volume
EDV	-end diastolic volume
ESV	-end systolic volume
USI	-ultrasound investigation
EF	-ejection fraction
CSS	-circular systolic speed
CSF	-circular systolic fraction
ECG	-electrocardiography
Echo CG	-echocardiography
HI	-heart index
HB	-heart beats
LVSO m/s.	-mean speed of left ventricular systolic output
LFQF m/q/p	- mean quality parameter of left ventricular systolic function

INTRODUCTION

A significant section of children's thoracic surgery is devoted to the problems of elimination of congenital chest deformities, which are common among children, accompanied by functional disorders in the cardiovascular and respiratory systems. Defects in the chest bones and cartilages lead to weakening of its protective function.

Cosmetic defects lead to the development of mental disorders, children become unsociable and try to avoid their peers. These aspects have a negative impact on the harmonious development and the social adaptation of these children.

The pigeon chest is the second most common deformity of the chest after the funnel deformity. This defect is considered to be a complex bone pathology and has various clinical manifestations.

It is characterized by a symmetric or asymmetric tilting of the sternum, as well as the ribs attached to it, anteriorly. This pathology is detected in 30% of patients at birth, and in almost 50% of cases, it appears at the beginning of puberty. Hereditary genesis of deformity is observed in 26% of cases. PCD is considered to be related to systemic dyschondroplasias, which often occurs along with many pathologies of connective tissue dysplasia (BTD).

In sick children, other pathologies are often not detected in functional tests, but they develop a feeling of decreased sense of fullness. In this pathology, along with a cosmetic defect, significant functional disorders in the cardiovascular and respiratory systems occur in most cases.

E.W. Fonkalsrud and S. Beanes, based on 30 years of treatment experience in patients with PCD, admit that the pigeon chest is not only a cosmetic defect.

Such types of deformities cause the sternocostal complex to be in a state of "permanent breathing" and cause functional disturbances (reduction of the vital capacity of the lungs, increase of the minute respiratory volume, decrease of the O_2 absorption coefficient, etc.) due to the restriction of the rib movement. In the conservative treatment of PCD, various external compression devices are used. However, with these devices, patients are required to walk for several years (at least 2 years), and the recurrence rate of the disease is 50%. In addition, when these technologies are used, complications such as necrosis of the chest wall skin can be observed.

Currently, methods of surgical treatment of PCD with external and internal devices or without any devices are widely used. The first two of the listed methods require re-surgery to remove the fixation devices. The last method allows to achieve a good cosmetic effect with the help of a one-time surgical procedure.

Regarding the problem of PCD, early diagnosis using modern methods, development and implementation of minimally invasive surgical methods and technologies that significantly reduce the recurrence of the disease are considered to be one of the urgent directions.

Chapter I
MODERN PRINCIPLES OF STUDYING PIGEON CHEST DEFORMITY IN CHILDREN

1.1. Prevalence of pigeon chest deformity among children.

Chest deformities in children are pathological changes in its shape, size and dimensions, which are defects that lead to shortening or lengthening of the chest-spinal distance and, as a result, disruption of the topography of internal organs. 90% are funnel deformities [7, 27, 36, 46, 70, 107]. In the second place is pigeon chest deformity and then various anomalies of the ribs, Poland and Caffey-Silverman syndromes, sternal diastasis, etc. [7, 9, 10, 27, 42, 48, 73, 87, 109, 137, 145]. In general, chest deformities occur in 1-4% of the population. Among children (mostly in boys), this rate is 0.6-2.3%, explained mainly due to cosmetic defects of the chest, functional disorders in the respiratory and cardiovascular systems [16, 81, 91, 102, 143], their mental depression . Defects in bones and cartilages lead to a decrease in the framework and protective functions of the chest, and cosmetic defects lead to the development of mental imbalances. Such children are introverted, restrained from their peers, and sometimes have an aggressive character [21, 68, 73, 89]. These conditions have a negative impact on the harmonious development of children and their social adaptation.

Therefore, the problem of chest deformities is one of the current problems of children's thoracic surgery, traumatology and orthopedics, cardiology and psychology.

Pigeon chest deformity (PCD) (syn. pectus carcinatum, "pigeon") is characterized by symmetric or asymmetric tilting of the chest and ribs attached to it anteriorly [46]. This condition can have several deformity components, with unilateral or

bilateral damage to the rib cages and forward bulging in the upper and lower parts of the chest.

In contrast to funnel chest deformity, the pigeon chest deformity is less common and accounts for 6-22% of all chest deformities [3, 6, 10, 46, 72, 116].

According to all authors, PCD is more common in boys [7, 10, 72, 110, 137, 144]. in 1987 R.C. Shamberger observed PCD in 119 (78.3%) boys and 33 (21.7%) girls [137]. K. J. Welch found it in 23 (88.5%) men and 3 (11.5%) women in 1973 [145]. On average, PCD is three times more common in boys than in girls. According to many authors, PCD, unlike pigeon chest deformity, can be detected in childhood and adolescence, even at birth or in the first year of life. This defect is detected at birth in 1/3 of patients, and in almost half of them, it begins to manifest after puberty "jump" [136, 137].

According to the results of research by K. J. Welch [145], 26% of patients with "pigeon chest" had chest deformities in other family members. As concomitant diseases, he cites kyphosis, tibia vara, arachnodactyly, talipes planovalgus, Osgood-Schlatter disease, microcephaly, diffuse hypotony. In 10 out of 26 patients, he observed diseases of the respiratory organs, and in 3, severe type of bronchial asthma. Concomitant diseases of the cardiovascular system were found in 1 out of 26 people. Data from R. C. Shamberger [137] also K.J. It is consistent with the data reported by Welch [145]. According to his observations, in 26% of these patients' family members had some type of chest deformity, 12% had scoliosis, 32 out of 152 patients (51.7%) had musculoskeletal disorders.

In PCD, the deformity of rib cages, arches and sternum is noted, and dislocation of the sternum and the ribs attached to it to the front is observed. This leads to an increase in the anterior-posterior dimensions of the chest and pigeon deformity.

In X-ray examinations, the heart rotates around its axis and has a droplet appearance along with the expansion of the retrosternal space in these patients. In radiographs taken from the side, the chest is visible as separate segments [3, 19, 92, 102].

In patients, PCD is diagnosed on the basis of anamnestic and objective data collected in each case, analysis of results of clinical-instrumental examination, as well as laboratory data.

In all sick children, chest deformity is detected at an early age, and its rate of acceleration corresponds to 11-16 years, that is, the period of intensive growth of the child. By this age, gross cosmetic defects were detected in patients, which served as an indication for operative treatment. This fact once again proves that congenital PCD is a disease of children at the age of puberty.

The main complaints of children diagnosed with this pathology are often significant shortness of breath and weakness after physical exertion, sometimes asthmatic attacks, as well as increased heart rate for no reason. Even in "asymptomatic" patients, significant changes in the cardiovascular system were observed on ECG paper. The main ECG-phenomenon was considered to be an incomplete bundle branch block (in 1 out of 4 cases). 8 (8.8%) patients had straining of the right heart chambers, giving symptoms such as chronic stasis of the pulmonary circulation.

Heart rhythm disorders were manifested as sinus bradycardia (17 cases, 18.7%), ectopic atrial rhythm (2 cases, 2.2%), pacemaker migration and Wolff-Parkinson-White syndrome (3 cases, 3.3%). Most patients (57, 62.6%) have an asthenic body type. In echocardiographic examinations, 14 (15.4%) children with hemodynamically insignificant regurgitation were found to have minor congenital heart defects (CHD) such as mitral and tricuspid valve prolapse, anomalous

position of the chords, and in 7 (7.7%) patients it turned out that there is a ventricular pseudochord.

Certain aspects of the mental state of children with PCD are also of particular note. They are usually thin, timid, shy, often avoid to bathe with their peers as they always feel unsatisfied because of a defect in themselves, and wear oversized clothes to hide the defect. These factors have a negative impact on the physical and mental state of the sick child.

According to the disease, syndromic pathologies such as Marfan (2; 2.2%), Ehlers-Danlos (8; 8.8%), Marfan-like (5; 5.5%) were identified. It has been found that in many cases, PCD occurs together with other organ and tissue diseases.

It should be noted that in these anomalies, along with the changes in the anatomical architecture of the heart and magistral blood vessels, it has been proven that it often occurs together with diseases of the musculoskeletal organs. Therefore, the coexistence of minor cardiac anomalies in children diagnosed with PCD is one of the risk factors, such as the occurrence of heart rhythm and conduction disorders with the risk of hemodynamic changes.

1.2 . Classification of pigeon chest deformity

Several classifications of PCD from a clinical-anatomic point of view have been proposed.

C. W. Lester [122, 123] proposed two forms of PCD, i.e. upper arcuate and lower curved.

H.A. Brodkin [90] also divided PCD into high - chondromanubrial prominence and - chondrogladiolar prominence forms.

F. Robichek [133, 134] divided PCD into the following types:

- symmetric - the sternum is elongated and attached to the xiphoid process at an abnormal angle, as a result of which the bulge of the sternum is visible, and the lower ribs are very deformed and are not visible due to their overlap.

- lateral deformities of the chest - the bulge of the rib arches is more obvious, and the wrong position of the chest is less visible. These deformities can be unilateral and bilateral;

- chondromanubrial deformity - only the sternal body and two rib cages protrude forward.

K. J. Welch, A. Vos [145] divided PCD into chondromanubrial or arcuate and chondrogladio or sloping deformities based on their research results. In addition, symmetric and asymmetric forms are noted.

In 1983, G.A. Bairov and A.A. Those suggested a comprehensive classification from clinic-anatomical point of view [9, 10, 46, 75, 139].

Congenital pigeon chest deformities:

I. Manubryocostal type:
1. With sunken sternal body
2. With non sunken sternal body
   a) triangular
   - symmetric; asymmetric;

II. Corporocostal type:
1. Sunk to the side
2. Non sunk to the side
   a) round shape
   b) pyramidal
   - symmetric; asymmetric;

III. Costal type:
1. With sternal rotation
2. Without sternal rotation
a) elliptical, with lateral protrusion;
- asymmetric;

Acquired pigeon chest deformities:

I. Postoperative:
1. Funnel deformity hypercorrection
2. After the operation carried out in the deviated sternum
a) in different forms
- symmetric, asymmetric;

II. Post-traumatic:
1. With a pseudo joint in the sternum
2. Without a pseudo joint in the sternum
a) in different forms
- symmetric, asymmetric;

The manubriocostal type is characterized by forward protrusion of the sternal body and 2-3rd ribs and simultaneous intrusion of the sternum.

There are two forms of the corporocostal type:

- Pyramidal - the chest is slanted in a straight line down and forward towards the xiphoid process. The maximum protrusion occurs at the joint of the middle and lower parts of the sternum.
- Round - the sternum is curved forward in its middle and lower parts.

Costal type is characterized by lateral bulging of the chest wall due to the deformed ribs. In this form, the sternum is rotated along the longitudinal axis. In this case, the ribs are bulging forward on one side and moderately or sunken on the other side, and the sternum is twisted in relation to the less

bulging side. That is why this type of deformity always occurs in an asymmetric form.

Acquired pigeon deformities can occur after surgery or injury.

G.A. Bairov and A.A. Fokin [9, 10, 75, 106] divided all PCD into symmetric and asymmetric forms, except for the costal type mentioned above. The followings can be the causes of asymmetry : unequal degree of rib bulging on both sides, chest rotation along the longitudinal axis, and the difference in the number of ribs involved in the deformity on the right and left sides.

According to the changes in dynamics, these authors also divided the deformity into types that decrease over time, grow in parallel with the child's age and development, and surpass the child's age and development.

All authors point out that sternochondral form is the commonest deformity in clinical practice [7]. It is characterized by symmetric bulging of the lower ribs and sternum on both sides. The upper or "manubriocostal" type is relatively less common.

According to some authors [7, 93, 98, 114], PCD is only a cosmetic defect, and it is not associated with dysfunction of the thorax organs.

N.I. A number of authors, such as K.U. Ashcraft, T.M. Holder [7], Kondarshin [48], consider [93, 98, 114, 116] PCD to be only a cosmetic defect, since no deviations from age-related norms were observed in functional examinations. However, many authors [19, 92, 95, 135, 144] believe that functional disorders occur due to a decrease in the decreased mobility of the chest as the child ages.

