

RESULTS OF TREATMENT OF SPRENGEL DISEASE IN CHILDREN.

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Summary. *In the department of pediatric orthopedics of the RSSPMCTO over the past 15 years 82 children with a congenital high scapula at the age from 3 to 14 years have been treated. The osteoarticular and neuromuscular systems, X-ray, MSCT and electroneuromyographic studies were performed to assess the general condition of the patient. Surgical treatment was carried out in 34 children without reconstruction and in 40 with reconstruction of the shape of the scapula. Of all 74 patients examined in the long-term period after treatment, we obtained good results in 75.7% of cases, satisfactory in 21.6%, and unsatisfactory anatomical and cosmetic treatment results in 2.7%.*

Keywords: *scapula, clavicle, shoulder, spine, operation, results.*

Relevance. Congenital high standing of the scapula or Sprengel disease is a complex anomaly in the development of the shoulder girdle, spine and chest. The main clinical evidences of pathology are the high standing of the scapula with limited abduction in the shoulder joint and a pronounced cosmetic defect, with deformities of the head, neck, chest and posture of the child. More than 30 methods of surgical treatment of Sprengel disease have been proposed by far, which can be conditionally divided into the following groups: operations of lowering the outer part of the scapula; partial and subtotal resection of the scapula; scapula lowering operations without bone-based interventions. The development and implementation of new methods of operations allowed to reduce the percentage of unfavorable treatment outcomes for this disease. However, in our opinion, insufficient attention was given to the pathological deformation of the shape of the scapula.

The aim of the study is the analysis of the surgical treatment outcomes in children with congenital high scapula position.

Materials and research methods. The object of the study was 82 children with a congenital high scapula, treated in the department of pediatric orthopedics of the State Institution RSSPMCTO of the Ministry of Health of the Republic of Uzbekistan for the period from 2005 to 2020. 84 operations were performed in 82 children. The repeated operations were performed in 2 children due to partial recurrence of the pathology.

The clinical designs, radiological, MSCT and ENMG research methods in the diagnosis of Sprengel disease were applied [1,2,3,4]. Clinical and X-ray examinations were performed in all patients, MSCT - in 42, ENMG - in 34 children.

Our observations have shown that in the process of choosing the volume of surgical accommodations and achieving treatment results, the shape of the scapula is important. After analyzing the performed X-ray and MSCT studies, the patients were divided into 4 groups [Fig. 1]:

1. There were 4 patients with mild degree of Sprengel disease without deformation of the scapula.
2. There were 31 patients with concave deformation of the scapula along the length.
3. There were 21 patients with concave deformation of the scapula across the width.
4. There were 26 patients with mixed deformity of the scapula.

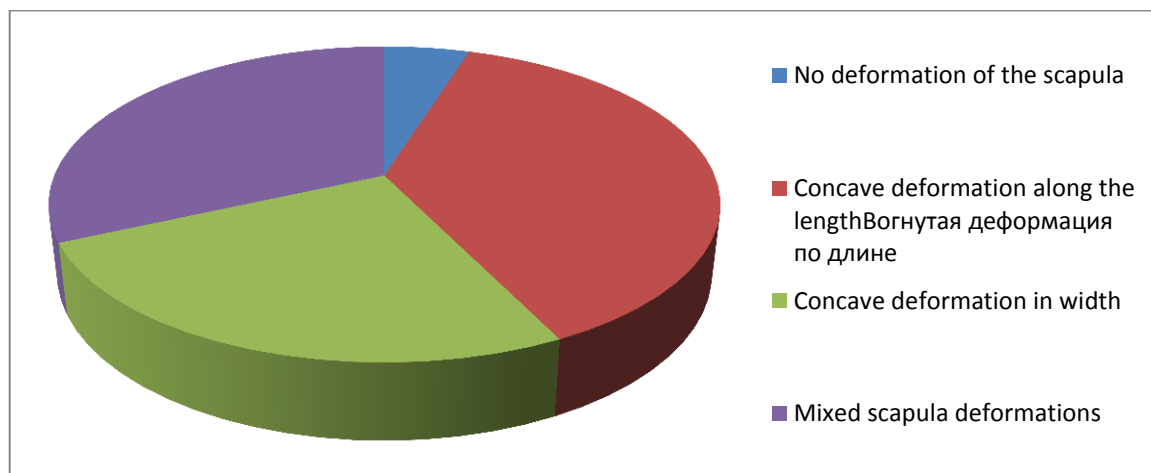


Fig. 1. Distribution of patients depending on changes in the shape of the scapula (description in the text).