G.A. Bairov and A.A. Fokin [9, 10, 75, 106] point out that these patients’ chests are in the position of maximum "breathing", and due to the reduction of the respiratory

excursion, bronchopulmonary diseases are often more common. In addition, lethargy, profuse sweating, difficult breathing through the nose, and the presence of adenoid hypertrophy are also observed in this category of children. Of course, congenital predisposition to the respiratory diseases also plays an important role.

According to these authors, 1 out of every 6 children with PCD has chronic pneumonia, topographic or developmental disorders of the bronchopulmonary system (polycyst, hypoplasia, lobar emphysema, bronchiectasis). In addition, these authors also observed cardiac pathologies such as ventricular septal defect, systolic murmur, sinus tachycardia, and various forms of arrhythmias in 6 of 17 patients. K.J. Welch [145] explains the last results in the long-term presence of PCD as follows: the anterior-posterior diameter of the chest increases, and the ineffectiveness of respiratory movements appear due to its low mobility. These movements are now performed at the expense of the diaphragm and accessory muscles. As the lung capacity continues to decrease, progressive emphysema and lung infection develop. S.F. Robichek called such a chest "frozen" [133, 134]. He points out that in PCD, gradual worsening of deficiency symptoms can lead to shortness of breath, asthmatic attacks and tachycardia. Even "asymptomatic" patients can develop early emphysema, pneumothorax, and cor pulmonale. Californian E.W. Fonkalsurd and S. Beanes [112], based on the experience of treating 90 patients with PCD during 1970-2000, proved that this pathology is not only a cosmetic defect, but a disease with serious functional changes. 85 out of 90 patients had decreased lung capacity (94.4%), respiratory disease (54%), bronchial asthma (26.6%), chest pain and discomfort (42.2%), lung changes requiring additional oxygenation (2.2%).

In PCD, the chest is partially enlarged, with emphysematous expansion, which in turn reduces its elasticity and tolerance to stress [40, 52, 53, 128].

Psychological aspects of deformity are also not less important. According to S.F. Robichek [133, 134], patients with pectus carcinatum are thin and shy. They always walk and sit in a stooped, slouched position to hide their flaws. During school, they become an object of ridicule, avoid to participate in the pool and various activities. A cosmetic defect leads to dehumanization, and sometimes to extreme aggression [21, 73]. All these factors lead to deterioration of mental and physical condition. In 22 out of 26 patients with PCD (84.6%), shyness towards the deformity, self-doubt, inability to undress, disgusting towards the bulge in the chest and a strong emotional reaction were observed [145].

1.3. Etiopathogenetic mechanisms of the development of pigeon chest deformity in children.

Despite the existence of many assumptions and theories, there is still no single opinion about the origin of PCD. There are many assumptions that the development of this disease can be associated with conditions such as rickets, respiratory obstruction, high intrauterine pressure during pregnancy [137], imbalance between chest strength and diaphragm traction [90].

In addition, there are theories about the changes in the quantitative and qualitative composition of collagen [12, 54, 55], glycosaminoglycan and water, which lead to a decrease in the strength of the rib cages. According to many scientists, the reason for the development of PCD is chondrodysplasia of the rib cages, and this condition is manifested by rapid growth of the ribs [52, 68, 145]. Although the theory that rib cage overgrowth leads to PCD is supported by many scientists, the

exact cause remains unknown, and the results of studies contradict each other. For example, T. Nakaoka [127] and his co-authors point out that the children's ribs with PCD are not longer than those of healthy children.

Some authors consider PCD to be one of the phenotypic manifestations of connective tissue dysplasia (BTD) [2]. The term "dysplasia" refers to the abnormal growth and development of an organ or tissue. The diagnosis of connective tissue dysplasia is based on a thorough analysis of symptoms or clinical studies. Nevertheless, in clinical practice, this diagnosis is combined with the main disease if it is not clearly confirmed histologically. Therefore, clinically detected dysplasia can be the result of many changes in tissue structure [75].

Connective tissue dysplasia is a genetically determined process, which is based on the mutation of the genes responsible for the synthesis of collagen fibers. As a proof of this, it can be noted that scoliosis with connective tissue anomalies and Marfan's syndrome are accompanied with deformities of the anterior chest wall in most cases [137]. In probands with PCD, the presence of an aggravating family history is 37% [137] indicates that this pathology is genetically determined. As a result of the mutation, the initial three-helix conformation of the collagen macromolecule is disturbed, and its stability decreases. Fibrils and fibers are formed defectively, fibrous structures cannot withstand mechanical stress.

J.Feng and his co-authors' draw particular attention to the comparative histochemical, morphological, and biomechanical characterization of rib cages in PCD [105]. As a result of the research of these authors, it was proved that the roughness of the rib cages (Young's modulus, compression and displacement) the strength quality decrease when they are stretched, crushed and displaced.

Another area of research focuses on the study of the chondrocytes structure and intercellular substances in PCD and in the norm. Some researchers believe that hyperplasia of the rib cages is the basis of the formation of PCD[2, 33], while other group of authors state that there is no sharp difference between the chondrocytes of PCD and the control group [12, 99, 105].

According to G.Melean [126], biomechanical disorders of the ribs are secondary, the main cause of which is the violation of chondro- and osteogenesis.

Rib cages in patients with PCD have blood vessels in all sections. The amount of blood vessels is the same in deformed and normal rib cages. The amount of chondrocytes in one chondron increases dramatically with age compared to the control group.

J. Feng and co-authors [105] reported that the above data had not been confirmed. According to them, chondrocytes and their nuclei are intact [105]. Also, no signs of hypo- or hyperplasia of rib cages were detected in electron and transmission microscopy. The amount of blood vessels in the children' ribs with PCD does not differ from that of healthy children [34]. The claim that the number of chondrocytes in one chondron increases dramatically with age has not been confirmed. According to J. Feng and co-authors [105], the number of cells does not change.

E.A. Bardakhchyan and co-authors [12] put forward the opinion that the morphological substrate of chest deformity is the disturbance of chondrocytes, intercellular substance and fibers at an ultrastructural level. Changes in chondrocytes in PCD are mainly associated with the development of dystrophic process - fatty and carbohydrate dystrophy, formation of atypical asbestos-like fibrils in pericellular zones. In this case, almost no organelles remain, and those that can be identified are functionally disproportional.

V.M. According to Kuritsin and his co-authors [50], the common morphological features of the rib cage in normal and PCD are the same, and are acellular, map-like areas, desquamated chondral fibers and "brain-like" spaces. However, in PCD they develop 6-7 years earlier and are detected 3 times more than in healthy children.

Thus, despite the fact that there are many studies and scientific publications devoted to the study of the structure and characteristics of the rib cages in the norm and in PCD, the causes and pathogenesis of PCD still remain unknown. It is also worth noting that most of the research on rib cages was done decades ago.

E.A. Abalmasova, E.V. Luzina [43] have focused on the fact that heredity is the leading factor in the origin of congenital chest deformity. According to them, these pathologies are a congenital systemic process. This opinion is supported by the fact that chest deformities are often found in Marfan syndrome, dysraphic status, and neurofibromatosis [43].

F.L. Westphal [146] and co-authors report that chest deformities are caused by as yet unidentified genetic disorders. When they examined 1332 Brazilian schoolchildren, they found that 26 (1.95%) had FCD (1.275%) and PCD (0.657%). The relatives of 17 (65.4%) of these students also had various chest deformities.

Currently, there are two expert systems, POSSUM (Pictures Of Standard Syndromes and Undiagnosed Malformations) and WBDD (Winter-Baraitser Dysmorphology Database), which are widely used for the diagnosis of rare genetic syndromes worldwide [30, 31, 35].

The results of examination in these systems showed that 39 patients with chest deformities had monogenic syndromes, 8 had digital chromosome aberrations, and 44 had systemic chromosomal rearrangements.

FCD and PCD occur as part of many hereditary syndromes and diseases associated with sporadic gene mutations. Chest deformities are most commonly seen in Noonan and Marfan syndromes [61, 67, 118, 119, 129, 142].

Marfan syndrome (MS) is an autosomal dominant connective tissue disorder [18, 118] with a population prevalence of 1:3000-10000. MS is caused by mutations in the fibrillin-1 gene. This gene mutation causes tubular bones to grow rapidly. In addition, characteristic features of MS in the musculoskeletal system (Marfan-like phenotype): dolichostenomelia, dolichocephaly, arachnodactyly, joint laxity, scoliosis, chest deformities, high palate, dental anomalies. A left anterior congenital diaphragmatic hernia (Morgagni-Larrey) thought to be the cause of bowel obstruction in a patient with MS.

Noonan syndrome (NS) is a heterogeneous genetic disease characterized by postnatal height reduction, characteristic facial dysmorphism, heart defects, and various psychosomatic disorders. In addition, there may be ectodermal and bone system defects, cryptorchidism, lymphatic dysplasia, tendency to bleeding, and in rare cases, malignant tumors of the blood system. The incidence rate is 1:1000-2500. Noonan syndrome is caused by mutations in PTPN11, SOS1, KRAS, RAF1, BRAF and MEK1 (MAP2K1) genes [142]. Orthopedic changes include FCD, PCD, spine deformity, elbow joint valgus deformity [13, 17, 23, 25, 26, 34, 41, 140].

Thus, there is a lot of information about various genes and hereditary diseases that cause chest deformities. Although breast deformities are the main component of hereditary syndromes and chromosomal disorders, the genetic causes of this pathology in most patients remain unknown. Also, the causes of sporadic cases of FCD and PCD are not clear. These scientific points of view require a thorough study of the genes and their changes that are involved in the origin of breast deformities.

Chapter II
MODERN APPROACHES TO DIAGNOSTICS OF PIGEON CHEST DEFORMITY IN CHILDREN

2.1. Modern methods of examination of the thorax and intrathoracic organs

Indications for thoracoplastic surgery in PCD are not the changes in the cardiovascular and respiratory system, as in the case of the funnel chest deformity, but the progressing defect, the bulge, in the chest, and this surgical procedure is only cosmetically noteworthy. Failure to detect this disease in time and in full will cause deterioration of the general condition of sick children and early disability. However, the research results of a number of authors show that the occurrence of chest deformity is influenced by the time of formation of the birth defect, all features of the fetal period. In addition to the local deformative changes in PCD, the processes leading to the distortion of the stature associated with the curvature of the spine are also manifested. Distortion of the stature causes disturbances in the functioning of the internal organs with a decrease in the vital capacity of the lungs, intrathoracic pressure, and the excursion of the chest and diaphragm [7-9]. These changes, in turn, have a negative effect on the cardiovascular and respiratory systems, leading to a decrease in their physiological reserves, as a result of which the body's adaptive capabilities are impaired, and the quality of physical health and endurance is reduced.

At the same time, the literature does not provide specific recommendations for the use of examination methods in children with PCD, because the etiological causes of this pathology depend on many factors, the coexistence of additional

anomalies and birth defects, and often the presence of connective tissue dysplasia. Therefore, it is appropriate to discuss the clinical and functional changes in children diagnosed with PCD due to the fact that the conditions and principles of examination in children belonging to this group are different, and there is no uniform program and recommendations for their examination.

Currently, there are no approved universal standards for the diagnosis of chest deformities [3, 14, 29, 51, 77, 79, 96, 97, 120]. Many authors think that there is a need to use computed tomography (CT) and magnetic resonance imaging (MRI) in order to accurately diagnose the form and degree of deformity, determine indications for thoracoplastic surgery, calculate the optimal volume of resection of deformed ribs and thoracoplastic surgery, as well as evaluate the shape of the chest before and after examination. Chest index (KI), which determines the ratio of the anterior-posterior and transverse dimensions of the chest, as well as parameters such as volume index of deformity, chest size, heart compression index, are used more often.

Also, there is a method of x-ray imaging of the spine in the direct projection, as well as in the lateral projection, while the patient is standing, and it is possible to determine the degree of deformity with the help of the x-ray images taken in the lateral projection. According to the roentgenogram, the Louis angle (the angle formed between the sternal manubrium and body, in °) is found to determine the deformity degree of the anterior chest wall (Fig. 2.1).

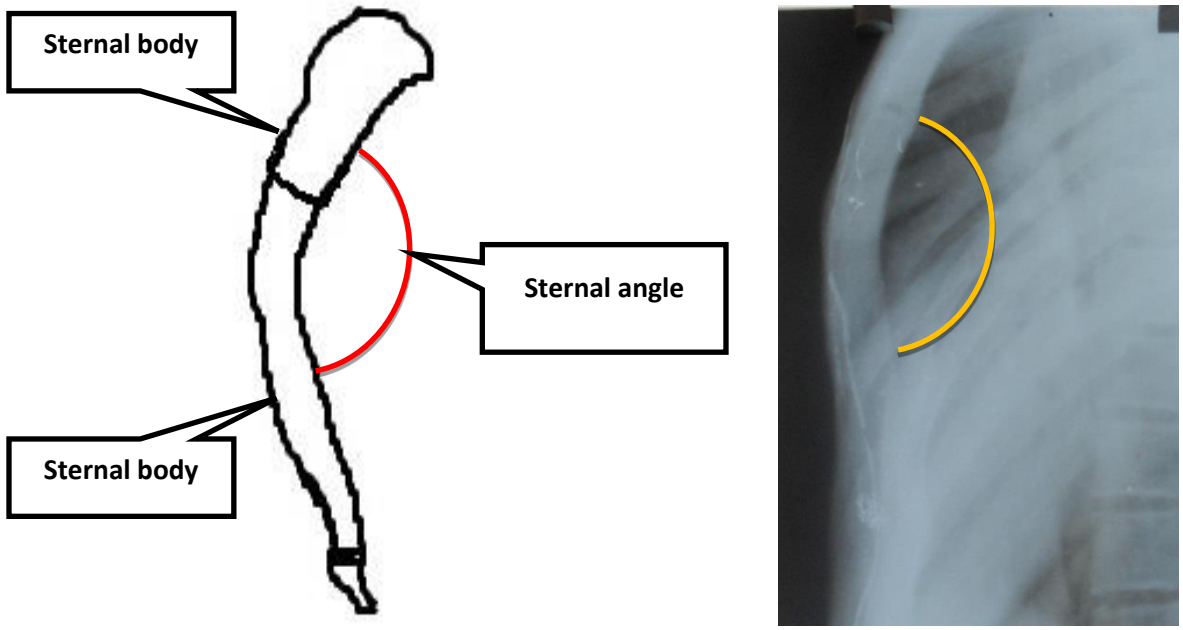

Figure 2.1. Louis angle determination scheme

Depending on the sharpness of this angle, 3 levels of pigeon deformity are distinguished. (Table 2.1).

Table 2.1

Degrees of pigeon deformity according to the Lois classification (°)

1	Degrees of chest deformity	I st Degree	II nd degree	III rd degree	Normal values
2	Louis angle	145-130°	130-115°	115° and <	175-145°

According to special instructions, a multispiral computed tomography is performed on a Philips, Briliance 16P

(Netherlands) computed tomography. This instrumental examination method can provide an opportunity to dynamically study the changes in chest deformity, and also determine the extent of operative treatment through 3D reconstruction (Fig. 2.2).

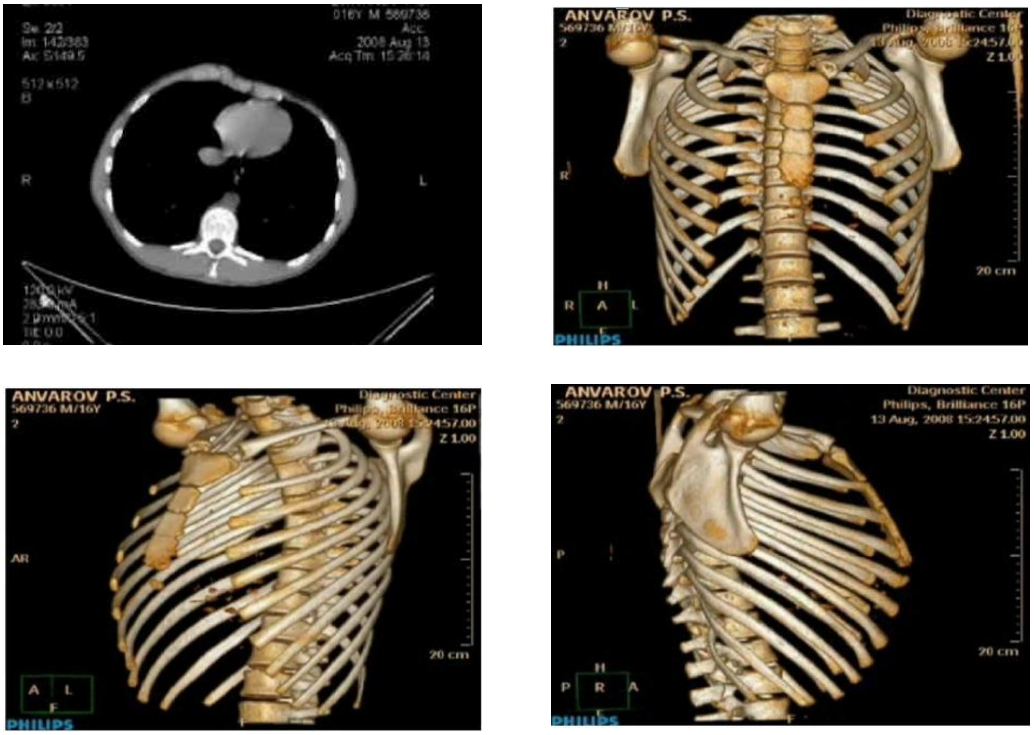

Figure 2.2. MSCT. Forward bending of the rib cage and forward expansion of the anterior-posterior dimensions of the ribcage. 3D reconstruction of the chest.

"Equipment for detecting chest deformities in children" was patented as an invention by the staff of the Children's Surgery and Orthopedics Clinic of Rostov State Medical University [78, 79, 80]. This equipment consists of two plates with a handle, which is made up of freely moving measuring metal rods with scales. Measurements with the help of the equipment are carried out as follows: the patient lies on a hard plane. The measurement is made segmentally from the III rd pair of ribs to

the VIII th pair of ribs. The number of segments can change depending on the duration of the deformity of the anterior chest wall. The obtained measurement results are submitted into the computer, and graphical images of the measured segments are obtained using a specially developed computer program.

Joint study of chest and spine deformities allows to determine the dynamics of pathobiomechanical changes, their influence on respiratory and cardiovascular systems activity.

Researchers of the Novosibirsk Institute of Traumatology and Orthopedics have proposed a computerized optical topography method for diagnosing pathologies of the anterior chest wall in order to determine indications for the operative treatment of chest deformities in children and adolescents and to monitor their dynamic changes. This method is based on taking optical measurements of the body surface. Body surface shape information depicts a graphical representation of the chest relief in the form of isolines. Such a topographic view is used to analyze the shape of the chest deformity along with obtaining general information about the surface relief. This makes it possible to avoid the X-ray examination method, which has a high radiation risk in children and adolescents in most cases. With this method, it is possible to obtain information about the shape and type of the anterior wall of the chest in a correct and high spatial resolution in the sagittal section.

In addition, this method is harmless to the patient, and it is characterized by the fact that it is much better than radiological examinations, as well as conducting screening examinations and creating a database.

O.A. According to Sudeykina [72], X-ray examinations of pigeon chest deformities do not always allow to determine the type of deformity. Laser stereolithography is the most informative method.

This method of examination allows not only to identify anatomical disorders, but also to determine the optimal methods of resection and osteotomy of deformed ribs in advance. With the help of this technology, it is possible to obtain a plastic copy of the object according to the computerized image of the object using liquid photopolymerizable compositions and a laser. With the help of stereolithographic biomodeling based on computed tomography data, it is possible to visually see the form of deformity (chest width, depressions, shifts, etc.) in the plastic copy of the deformed chest wall and implement the method of surgical correction provided for in this model. The disadvantage of this method is its cost and the impossibility of performing it in ordinary medical institutions.

2.2. Methods of anthropometric examination

In chest deformities, anthropometric methods can be effectively used to evaluate its shape, development rate, effectiveness of rib resection and reconstructive thoracoplasty performed [77].

Numerous sources of literature [11, 43, 44, 59, 61, 74, 82, 83, 84, 115, 139] show that connective tissue dysplasia (CTD) is of great importance due to its prevalence, its various manifestations, severity, and lack of timely diagnosis. Until now, there is no generally accepted diagnostic algorithm of connective tissue dysplasia syndrome (CTD). This prevents timely detection of pathological aspects associated with this condition [15, 28, 38, 124,125, 138].

During their work, doctors often diagnose connective tissue dysplasia as an independent disease or syndrome [15, 82]. From a clinical point of view, visceral, skin and musculoskeletal changes of organs and tissues caused by dysplasia are distinguished [38]. At the same time, the research materials of

many authors confirm the occurrence of various changes in musculoskeletal organs related to dysplastic changes [52].

A number of dysplastic diseases of the musculoskeletal system are characterized by a poor outcome and a severe course. However, researchers often consider musculoskeletal dysplasia as external markers of connective tissue deficiency, and complex clinical-instrumental examinations are not conducted in this regard. According to some researchers, [33, 74] the patterns revealed as a result of a comprehensive study of the orthopedic status of children with pigeon chest deformity are important in the diagnosis of connective tissue dysplasia.

In addition to the local deformative changes in PCD, the processes leading to the distortion of the stature associated with the curvature of the spine are also manifested. Distortion of stature is the cause of internal organ dysfunctions with a decrease in lung capacity, intrathoracic pressure, and chest and diaphragm excursions. These changes, in turn, have a negative impact on the cardiovascular and respiratory systems, leading to a decrease in their physiological reserves, as a result, a violation of the body's adaptive capabilities, a decrease in the quality of physical health and endurance.

A number of dysplastic diseases of the musculoskeletal system are characterized by a poor outcome and a severe course. However, researchers often consider musculoskeletal dysplasia as external markers of connective tissue deficiency, and complex clinical-instrumental examinations are not conducted in this regard.

Therefore, there is a need for a comprehensive study of the orthopedic status of children with pigeon chest deformity and to evaluate the significance of the identified patterns in the diagnosis of connective tissue dysplasia.

Examinations included patient complaints, vital and genetic anamnesis, objective vision, somato- and anthropometry, as well

as radiographic data. Since there are no official diagnostic criteria for connective tissue dysplasia, the external signs and criteria of dysplastic changes, which are clinically and instrumentally convenient, are used.

Children's height was checked in the traditional way, that is, a vertical line passed through the occipital protuberance on a sagittal section, the vertebra prominence, the crease between the buttock muscles and the base quadrangle (the area occupied by the paired feet and the space between them). All patients with PCD have diagnostically significant external dysmorphogenetic signs, which proves the presence of CTD in these children.

Analysis of anthropometric measurements shows that children with CTD are underweight. In addition, in children of the main group, in contrast to the control group, a higher body length, lower Ketle's index, and less than the length of the II nd finger of the circumference of the wrist-palm joint are determined.

Special examinations, such as goniometric measurements and the study of external dysembryogenetic stigmas, were carried out in all patient children in the main group. In order to determine the sternovertebral distance and the deformity type in the examined children of this group, chest X-ray performed in a lateral projection.

It is worth mentioning that scoliosis and flat feet are the most common and occur almost equally. This shows that the prevalence of these pathologies is much higher than the average. However, according to some researchers, the prevalence of scoliosis in the population is 3.6%, and flat feet is 7.8% [9]. The results of the study of the nosological forms diagnosed by us among the pathologies of the musculoskeletal system are presented in Table 2.3.