Among our patients, children with concave and mixed deformation of the scapula prevailed. Therefore, all these children in the process of surgical treatment were need to correct the shape of the scapula. Sprengel disease without deformity of the scapula were observed only in children of younger age groups, with soft tissue and mild deformity. We consider that with the growing due to the disturbed balance of forces of various muscle

groups the secondary deformities of the scapula are developed over time in children. Therefore, early diagnosis and early radical treatment of Sprengel disease in children is the prevention of the development of secondary deformities and the progression of the pathological process.

We consider all cases of Sprengel disease to be an indication for surgical treatment in the absence of a visible positive effect of conservative treatment in children over 3 years old. Conservative treatment can be provided in children under 3 years and, in general, it is a preparatory stage before the main surgical treatment.

The surgical treatment of the high standing of the scapula was performed in the following order: first of all, the scapula was mobilized, specifically its medial edge, the superior medial and inferior angles, the anterior and posterior surfaces of the scapula, then pathological rotation was eliminated with bringing the scapula down to a healthy level, the adaptation of the scapula surface to the chest was checked, the scapula was fixed with stitching of its lower angle in the "pocket" between the chest and the broadest muscle of the back [8,9,10,11,12,15].

Our observations have shown that in order to prevent the development of scapula deformities as children growing, it is advisable to assess comprehensively the state of changes in the shape of the scapular bone and make corrections during surgery. The methods for correcting hook-shaped, concave and mixed deformities of the scapula are proposed. The essence of the method is that after complete mobilization of the scapula with the use of bone holders, we make a fracture of the scapula according to the type of "fracture of the green branch" and fix the fragments with intercrossing knitting needles for 3 weeks. We applied the technique for the surgical correction of scapula deformity in 42 children (Fig. 2).