Table 2.3

Associated musculoskeletal diseases in children with pigeon chest deformity

№	Nosologic forms	Deformity degree				Abs	%
		I	II	III	IV		
1	**Scoliotic spine deformity**						
	Mixed	3	1	1	-	5	8,1
	Thorax	4	2	1	-	7	11,3
	Lumbar	1	1	-	-	2	3,2
	Total	8* (57,1%)	4(28,6%)	2(14,3%)	-	**14**	**22,6**
2	**Flat feet**						
	Vertical	3	6	2	1	12	19,3
	Horizontal	1	-	-	-	1	1,7
	Total	4(30,8%)	6*(46,1%)	2(15,4%)	1(7,7%)	**13**	**21,0**
	With talonavicular joint arthrosis					22*	35,5
	Without talonavicular joint arthrosis					30*	48,4
3	**Stature disorder**					**20**	**32,2**
	Scolioitc					14	22,6
	Kyphotic					2	3,2
	Other					4	6,4
4	**Degenerative-dystrophic diseases of the spine**					**50***	**40***
	Polysegmental osteochondrosis					18	29,0
	Spondylosis					5	8,1
	Intervertebral disc hernia					2	3,2
	Dysplastic coxarthrosis					**4**	**6,4**

It is worth noting that in children with the first degree scoliosis, there was a slight lateral deviation of the spine and torsion at the initial stage, which could be determined only by

X-ray examination. It is known that detection and systematic treatment of scoliosis in the earliest stages of its development can prevent the acceleration of the spine curvature [2, 7]. The obtained results confirm the necessity of X-ray examinations in order to start orthopedic treatment measures in time in patients with musculoskeletal disorders accompanied by connective tissue dysplasia. It is known from the literature that the weakening of the musculoskeletal system associated with CTD is one of the main etiopathogenetic aspects of the spine curvature and the flat feet formation. This process in the vertebrae, joints, and muscles leads to stature disorder[7].

Recently, a significant "rejuvenation" of degenerative-dystrophic changes such as osteochondrosis and spondylosis of the vertebrae has been observed [5]. Polysegmental osteochondrosis was found in 18 (29.0%) children, spondylosis in 8.1% (5 cases) and intervertebral disc herniation in 3.2% (2 cases). Hip dysplasia of various degrees was found in 6.4% of cases.

In many cases, it became clear that the above-mentioned orthopedic pathologies meet in a polfocal form. That is, the co-occurrence of scoliosis and vertical flat feet was found in 29.0% of patients, stature disorder and flat feet in 25.8%, scoliosis and polysegmental osteochondrosis in 12.9% of children.

In the literature, special attention is paid to the injury characteristics of patients with CTD [3, 9]. It has been shown that the more obvious the symptoms of CTD during any dynamic and static stress, the higher the degree of injury, while there is a correlation between the intensity of stress and the injury index. [3]. According to the data obtained in the ASMI, 38.7% of the children's anamnesis revealed that they had suffered injuries such as tubular bone fractures of various degrees, while in the control group, only 4.5% of the subjects had this condition.

In conclusion, it can be noted that the obtained data prove that there is a clinical-functional connection between the pathologies of the musculoskeletal organs associated with congenital pigeon chest deformity and concomitant connective tissue dysplasia. The laws taken from the analysis of the cited correlative relationships are of great importance in the complex diagnosis of connective tissue dysplasia syndrome.

2.3. Methods of electrophysiological examination of the cardiovascular system in children with clavicle deformity of the chest

According to the literature, changes in the cardiovascular system accelerate as a result of timely and inadequate elimination of chest deformities [24]. At the same time, researchers have not specified the main and additional diagnostic criteria that determine the exact changes in the cardiovascular system that cause the decompensation of children in this group, deterioration of the life quality, as well as indications and obstacles to surgical treatment of this defect. The above scientific points of view require a deeper study of the changes in the cardiovascular system in children with PCD. Among this contingent of children, the use of electrophysiological testing methods such as ECG and Exo CG allows not only objective assessment of central hemodynamic parameters, left ventricular pumping and contractility, but also changes in heart chambers, compensatory and pathological changes in cardiac hemodynamics [24, 49].

The rate of occurrence of heart rhythm and conduction disorders among children diagnosed with PCD did not significantly differ from those reported in the literature, both nomotopic (sinus tachy-, bradycardia and arrhythmia) and heterotopic (rhythm controller migration, extrasystole) type of

rhythm disorders in patient children [49, 57]. According to the data of ASMI, in the post-examination period, the occurrence of sinus tachycardia among 12-16-year-old children decreased slightly (pϕ<0.021), while it was found to be high among 7-11-year-old children (p>0.05). In our opinion, this situation is related to the end of the strong excitation of the parasympathetic part of the autonomic nervous system in the period before the examination. The incidence rate of interventricular impulse conduction (AV blocks) and mixed arrhythmias among children with PCD did not differ from literature data.

Complete right bundle branch block is defined by QRS complex expansion of up to 0.010 per second and more, expansion of S wave in I, AVL and left precardial leads (V_5, V_6), ST-T segment in relation to the main QRS complex and is characterized by a discordant secondary change of the T wave, as well as morphological changes in the form of rSR^I and rR^I of QRS complex. A morphological change in the form of rSr^I in V_1 extension of QRS complex was considered as a differentiating aspect of the right bundle branch block.

The ECG sign of the left anterior fascicular block deviates the heart electrical axis (HEA) from 0˚ to -10˚ in children aged 12-16 years, and to >-15 in children aged 7-11 years, qR in the I and AVL leads of the QRS complex, II, III and AVF transmissions were manifested in the form of morphological changes in the form of rS. The left posterior fascicular block is represented by the QRS complex in the form of RS in lead I, qR in the standard lead III.

In many cases, values of the diastolic function of the left ventricle, such as the maximum speed of early filling (ε, m/s), atrial filling (e, m/s), their ratio (e/A), are not studied, however, having high EF values against the background of a slight

increase in the diastolic volume of the left ventricle, indicates the presence of diastolic dysfunction of the left ventricle.

In general, values in 7-11-year-old children do not almost differ from changes in 12-16-year-old children, only in 7-11-year-old children, the changes in MV (2.39±0.30 and 1.90±0.02 l/min; p>0 ,05) is accompanied by a decrease in left ventricular systolic volume – ESV (13.8±0.95 and 15.8±0.20 ml; p<0.05). These shifts indicate that hemodynamics is normodynamic in nature. Compared to 12-16-year-old children, 7-11-year-old children after thoracoplasty have significant positive changes only in SV (p<0.05), % D (p<0.05) and DVI (p<0.01) values, In 12-16-year-old children, positive results were observed for EDV, ESV, SV, HI. In addition, when comparing intracardiac hemodynamics in the postoperative period according to the age norms, there is a deepening of EDV (p<0.01), MV (p<0.001) and HI (p<0.001) values, which turns the normodynamic hemodynamics, observed in the preoperative period, into a hyperdynamic type, is a sign of change.

These data indicate that 12-16-year-old children have less pronounced changes in Exo CG values compared to 7-11-year-old children, and disturbances in intracardiac hemodynamic parameters are better tolerated. In a word, after thoracoplasty in children aged 7-11 years, only systolic (right ventricle) and diastolic (left ventricle) tension and acceleration of Exo CG values such as dilatation and hypertrophy of heart ventricles and chambers are reduced. Taking into account significant changes in heart rhythm and conduction, intracardiac hemodynamics in children diagnosed with PCD, it is necessary to learn the clinical-functional relationships between chest deformity and CTD using electrophysiological research methods such as ECG and Exo CG. [4, 8].

The scientific aspects discussed in this chapter made it possible to put forward the following general conclusions:

1. Children diagnosed with PCD have less body weight than body length before diagnosis. It is more obvious in children aged 12-16. In the postoperative period, the weight deficit is eliminated, but the body length values remain unchanged.

2. There is a clinical-functional connection between the pathologies of the musculoskeletal organs associated with congenital pigeon chest deformity and connective tissue dysplasia.

3. Impulse conduction along the bundle branch legs is disturbed in children diagnosed with PCD. This is related to the systolic and diastolic tension of the ventricles, which are eliminated in the postoperative period.

4. Electrophysiological and visualization methods, such as ECG and Exo CG, allow to evaluate not only central hemodynamic parameters, but also left ventricular pumping and contractility characteristics, as well as changes in the heart chamber in children diagnosed with PCD.

Chapter III
MODERN APPROACHES TO THE TREATMENT OF PIGEON CHEST DEFORMITY IN CHILDREN

3.1. Elimination methods of pigeon chest deformity in children

Many methods of surgical treatment of PCD have been proposed in the literature [32, 33, 39, 45, 48, 56, 58, 60, 62, 63, 64, 65, 66, 67, 69, 70, 76, 85, 86, 88 , 94, 100, 101, 103, 104, 113, 117, 121]. This indicates that this problem is very complicated and has not yet found its solution. Currently, the available methods of surgical treatment of PCD can be divided into four main groups:

1. Without the use of fixing equipment
2. Using internal fixators
3. Using external fixators
4. Using external compression equipment

The first three methods are surgical methods, and the last one is a conservative treatment method. The leading method of treatment of PCD is surgery - thoracoplasty [70].

Among the various treatment methods offered, the leading place is occupied by surgical treatment - thoracoplasty method. There is no need for complex examination in conservative treatment methods with the use of external compression equipment. However, the long-term wearing of this type of equipment, which severely restricts breathing, as well as the possibility of relapse and their ineffectiveness, have somewhat limited the treatment with these methods. Because the design of this equipment is complex, it is very uncomfortable for patients,

and it requires constant supervision by specialists during the treatment period. In addition, it causes many social and household inconveniences for children, as well as disability for a long time. Because of this, until now these treatment methods are not widely used.

The use of internal fixators provides a good result for a long time after the operation and prevents the deformity recurrence, but in these methods it is necessary to conduct a re-examination - to remove the metal plates.

V.A.Timoshenko [73] performed metallosternochondroplasty in corporocostal and costal forms of PCD. The gist of this method is that after the xiphoid process is cut from the chest, the parietal pleura from both sides is separated in a retrosternal way. Then only the deformed ribs are opened and is cut subperichondrally. Rib arches are separated from the sternum. In order to eliminate the chest deformity, a transverse sternotomy is performed. For firm fixation of the sterno-costal complex in the corrected position, a titanium plate shaped like the patient's chest is placed in front of the chest and fixed to the bony part of the ribs. Rib arches are attached to the sternum using capron threads. The plate is removed after 6 months.

The method of using external "crushing" corsets [47, 56, 88, 141] is based on the plastic properties of the sternocostal complex in children, in which the chest is slowly crushed in the sagittal direction by reducing the length of the lateral handles of the external corset with the original design and at the same time, correction of the pigeon deformity is achieved. The period of walking with a corset is up to 2 years.

V.B. Shamik [78,79,80] proposed a technologically based method of surgical correction of PCD using an apparatus with an original design. In this method, a transverse or longitudinal sternotomy of the most bulging area of the chest is performed

without opening the retromediastinum. Chondrotomy of deformed ribs is performed on both sides of the chest. Then, correction needles are attached to the mattress in such a way that one end of the needle is fixed to the mattress, and the other end is fixed to the external correction device. With the help of corrective needles and apparatus, controlled continuous pressure is applied from front to back on the deformed sternum and ribs until the chest is in the correct position.

In the method of sternochondroplasty [4, 20, 73, 83, 84, 91], one (in boys) or two (in girls) transverse submammary incisions are made along the line passing through the deformity center. The xiphoid process is separated from the sternum. The parietal pleura layers are separated from both sides retrosternally in order to prevent further injury. Subperichondral resection is performed only in deformed rib cages. The rib arches are separated from the sternum and each is shortened by 3-5 cm. At the peak of the deformity, a wedge-shaped sternotomy is performed. In case of S-shaped curvature of the chest, two sternotomies are performed along the anterior and posterior surfaces of the chest. Shortened rib arches are sutured to the mattress using lavsan threads. Thus, the immobilization of the chest in the corrected position is carried out due to the traction force of the shortened rib arches.

In conclusion, choosing operative treatment methods and improving them still remain relevant.

One of the methods of thoracoplasty involves resection of the sternal body, sometimes total resection, which is not only extremely traumatic, but also leads to prolonged duration of the operation and bleeding.

As a result of wide resections of the sternum, the protective and frame function of the sternum-rib complex is disturbed. Some surgeons perform not only bilateral resection of deformed ribs, but also bilateral resection of ribs II-VII. This, along with

the extension of the operation time, also increases the level of its injury.

The lack of reliable fixation of the sternocostal complex in surgical treatment methods performed without fixating equipment, in turn, increases the rate of recurrence and complications, worsens the cosmetic result. Transverse, wavy, "Mercedes" type incisions lead to the formation of rough scars and spoil the aesthetic appearance of the patient. Therefore, the question of developing new thoracoplastic methods that enable good cosmetic and functional results without the use of fixating equipment remains relevant.

3.2. Elimination of pigeon chest deformity in children without fixating devices.

Currently more than 10 thoracoplasty methods are used in the surgical treatment of PCD. The most populars are: G.A. Bairov [9,10], M.M. Ravitch [130, 131], K.J. Welch [145], N.I. Kondrashin [48].

M.M. Ravitch in 1948, was one of the first to perform surgery in this pathology. performed thoracoplasty on an 18-year-old patient with manubriocostal deformity. Clinically, the patient also had tachycardia and shortness of breath. A lateral X-ray showed a Z-shaped sternum, i.e., the upper part is bulging and the lower part is sunken. An incision is made from the jugular fossa down to the xiphoid process. After the separation of the pectoral muscles, the upper part of the lower five ribs was resected (subperichondrally), the xiphoid process was separated from the sternum, and the rough fibrous ligaments between the lower part of the sternum and the diaphragm were separated from the sternum. A transverse wedge-shaped osteotomy was performed after some portion of the most convex part of the sternum was removed. By breaking the posterior cortical plate,

the upper third of the chest and body were elevated until they were in one horizontal plane. A second osteotomy was performed at the most concave point to elevate the sternal concavity, and a section was removed, the posterior cortical plate was broken and elevated. The fragment extracted during the bony osteotomy was placed in the space where the osteotomy was made. This made it possible to bend down the peripheral part of the mattress and keep the mattress in the correct position. After elevation, the sternum was fixed in the correct position with knotted sutures using silk threads [4, 22, 130, 131].

In addition, M.M. Ravitch [130] performed a transverse sternotomy of the anterior sternal plate in the region of the III rib in children. After removing the xiphoid process from the sternum, the deformed IV to VIII ribs on both sides were subjected to subperichondral resection, and corrugated sutures were placed on the periosteum under the removed rib segments. After that, in the correction position, the sternum was sutured to the sternum of the resected ribs in order to form a single frame of the chest. Most often, patients are operated at 10-15. However, M.M. Ravitch [130, 131, 132] believes that these surgical methods should be performed much earlier, without worsening the degree of deformity.

In 1953 S.W. Lester [122, 123] proposed two methods in PCD. In the first method, he performed a resection of the anterior part of the sternum, while in the second method, he performed a subperichondral resection of the deformed rib bones along with a subperiosteal incision along the entire sternal body. Both of these methods have not been widely used due to the high rate of injury and excessive bleeding. In addition, the rate of recurrence after operations performed in these methods was high [4, 71].

In 1958 E.F. Chin and H.A. Brodkin [90, 94] suggested resection of VI and VII th rib cages. After that, he performed a subperiostal resection of the sternal body till the IV th rib, and sutured the previously resected xiphoid process together with the rectus abdominis muscle. In this method, only the cosmetic defect was eliminated, and the function of the mediastinal organs was not improved.

According to the method of G.A. Bairov and I.A. Marshev [9, 10], 1-1.5 cm length is cut from the most sunken place of the V-VIII th ribs on both sides together with the anterior part of the periosteum. 1-2 cm width is resected from the most prominent part of the sternum. If there is a concavity in the lower part of the sternum, the outer plate is cut, and the back plate is broken under finger control. Cut ribs and sternum are sutured with separate capron threads. A compressive plaster bandage is then applied for 14-24 days.

N.I. Kondrashin proposed the following thoracoplasty method [48]: 0.5 cm is left from the point where the ribs are attached to the sternum, and longitudinal resection of the sternum is carried out in the I-II nd intercostal area. Later, incisions are made on the II-VII ribs on both sides at the transition to the bony part of the ribs. After that, the cut ribs with the bone plate of the sternum are transpositioned towards the center, adjusted to the resected edges of the sternum, and the II nd rib is attached to the sternum and sutured using lavsan threads.

The authors suggested subperichondral resection of the ribs II-III to the VII th rib on both sides. In this method, a transverse sternotomy is performed in the III-IV th intercostal space. After that, the distal end of the sternum is resected by forming a triangular incision at a distance of 1-1.5 cm, and xiphoid process is attached and sutured to it. The contraction of the rectus abdominis muscles causes the sternum to sink back. The large

muscles of the chest are sutured to each other along the middle line, and the rectus and oblique muscles of the abdomen are attached and sutured to them from below. In this way, a special muscle frame is formed, which presses on the front of the chest.

In 2001, a thoracoplasty method was proposed, with subperichondral-parasternal segmental resection (0.5-2.0 cm) of the II-VII th ribs leaving 0.5 cm from the sternum [72]. A bilateral chondrotomy is performed medially 1.0-1.5 cm from the border of the ossified and cartilagenous part of the ribs. When concavity is detected in the upper part of the ribs, additional chondrotomy is performed on them, if necessary. In the most prominent part of the sternum, with the base of the sternum directed forward, the wedge-shaped resection is performed. In the concave area of the sternum, the posterior cortical plate is undergone to additional sternotomy, and the anterior one is broken. The resected areas of the rib and sternum are adjusted and sutured with lavsan threads.

According to R.C. Shamberger [137], at present, operations performed using the thoracoplastic technique, proposed by K.J. Welch [145] in 1973, allowed to obtain good results. In this technique, a subperichondral resection is performed at the bulging rib cages, but the sternum is preserved, i.e., the sternum is not resected. The bulging part of the sternum is corrected by an anterior transverse osteotomy and a posterior fracture, which allows the sternum to move backwards.

Despite the fact that many methods of operative treatment of PCD have been proposed, according to literature sources, complications occur in 5-10% of cases. Firstly, the insufficient mobilization of the sternocostal complex, the formation of an angle in the areas of the ribs attached to the sternum cause the recurrence of deformity, and secondly, the penetration of infection from the places where the bone-retaining equipment is fixed leads to purulent complications in the postoperative

period. Thirdly, the operative treatment itself is considered to be complex and traumatic, so it is advisable to carry out these operative methods in medical institutions that have developed and implemented them.

Thus, taking into account the above shortcomings, even today, the issues of improving surgical correction methods of pigeon chest deformity in children are considered to be one of the actual issues in children's surgery.

In our clinic, the surgical treatment of PCD is based on the thoracoplasty method by Ravitch. From 2007 to 2016, significant changes were made to thoracoplasty techniques.

When analyzing the results of surveys conducted with Ravitch and his modifications (G.A. Bairov and I.A. Marshev, 1985) and the course of early and late postoperative periods, as well as the long-term results, we found the following: a wide access to the surgery area and restoration of the anatomic-topographic proportions of the tissues at the end of the surgery make its duration 1.5-2 hours. The volume of intraoperative bleeding is 150-200 ml due to opening a wide access to the sternocostal complex, resections of the sternum and ribs. In the early hours postoperatively, the pain syndrome lasts longer. Some patients were prescribed narcotic analgesics for 5-7 days to relieve the pain syndrome. Due to the same reasons, the period of activation of patients has been extended to 3-4 days.

At 1-year follow up after surgery, the results in some patients were not considered satisfactory. 1 patient had a partial recurrence of the deformity up to grade I, and another patient had a rough keloid scar. In the third patient, diagnosed with PCD along with the deformity of rib arches, the rib deformity did not resolve. These results did not provide physical and aesthetic satisfaction for both patients and their parents.

The above cases prompted us to find a technically new solution for the surgical treatment of pigeon chest deformity in children, using minimally invasive methods.

3.3. A new thoracoplastic method in PCD

The main principle of plastic surgery is to achieve the maximum functional result with minimal damage. The technical result of the developed and proposed thoracoplastic method is the improvement of treatment and cosmetic efficiency due to the intraoperative elimination of the deformity of the sternocostal complex and its stable strengthening in the corrected position.

Preparation for the surgery began directly with premedication. Premedication was performed 30 minutes before by intramuscular injection of 0.1% atropine, 1% diphenhydramine and tranquilizers based on the patient's age and weight. All surgeries were performed under total intravenous anesthesia using artificial lung ventilation in controlled breathing modes. Anesthesiological station Fabius plus X (Drager, Germany) and Savina-300 respirators (Drager, Germany) were used for anesthesia. All patients underwent peripheral venous catheterization using KD-Fix polyurethane catheters. For induction, 2-5 mg/kg of 5% ketamine hydrochloride solution was administered intravenously. For the purpose of myoplegia, 0.4% pipecuronium hydrobromide solution was used in 0.05 mg/kg. Propofol solution in 10 mg/kg was injected intravenously to maintain the level of anesthesia. A 0.005% fentanyl solution 0.1 ml/kg was used as the main anesthesia. One of the hydroxyethyl starches was used to replenish the blood volume lost intraoperatively. After the surgery, the patients were extubated and transferred to the post-anesthesia room under the supervision of an anesthesiologist.

In the literature, opinions on the choice of skin incision in the surgery of PCD remain controversial. In our observations, after thoracoplastic procedures performed with undulating or submammary incisions, patients mainly complained of a wide and coarse postoperative scar. The technical result obtained from the proposed thoracoplastic method was achieved first by changing the skin incision. The skin incision differs from other methods in that a vertical incision of up to 8 cm in length is made from the area of the maximum bulge of the chest, not wavy or submammary up to 12-15 cm (Fig. 3.3).

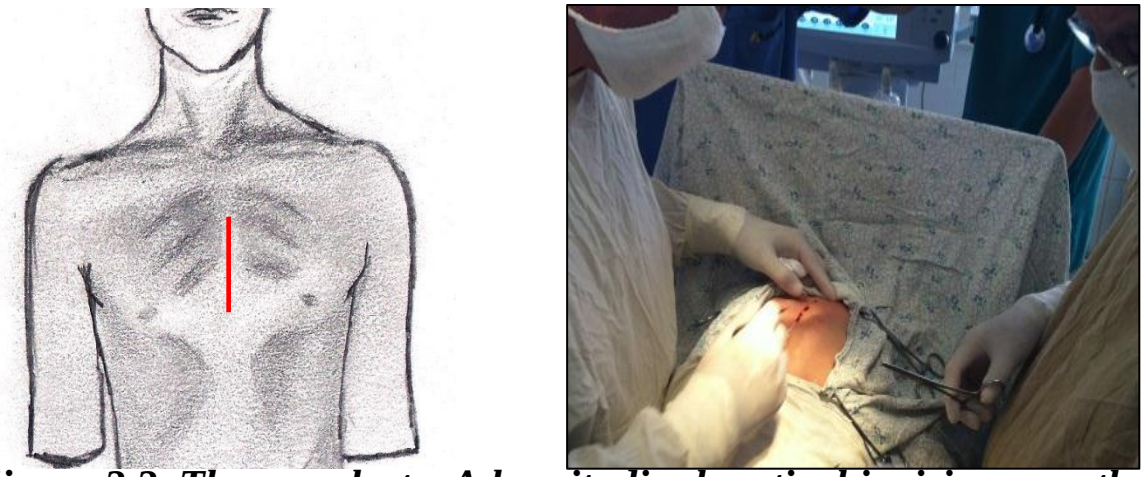

Figure 3.3. Thoracoplasty. A longitudinal vertical incision over the sternum.

In addition, unlike other methods, tissues are not mobilized layer by layer (skin, subcutaneous fat layer, muscles), but as a single clot (Fig. 3.4).

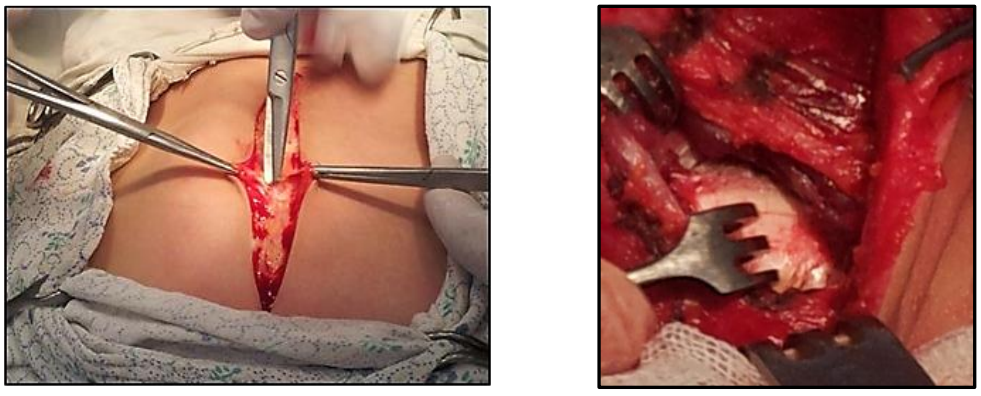

Figure 3.4. Thoracoplasty. Tissues to be mobilized as a single clot

At the next stage, a technically positive result is achieved through bilateral resection of the ribs. The main difference of the applied method is that the bilateral resection from the 2nd to the 8th rib is not subperichondral or subperiostal, but a complete resection of the ribs (along with the cartilage or periosteum) is performed (Fig. 3.5).