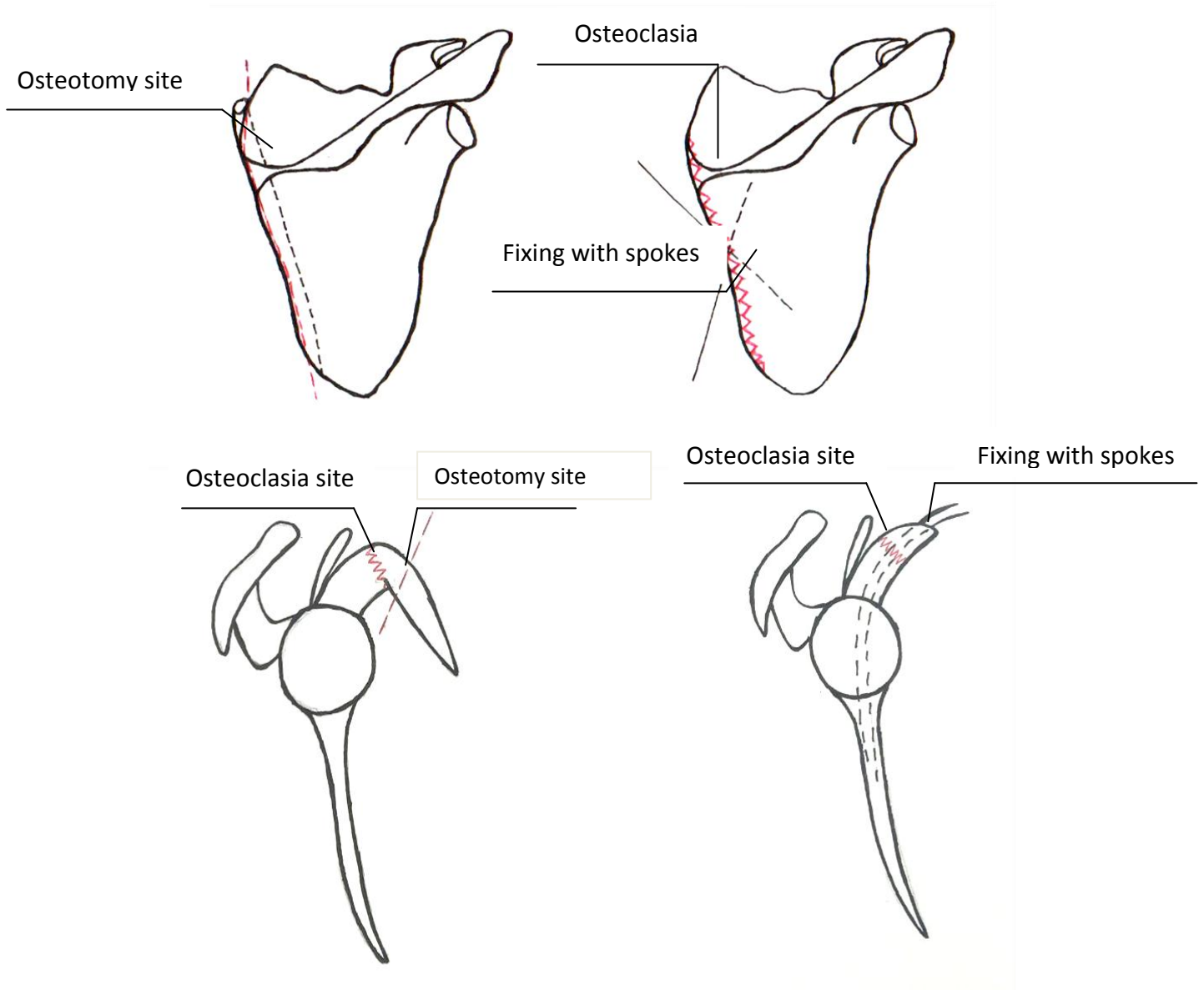


Fig. 2. Method of correction of concave deformation of the shoulder blade in width and length (description in the text).

We consider it is appropriate to divide the postoperative management of patients into four periods:

1. Early postoperative period is up to 1 week
2. Second postoperative period is up to 3 weeks
3. The first rehabilitation period after surgery is from 1 month to 3 months
4. The period of medical and social rehabilitation of children after surgery is from 3 months to 1 year.

Timely implementation of rehabilitation measures with the use of physiotherapy and exercise therapy are allowed to obtain good treatment outcomes.

The treatment outcomes for children with congenital high scapula position were assessed by the restoration of the anatomical and cosmetic shape and functional state of the upper limb [5, 6]. Our observations showed that the anatomical and cosmetic results after surgery in children with a high scapula position depended on the severity of the disease, the presence of concomitant diseases and the operations performed [13, 14, 16]. Long-term results of treatment were studied in 74 children after surgical treatment in the period of one to 14 years. Among these, there were 34 children from the first group underwent surgery without scapula reconstruction. And there were 40 patients from the second group, who underwent surgery for lowering the scapula with reconstruction of the shape of the scapula.

The anatomical and cosmetic results of the treatment were assessed according to a three-point system: good, satisfactory, and unsatisfactory.

The restoration of the anatomical and cosmetic shape of the neck and shoulder girdle was assessed by the symmetry of the right and left halves of the neck, by the height of both shoulder blades, by the symmetry of both halves of the chest, the ratio between the scapula and the spine, the comparative state of the scapular regions of the back, the state of the postoperative scar, the posture of the child, the presence of rotation and cranial mixing of the scapula.

Good anatomical and cosmetic results included when in patients, the right and left sides of the neck were symmetrical, and the cranial mixing of the scapula was up to 1 cm, the both halves of rib cage were symmetrical, the medial mixing of the scapula was insignificant and invisible on clinical examination, the scapular regions were symmetrical, postoperative scar smooth, the child's posture was correct, in addition, on the X-ray and MSCT there was no rotation of the scapula and cranial mixing up to 1 cm from the lower edge of the 2nd thoracic vertebra.