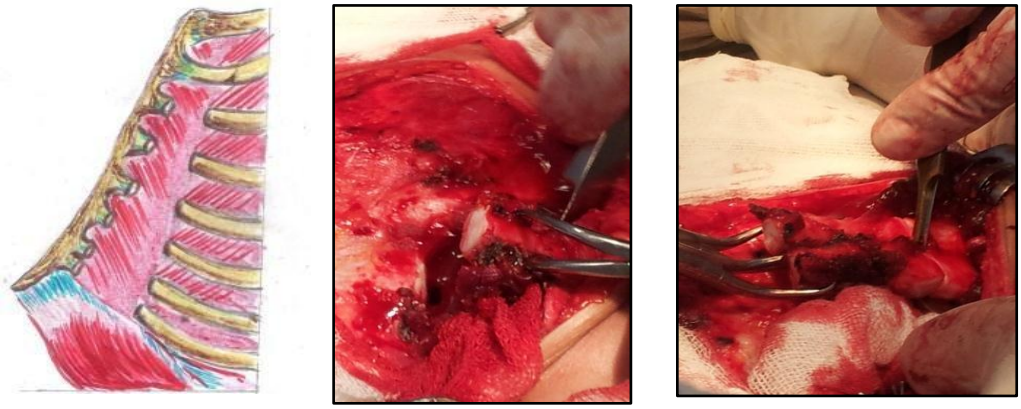

Figure 3.5. Thoracoplasty. Full of ribs bilateral resection

At the next stage, a semilunar incision is made in the area of the xiphoid process, unlike other methods, the xiphoid process is grasped with an instrument, separated from the tissues behind it (together with the retrosternal ligament) and the process is removed, and the shortening of this ligament is achieved by fixing it to the lower 1/3 of the sternum. (Figure 3.6).

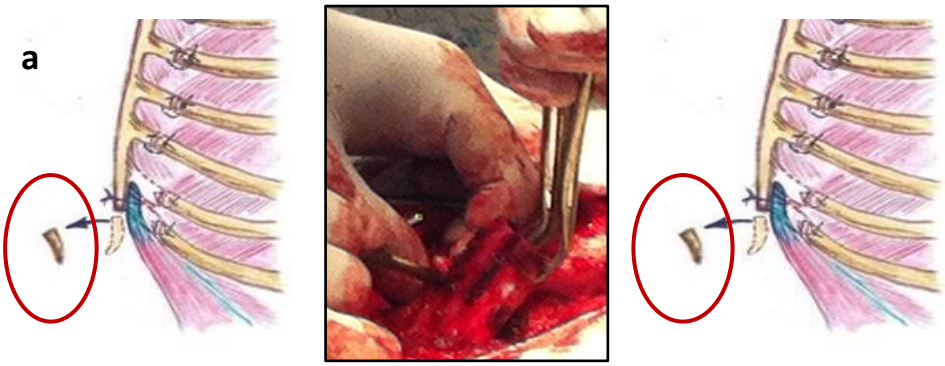

Figure 3.6. Thoracoplasty. The xiphoid process is removed (a).

Retrosternal ligament is shortened by fixating it to the lower 1/3 of the sternum (b).

At the next stage, in contrast to other methods, a positive result was also achieved by performing a transverse sternotomy with the help of a wedge-shaped incision, leaving the back plate of the sternum from its maximum convex part, depending on the type and degree of the deformity (Fig. 3.7).

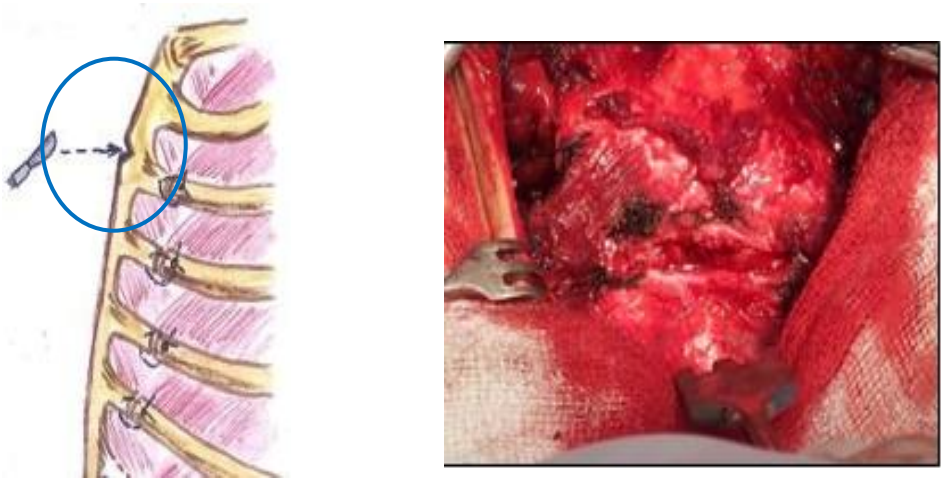

Figure 3.7 Thoracoplasty. Wedge-shaped sternotomy.

In asymmetric, pyramidal types of PCD, in order to ensure the location of the sternum along a flat surface, a longitudinal sternotomy is performed and a separate suture is placed. The resected ribs are sutured end to end (Fig. 3.8).

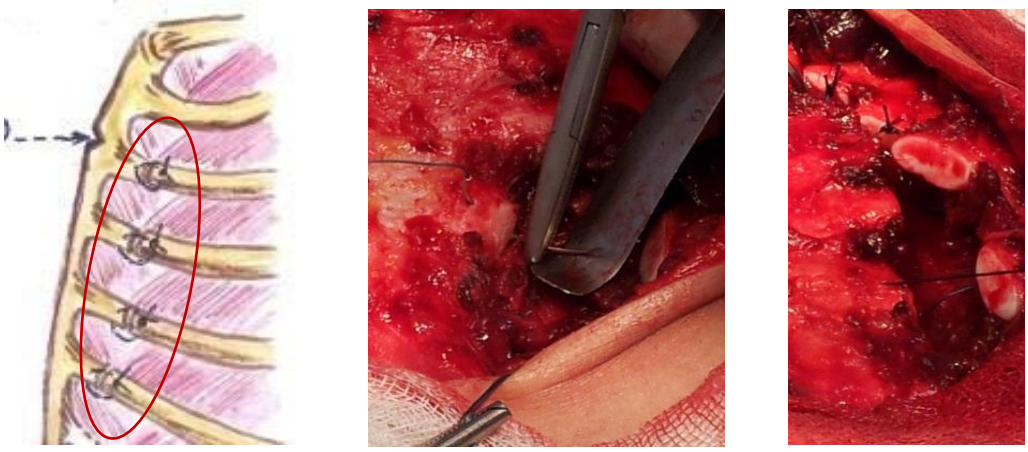

Figure 3.8. Thoracoplasty. End to end suturing of the resected ribs.

In order to more convincingly strengthen the sternocostal complex in its position, the proportion of the chest muscles is achieved out by suturing in the form of a ladder (Fig. 3.9). Then the tissues are restored anatomo-topographically, the subcutaneous fat layer is sutured, and the surgery is completed by placing a cosmetic suture on the skin. There was no need for drainage of the chest cavity due to less trauma, less intraoperative bleeding and hematomas.

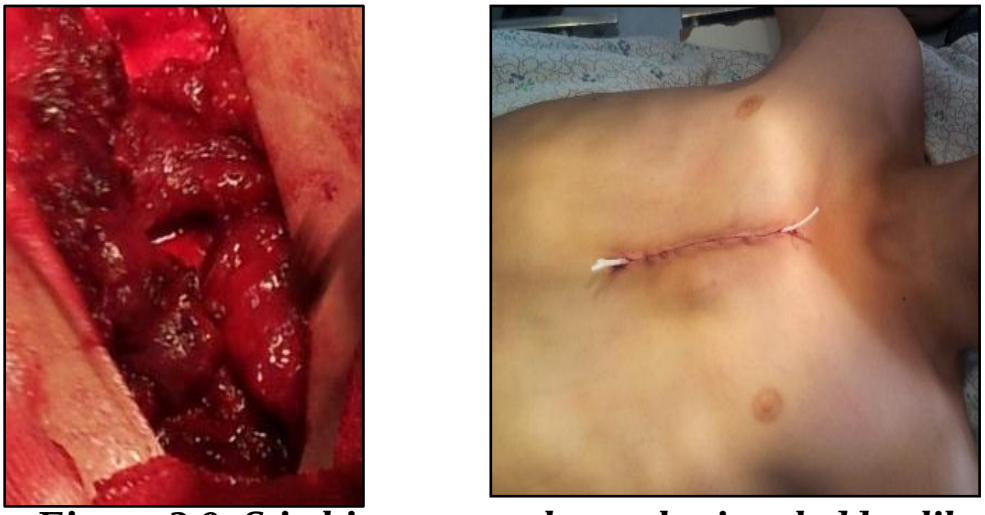

Figure 3.9. Stitching pectoral muscles in a ladder-like manner and cosmetic skin suture

Chapter IV
RESULTS OF SURGICAL TREATMENT IN CHILDREN WITH CONFIRMED PIGEON CHEST DEFORMITY

In order to evaluate the clinical effectiveness of thoracoplastic surgery in children with pigeon chest deformity, intraoperative, early and long-term results were analyzed.

The amount of intraoperative bleeding was 40-50 ml, and the surgery lasted for 46 minutes on average.

In the early postoperative period, the results of surgical treatment were evaluated according to the following parameters: duration of analgesia with narcotic analgesics or nonsteroid anti-inflammatory agents; the period of patient activation; changes in the Louis angle in radiographic data after thoracoplasty; degree of complication.

The following criteria were used to evaluate long term treatment results after the surgery: 1) cosmetic efficiency; 2) anthropometric parameters; 3) radiographic parameters; 4) functional parameters of intrathoracic organs; 5) the presence of complications such as bulging or sinking of some parts of the chest or ribs. Evaluation of the results obtained after thoracoplastic surgery was carried out based on a questionnaire using a 4-point scale:

A positive result - is the complete elimination of the deformity, the disappearance of its residual signs and chest asymmetry, the chest formation as in healthy children and normotrophic scarring of the skin, cosmetically smooth chest, correct stature, compliance of anthropometric parameters with age norms, chest axis is in the right position on X-ray of lateral projection, the functional parameters of the intrathoracic organs are close to the norm.

A good result - is the complete elimination of the deformity, the absence of its residual signs, the disappearance of chest asymmetry and hypertrophic scarring of the skin,

A satisfactory result - preservation of small deformity signs and chest asymmetry, slight deviation of the rib arches or keloid scarring of the skin, slight retardation of physical development, slight stature distortion, cosmetically a slight bulging or sinking of the sternum in the surgery area, exostoses in the area of rib resection confirmed on X-ray.

An unsatisfactory result - recurrence of pigeon chest deformity. The appearance of a cosmetical bulge on the anterior chest wall that needs to be removed again.

4.1. Comparative analysis of the results of surgical treatment of pigeon chest deformity in children

A number of authors have suggested the use of different drainage systems due to the high volume of blood lost during the surgery. However, the use of these systems may cause delayed wound healing after the surgery. By separating the tissue as a single clot, reduced volume of intraoperative bleeding has been achieved, so it is better to avoid the use of drainage systems.

4.2. Prevalence of pigeon chest deformity among children, its classification and terminology

Deformities of the chest in children are pathological changes in the shape, size and dimensions of the chest, which are defects that lead to shortening or lengthening of the chest-spine distance and, as a result, to the topographic disruption of internal organs. 90% of chest deformities are funnel chest deformities. In the second place are pigeon chest deformities and then various anomalies of the ribs, Poland and Currarino-Silverman syndromes, sternum deviation, etc. In general, chest deformities

occur in 1-4% of the population. Among children (mostly in boys), this is 0.6-2.3%, and it is mainly characterized by a cosmetic defect of the chest, functional disorders in the respiratory and cardiovascular systems, and their dissatisfaction with the spiritual aspect. Defects in bones and cartilages lead to a decrease in the framework and protective functions of the chest, and cosmetic defects lead to the development of mental disorders. Such children are introverted, restrained from their peers, and sometimes have an aggressive character (Timoshchenko V.A., 1995). These conditions have a negative impact on the harmonious development of children and their social adaptation. Therefore, the problem of chest deformities is one of the actual issues of children's thoracic surgery, traumatology and orthopedics, cardiology and psychology.

Pigeon chest deformity (syn. pectus carinatum, "pigeon", "chicken" breast, chicken-breast, keeled chest deformity) is characterized by forward symmetrical or asymmetrical tilting of the chest and the ribs attached to it. This condition can have several components of chest deformity with unilateral or bilateral damage to the rib cages and forward bulging in the upper and lower parts of the sternum.

Unlike funnel deformity, pigeon deformity is less common and accounts for 6-22% of all chest deformities. (Bairov G.A. 1983, Hecher M. Happ 1986).

According to all authors, PCD is more common in boys. Shamberger R. in 1987 observed PCD in 119 (78.3%) boys and 33 (21.7%) girls [74]. In 1973 Welch K. found it in 23 (88.5%) men and 3 (11.5%) women. On average, PCD is three times more common in boys than in girls. According to many authors, in contrast to the funnel chest, PCD can be detected in childhood and adolescence, even at birth or in the first year of life. This

defect is detected in 1/3 of patients at birth, and in almost half of them it begins to appear after puberty "jump" [4].

According to the results of K. Welch's research, 26% of patients with "chicken breast" had breast deformities in other family members. As concomitant diseases, he cited kyphosis, tibia vara, arachnodactyly, talipes planovalgus, Osgood-Schlatter disease, microcephaly, diffuse hypotony. In 10 out of 26 patients, he observed respiratory diseases and in 3 of them, severe type of bronchial asthma. Cardiovascular system concomitant diseases were found in 1 out of 26 people [81].

The R. Shamberger data (1987) is also consistent with the data given by K. Welch. According to his observations, family members of 26% patients had some type of chest deformity, 12% had scoliosis, 32 out of 152 patients (51.7%) had musculoskeletal disorders [74].

In PCD, the deformity of rib cages, arches, and sternum is observed, and the sternum and the ribs attached to it are displaced forward. This leads to an increase in the anterior-posterior dimension of the chest and pigeon chest deformity.

In X-ray examinations, retrosternal space is expanded in these patients, the heart is rotated around its axis and has a droplet appearance. On lateral X-rays, the chest has the appearance of separate segments [7, 8].

Several classifications of PCD from a clinical-anatomic point of view have been proposed.

C. W. Lester [122, 123] proposed two forms of PCD, i.e. upper arcuate and lower curved.

H.A. Brodkin [90] also divided PCD into high - chondromanubrial prominence and - chondrogladiolar prominence forms.

F. Robichek [133, 134] divided PCD into the following types:

- symmetric - the sternum is elongated and attached to the xiphoid process at an abnormal angle, as a result of which the bulge of the sternum is visible, and the lower ribs are very deformed and are not visible due to their overlap.

- lateral deformities of the chest - the bulge of the rib arches is more obvious, and the wrong position of the chest is less visible. These deformities can be unilateral and bilateral;

- chondromanubrial deformity - only the sternal body and two rib cages protrude forward.

K. J. Welch, A. Vos [145] divided PCD into chondromanubrial or arcuate and chondrogladio or sloping deformities based on their research results. In addition, symmetric and asymmetric forms are noted.

In 1983 G.A. Bairov and A.A.Fokin proposed a comprehensive classification from clinic-anatomical point of view [9, 10, 46, 75, 139].

Congenital pigeon chest deformities:

I. Manubryocostal type:
1. With sunken sternal body
2. With non sunken sternal body
a) triangular
- symmetric; asymmetric;

II. Corporocostal type:
1. Sunken to the side
2. Non sunken to the side
a) round shape
b) pyramidal
- symmetric; asymmetric;

III. Costal type:
1. With sternal rotation
2. Without sternal rotation
a) elliptical, with lateral protrusion;
- asymmetric;

Acquired pigeon chest deformities:

I. Postoperative:

1. Funnel chest hypercorrection
2. After the operation carried out in the deviated sternum

a) in different forms

- symmetric, asymmetric;

II. Post-traumatic:

1. With a pseudo joint in the sternum
2. Without a pseudo joint in the sternum

a) in different forms

- symmetric, asymmetric;

The manubriocostal type is characterized by forward protrusion of the sternum and 2-3 rd ribs and simultaneous inwardness of the sternum.

There are two forms of the corporocostal type:

- Pyramidal - the chest is slanted in a straight line down and forward towards the wedge-shaped growth. The place where the middle and lower part of the chest joins the wedge-shaped growth is considered to be the maximum protruding area.

- Round - the chest is curved forward in its middle and lower parts.

The costal type is characterized by the bulging of the chest wall from the side due to the deformed ribs. In this form, the sternum is undergone rotation along the longitudinal axis. In this case, the ribs bulge forward on one side, while on the other side they are normal or sunken inwards, and the sternum is twisted in relation to the less convex side. Thus this type of deformity always occurs in an asymmetric form.

Acquired funnel chest deformities can occur after surgery or injury.

G.A. Bairov and A.A. Fokin divided all PCD into symmetric and asymmetric forms, except for the above-mentioned costal

type. The followings can be causes: the bulging degree of the ribs is not the same on both sides, the chest rotation along the longitudinal axis, and the difference in the number of ribs involved in the deformity on the right and left sides.

According to the changes in dynamics, these authors also divided into types that decrease over time, grow in parallel with the child's age and development, and accelerate, that is, surpass the child's age and development.

All authors point out that in clinical practice, the sternocostal (chondrogladiolar) form is most common (Ashcraft, 1996). It is characterized by symmetrical bulging of the lower ribs and sternum on both sides. The high or "clumpy" deformity is relatively less common.

According to some authors [4], one of the unique features of PCD is that it is only a cosmetic defect, and dysfunction of mediastinal organs does not occur.

A number of authors such as Picard LR, Tepas JJ, Shermeta DW, Haller JA. [63], Ashcraft K.U. [2], Kondarshin N.I. [26] consider PCD to be only a cosmetic defect, since no deviations from age-related norms were observed in functional examinations. According to many authors [5], as a child ages, functional disorders occur due to a decrease in the mobility of the chest.

G.A. Bairov and A.A. Fokin [9, 10, 75, 106] point out that these patients’ chests are in the position of maximum "breathing", and due to the reduction of the respiratory excursion, bronchopulmonary diseases are often more common. In addition, lethargy, profuse sweating, difficult breathing through the nose, and the presence of adenoid hypertrophy are also observed in this category of children. Of course, congenital predisposition to the respiratory diseases also plays an important role.

According to these authors, one out of every 6 children with PCD has chronic pneumonia, disorders of the topography or development of the bronchopulmonary system (polycystosis, hypoplasia), lobar emphysema, bronchiectasis. In addition, these authors observed cardiac pathologies such as ventricular septal defect, systolic murmur, sinus tachycardia, and various forms of arrhythmias in 6 of 17 patients. K.Welch explains the final results in the long-term existence of PCD as follows: an enlarged anterior-posterior dimensions, chest becomes less mobile, which leads to inefficiency of aphasic movements, which are carried out only due to the diaphragm and accessory muscles. Pulmonary stability is gradually reduced, progressive emphysema and concomitant lung infection develop. Robichek called such a chest "frozen". He notes that symptoms of PCD can range from a gradual progression to wheezing, asthma attacks, and tachycardia. Even "asymptomatic" patients can develop early emphysema, pneumothorax, and pulmonary heart disease. Californian E.W. Fonkalsurd and S. Beanes, based on the experience of treating 90 patients with PCD during 1970-2000, proved that this pathology is not only a cosmetic problem, but a disease with serious functional changes. In 85 out of 90 patients, a decrease in the vital volume of the lungs (94.4%), respiratory diseases (54%), bronchial asthma (26.6%), pain and discomfort in the chest area (42.2%), changes in the lungs requiring additional oxygenation (2.2%) were detected.

In PCD, the chest is partially enlarged and emphysematous expanded, which in turn reduces its elasticity and tolerance to strain [53].

The psychological aspects of deformity are also not less important. According to Robichek, patients with pectus carinatum tend to be thin and shy. They always walk and sit in a hunched over position to hide their flaws. During school, they become an object of ridicule among their peers, they are shy to

participate in the pool and various activities. A cosmetic defect leads to inhumanity, and sometimes extreme aggression [70]. All these factors lead to deterioration of mental and physical condition. 22 out of 26 patients (84.6%) with PCD had a strong mental reaction to the deformity (K. Welch): shyness, self-doubt, inability to undress, disgust for the bulge in the breast area, etc.

According to G.A.Bairov: "An initial cosmetic defect in a child later will lead to a significant decrease in the activity of an adult. In contrast to the funnel chest, the pigeon chest is permanently traumatizing because it cannot be hidden behind clothing. All of our patients were ashamed of their existing deformity from a certain age (often 10 years old), refused to go to the bathroom, to the beach, to play with their peers. Older people with such deformities always insist on being treated and operated."

Despite the existence of many assumptions and theories, there is still no single opinion about the etiology of PCD. There are many assumptions that its development can be caused by conditions such as rickets [27], respiratory obstruction [20, 27], high intrauterine pressure during pregnancy [37], imbalance between chest strength and diaphragm traction [15, 16].

In addition, there are theories about changes in the quantitative and qualitative composition of collagen [4,6], glycosaminoglycans, and water [11], which lead to a decrease in the strength of the rib cages. According to many scientists, the reason for the development of PCD is chondrodysplasia of the rib cages, and this condition is manifested by the rapid growth of the ribs [5, 7–9, 38]. The theory that rib cage overgrowth leads to PCD is supported by many scientists. The exact cause of rapid growth of the ribs remains unknown, and the results of studies contradict each other. For example, T. Nakaoka and co-

authors note that the ribs of children with PCD are not longer than those of healthy children [33].

Some authors consider PCD to be one of the phenotypic manifestations of connective tissue dysplasia (CTD) [1, 2]. The term "dysplasia" refers to the abnormal growth and development of an organ or tissue. A diagnosis of CTD is made based on a thorough analysis of symptoms or clinical studies. Nevertheless, in clinical practice, this diagnosis is put together with the main disease if it is not clearly confirmed from the histological point of view. Therefore, clinically detected dysplasia can be the result of many changes in tissue structure [10].

Connective tissue dysplasia is a genetically determined process, which is based on the mutation of the genes responsible for the synthesis of collagen fibers. As a proof of this, it can be noted that in almost all cases of scoliosis and Marfan syndrome with connective tissue anomalies, deformities of the anterior chest wall occur together [28, 29, 35, 37]. The presence of an aggravating family history in probands with PCD is 37% [35], which indicates that this pathology is genetically determined. As a result of the mutation, the initial three-helix conformation of the collagen macromolecule is disturbed, and its stability decreases. Fibrils and fibers are formed defectively, fibrous structures cannot withstand mechanical stress.

Of special interest is the comparative histochemical, morphological, and biomechanical description of rib cages in PCD by J. Feng et al. As a result of the research of these authors, it was proved that the roughness of the ribs (Young's modulus, compression and displacement) decreases and the strength threshold decreases when they are stretched, crushed and displaced [21].

In PCD, we will pay special attention to the works devoted to the biochemical and morphological researches of rib cages.