Anatomical and cosmetic results were considered satisfactory: both halves of the neck were symmetrical or there was slight asymmetry, the cranial mixing of the scapula was up to 1.5-2.0 cm, the thorax - the right and left halves had noticeable asymmetry, the scapular regions were symmetrical, the medial edge of the scapula protruded, Posture - residual lateral curvature of the spine, on X-ray and MSCT scapula rotation was up to 5-6 degrees, cranial displacement was up to 1.5-2.0 cm.

Of all 74 patients examined in the long-term period after treatment, we obtained good results in 75.7% of cases, satisfactory in 21.6%, and unsatisfactory anatomical and cosmetic treatment results in 2.7% (Fig. 3).

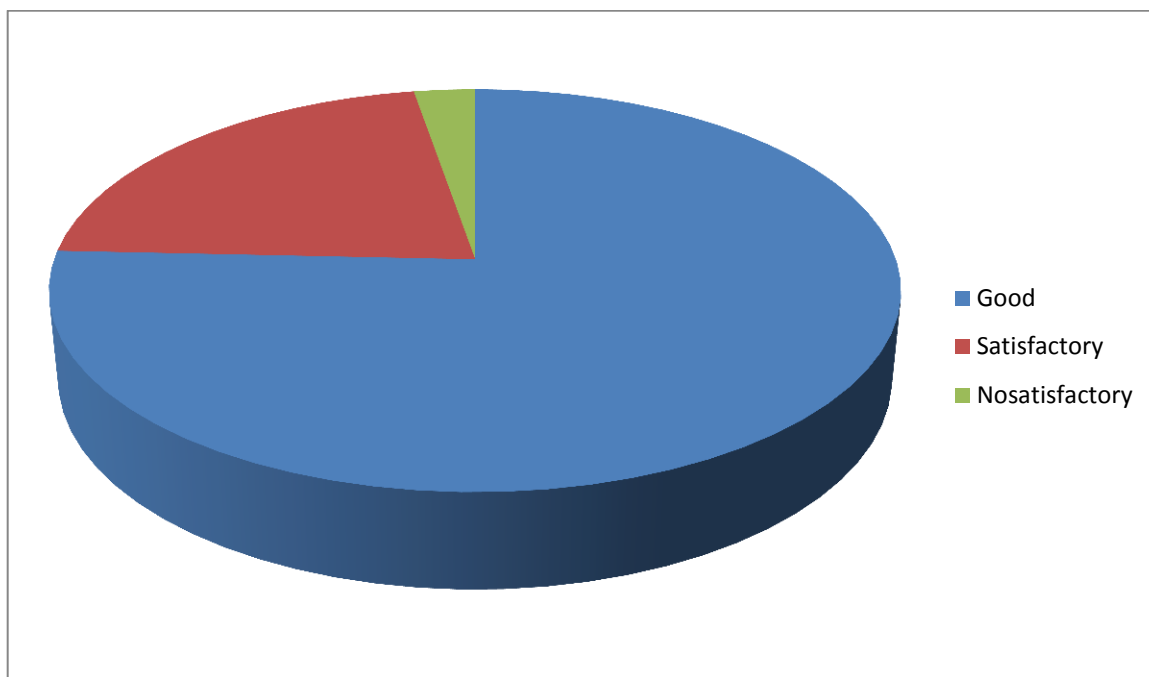


Fig. 3 Indicators of the results of surgical treatment of Sprengel disease.

The functional results of surgical treatment of Sprengel disease were assessed according to the following criteria:

The amount of abduction of the shoulder and upper limb, the presence of pain in the shoulder joint and in the area of the scapula during movement, the ability to use the upper limb for self-care and exercise, the presence or absence of neurological changes in the upper limb.

The overall assessment of the functional results of treatment was also carried out according to a three-point system: good, satisfactory, and unsatisfactory.

Functional results were considered good when:

abduction of the shoulder and upper limb in the shoulder joint up to 160 degrees, pain during movement of the upper limb is absent, the upper limb is used for self-service in full, there are no neurological disorders.