The cartilages in the rib complex belong to the hyaline group. Type II collagen and glycosaminoglycans are the main components of the extracellular matrix of hyaline cartilage. The core macromolecules of type II collagens are attached to fibrils and reinforced by intermolecular cross-links. The number of these connections increases with age [12]. A fine fibrillar network is considered to be the basis of the matrix framework in the hyaline. Glycosaminoglycans (GAGs) are polymer chains of sulfated amino sugars and carboxylated sugars (uronic acid). The degree and stereometry of sulfation and carboxylation determine the species composition of GAGs. Polysaccharide chains of GAG are attached to the core protein and form proteoglycan. In hyaline cells, proteoglycans together with hyaluronic acid form huge (mass up to 108 D) aggregates. Such aggregates strengthen the fibrillar network due to mechanical immobilization of fibrils. This ensures that the hyaline is stable against shear-related deformitys. In addition, the high concentration of anionic groups in GAG maintains a high osmotic pressure, which ensures the retention of water in the tendon and, in turn, the resistance of the tendon to compressive stresses [31].

There is very little information about the chemical composition of rib cages and how they change in PCD. According to Svetkova T.A. and her co-authors' information, the percentage of collagens in dehydrated preparations was 56.2±1.6. This value does not statistically differ from the collagen content in preparations obtained from isolated (55.7±0.5%) and syndromic forms (55.6±1.1%) of PCD. The percentage of collagens in wet preparations prepared from PCD was higher than in the control group (≈ 45 and 60%). The authors attribute this difference to the reduced ability of the rib cage matrix to absorb water in PCD. However, the level of uronic acids remained the same level in the control group

(3.3±0.5%), as well as in isolated (3.5±0.7%), and syndromic (3.6±0.1%) forms of PCD. The authors explain this situation with a change in the species-specific composition of GAG [11].

J. Feng and co-authors showed that no difference was detected when safranin histochemically stained with O and PAS to detect GAG in the cross-sections of the rib cages from the control group and from PCD [21]. However, this situation does not rule out the possibility of species-specific changes in GAG.

In 2011 this assumption was indirectly confirmed in the scientific work published by V.L. David and co-authors. When the authors stained the sections from the control group and from PCD patients together with safranin O and Alcian blue, they noticed that the preparations from patients with PCD patients bind more with alcian, while in the control group, safraninophilia was observed. The authors concluded that while in alcyanophilia the shift towards safraninophilia is determined by an increase in the level of GAG sulfation, it means that the level of GAG sulfation decreases in PCD. According to them, this condition causes a decrease in water absorption and deterioration of the mechanical properties of the rib cage [18].

As for the type-dependent composition of collagen, the results of immunohistochemical analyzes confirm that type II collagen is the main component in the structure of the cartilage tissue in both the control group and PCD [21]. A significant increase in type V collagen content was observed in both groups [4]. However, the following differences are observed in PCD compared to the control group:

The distribution of type II collagen in deep (away from the epidermis) zones is not same; the presence of areas with a high (strong staining) and low concentration (weak staining) of proteins and even none at all (discoloration) [21];

Presence of type III and IV procollagens [4];

Increased concentration of type V collagen and fibronectin [4].

In addition, it has been convincingly confirmed that the percentage of soluble collagen in the ribs of patients with PCD is significantly reduced. This collagen is extracted from tissue by saline solutions, acetic acid, proteolytic enzyme (pepsin). This evidence is the basis for the assumption of an increase in the amount of intermolecular and intramolecular crosslinks (premature aging).

Indeed, Borisova N.V. and studies of raw materials have shown that hydroxylysylpyridinoline linkages, specific for type II collagen, are increased in PCD [14]. In addition, the presence of lysylpyridinoline linkages, characteristic of type I collagen, has also been identified. These results indicate a severe impairment of the posttranslational modification of collagen networks in PCD. In addition to this opinion, it can be noted that there are data on the decrease in the urinary excretion of free oxyproline and oxyproline-binding peptides, which are considered collagen degradation products.

Therefore, it is necessary to pay attention to the results of the analysis of the rib samples. These samples, obtained from patients with PCD, had higher levels of magnesium and calcium and lower levels of spirit compared to the control group [37]. As well known, Zn^{2+} ion enters the active center of single enzymes that degrade three-helix core macromolecules of matrix metalloproteinases (MMP), i.e. collagen. Obviously, if MMP activity is reduced in the tissue, the remodeling of the collagen network is impaired and leads to the development of connective tissue imbalance.

Another research directions are aimed at studying the structure of chondrocytes and intercellular substances in PCD and normal. Some researchers believe that hyperplasia of rib cages is the basis for the formation of PCD [22, 34], while other authors state that there is no sharp difference between the chondrocytes of the PCD and the control group [3, 18, 21, 25].

According to K. Mullard, biomechanical disorders of the ribs are secondary, the main cause of which is the violation of chondro- and osteogenesis [32].

According to H. Rupprecht and N. Freiberger, the rib cages in patients with PCD have blood vessels in all sections. The number of blood vessels per unit area is same in deformed and intact rib cages. The number of chondrocytes in one chondron increases dramatically with age compared to the control group [34]. J. Feng and co-authors declare that the above data have not been verified. According to them, chondrocytes and their nuclei are intact [21]. Also, no signs of hypo- or hyperplasia of rib cages were detected in electron and transmission microscopy. H. Rupprecht and N. Freiberger noted that the number of blood vessels in the ribs of children with PCD does not differ between patients and healthy children [34]. The claim that the number of chondrocytes in one chondron increases dramatically with age has not been confirmed. According to J. Feng and co-authors, the number of cells does not change [21].

E.A. Bardakhchyan and Khammualifs put forward the opinion that the morphological substrate of chest deformity is the disturbance of chondrocytes, intercellular substance and fibers at the ultrastructural level [3]. Changes in chondrocytes in PCD are mainly associated with the development of dystrophic process - fatty and carbohydrate dystrophy, formation of atypical asbestos-like fibrils in pericellular zones. In this case, almost no organelles remain, and those that can be identified are functionally disproportionate.

According to the information provided by V.M.Kuritsin and co-authors, common morphological features of the rib cage in normal and PCD are acellular, map-like areas, desquamated chondrin fibers and "cerebral" spaces [4]. However, in PCD they develop 6-7 years earlier and are detected 3 times more often than in healthy children.

Thus, despite the fact that there are many studies and scientific publications devoted to the study of the structure and characteristics of rib cages in normal and PCD, the causes and pathogenesis of PCD are still unknown. It is also worth noting that most of the research on rib cages was done decades ago. Therefore, the study of rib cages in normal and pathological conditions with modern equipment may allow to determine their structure, appearance, characteristics, and finally the real cause of chest deformity.

CONCLUSION

Treatment of congenital malformations of the chest is still considered to be one of the actual issues of pediatric surgery. The funnel chest takes the second place among the chest deformities after the pigeon chest. This defect is considered to be complex bone and cartilage pathology, manifested by various clinical courses. It is characterized by a symmetric or asymmetric tilting of the sternum, as well as the ribs attached to it, towards the front. This pathology is detected in 30% of patients at birth, and in about 50% of cases, it appears at the beginning of puberty. Hereditary genesis is observed in 26% of cases. PCD is considered to be related to systemic dyschondroplasias, which often occurs together with many other pathologies accompanied by connective tissue dysplasia. In this pathology, defects in the chest bones and cartilages lead to the weakened protective function. Cosmetic defects cause mental disorders, and children try to avoid their peers. These aspects have a negative impact on the harmonious development and social adaptation of these children.

When examined by functional testing methods, other pathologies are often not detected in such sick children, but they develop a complex of decreased sense of fullness. Along with the presence of a cosmetic defect in this pathology, there are often significant functional disorders in the cardiovascular and respiratory systems. E.W. Fonkalsrud and S. Beanes 2001, based on 30 years of treatment experience in patients with PCD, admit that the skeletal deformity is not only a cosmetic defect. Such deformities can cause functional disorders (reduction of lung vital capacity, increase of respiratory minute volume, decrease of oxygen absorption coefficient, etc.) due to the chest-rib complex being in a state of "permanent breathing" and the limitation of the rib movement.

Currently, there are no approved universal standards for the diagnosis of chest deformities. Many authors think that there is a need to carry out computed tomography (CT) and magnetic resonance imaging (MRI) in order to accurately diagnose the form and degree of deformity, determine the indications for thoracoplasty, calculate the optimal volume of resection of deformed ribs and thoracoplasty, and evaluate the shape of the chest before and after the examination. Chest index (CI), which determines the ratio of the anterior-posterior and transverse dimensions of the chest, as well as parameters such as volume index of deformity, chest volume, heart compression index, are being used more often. It was found that the changes in the external dimensions of the chest when differentially compared with internal changes, recorded in anthropometric examinations and computed tomography, have a high correlation (r=0.99). Thus low-cost anthropometric methods can be effectively used to evaluate the shape, developmental progress, effectiveness of rib resection and performed reconstructive thoracoplasty examinations in chest deformities.

Until now, the surgical method has been the leader in the treatment of PCD, and many of them have been proposed. This indicates that this problem is very complicated and has not yet found its solution. Most of the proposed surgical methods are characterized by the fact that they are traumatic, and there is a high probability of intra- and post-surgical complications. In practice, almost all surgeries require a wide skin incision, resection of deformed rib arches. The formation of a rough post-operative scar impairs the aesthetic result of thoracoplasty, and rib resection cannot eliminate the risk of bleeding, even with the use of modern electrosurgical instruments. Currently, at the stage of modern development of surgery, attention is paid to surgical methods that are less invasive, less traumatic, and at the

same time, aimed at obtaining the maximum cosmetic effect and the complete elimination of chest deformity.

The above scientific aspects determine the prospective directions and practical importance of the research aimed at the development and implementation of new surgical methods in various forms of PCD in children.

A number of dysplastic diseases of the musculoskeletal system are characterized by a poor outcome and a severe course. However, researchers often consider musculoskeletal dysplasia as external markers of connective tissue deficiency, and comprehensive clinical-instrumental examinations are not conducted in this regard. Therefore, there is a need for a comprehensive study of the orthopedic status of children with pigeon chest deformity and to evaluate the significance of the identified patterns in the diagnosis of connective tissue dysplasia.

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KEEL SHAPED DEFORMITY IN CHILDREN

O‘QUV ADABIYOTINING NASHR RUXSATNOMASI

O‘zbekiston Respublikasi Oliy ta’lim, fan va innovatsiyalar vazirligi, Andijon davlat tibbiyot instituti rektorining 2023 yil “30” sentyabrdagi “01/08/67-Sh”–sonli buyrug‘iga asosan

K.S.Yulchiyev, K.Z.Kadirov, D.K.Karimov
(muallifning familiyasi, ismi-sharifi)

Pediatric work - 5510200
(ta’lim yo‘nalishi (mutaxassisligi)

_____
_____ ning
talabalari (o‘quvchilari) uchun tavsiya etilgan.

Keel shaped deformity in children *nomli o‘quv qo‘llanmasi*
(o‘quv adabiyotining nomi va turi: darslik, o‘quv qo‘llanma)

_____
_____ ga
O‘zbekiston Respublikasi Vazirlar Mahkamasi tomonidan litsenziya berilgan nashriyotlarda nashr etishga ruxsat berildi.

Rektor _____ M.M. Madazimov
(imzo)

Ro‘yxatga olish raqami: 100233

K.S.Yulchiev, K.Z.Kadirov, D.K.Karimov.

Ilmiy nashr

YULCHIEV K.S. KADIROV K.Z. KARIMOV D.K.

KEEL SHAPED DEFORMITY IN CHILDREN

O’quv qo’llanma

Muharrir: D.Axmedova
Dizayner: Z.Axmedova
Musahhih: B.Axmedov

“STEP BY STEP PRINT” MChJ
Andijon shahri. Xrabek ko’chasi 94 b uy.
Nashriyot litsenziya raqami: № 040955
2022-yil 29 sentabrda berilgan.

Bosmaxonaga 2023 yil 25 sentabrda berildi. Bosishga 2023 yil 05 oktabrda ruxsat etildi. Bichimi 84x108 1/32. Hajmi 5. bosma taboq. Times New Roman garniturasi. Ofset qog’ozi, offset usulida chop etildi. Buyurtma 11. Adadi 100 dona.

“Step by step print” MChJ bosmaxonasida chop etildi.
Andijon shahar Xrabek ko’chasi 94-b uy.
O’zbekiston Respublikasi Prezidenti admintratsiyasi huzuridagi Axborot va ommaviy kommunikatsiyalar agentligining 12.07.2019.dagi 12-3299. Raqamli guvohnoma.