The functional results were considered satisfactory when:

1. Abduction of the shoulder and upper limb in the shoulder joint is up to 140 degrees
2. There is mild pain in the scapula or shoulder joint with maximum abduction and rotation of the shoulder

3. The upper limb is used in full self-service

4. Neurological disorders: at times numbness in the fingers and paresis are noted.

Good functional results of treatment were obtained on average in 81.0% of patients, satisfactory - in 16.2%. Comparative analysis of the two groups of patients showed that the functional results of treatment depended on the reconstruction of the shape of the scapula. The adaptation of the scapula to the chest surface allows the scapula to slide smoothly during shoulder abduction. If in the first group of patients we noted good results in 76.4% of cases, then after the reconstruction of the scapula good results were observed in 85.0% of patients.

As an example, we present our observation with a good result of a patient with Sprengel disease on the right (Fig. 4).

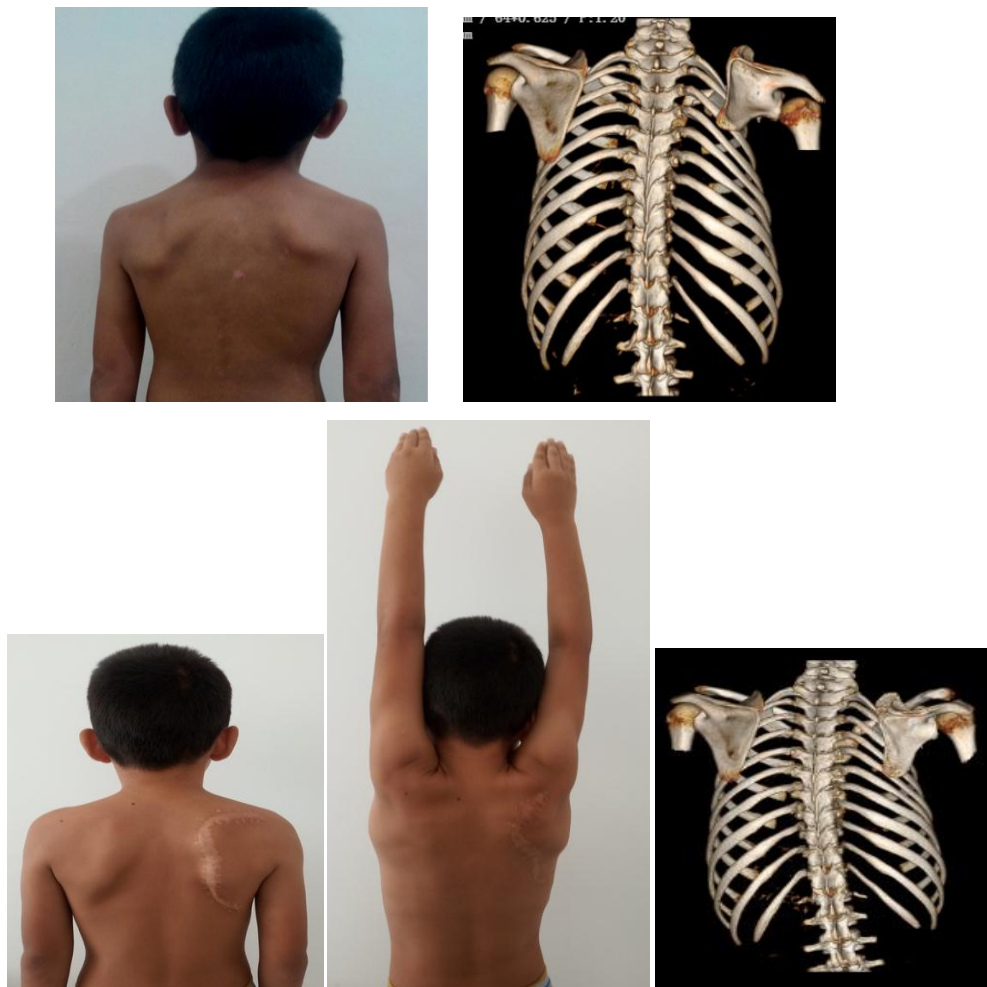


Fig. 4. Umurzakov From 8 years old photo and MSCT examination before and after surgery

The second observation with a satisfactory result of a patient with Sprengels disease on the left (Fig. 5).

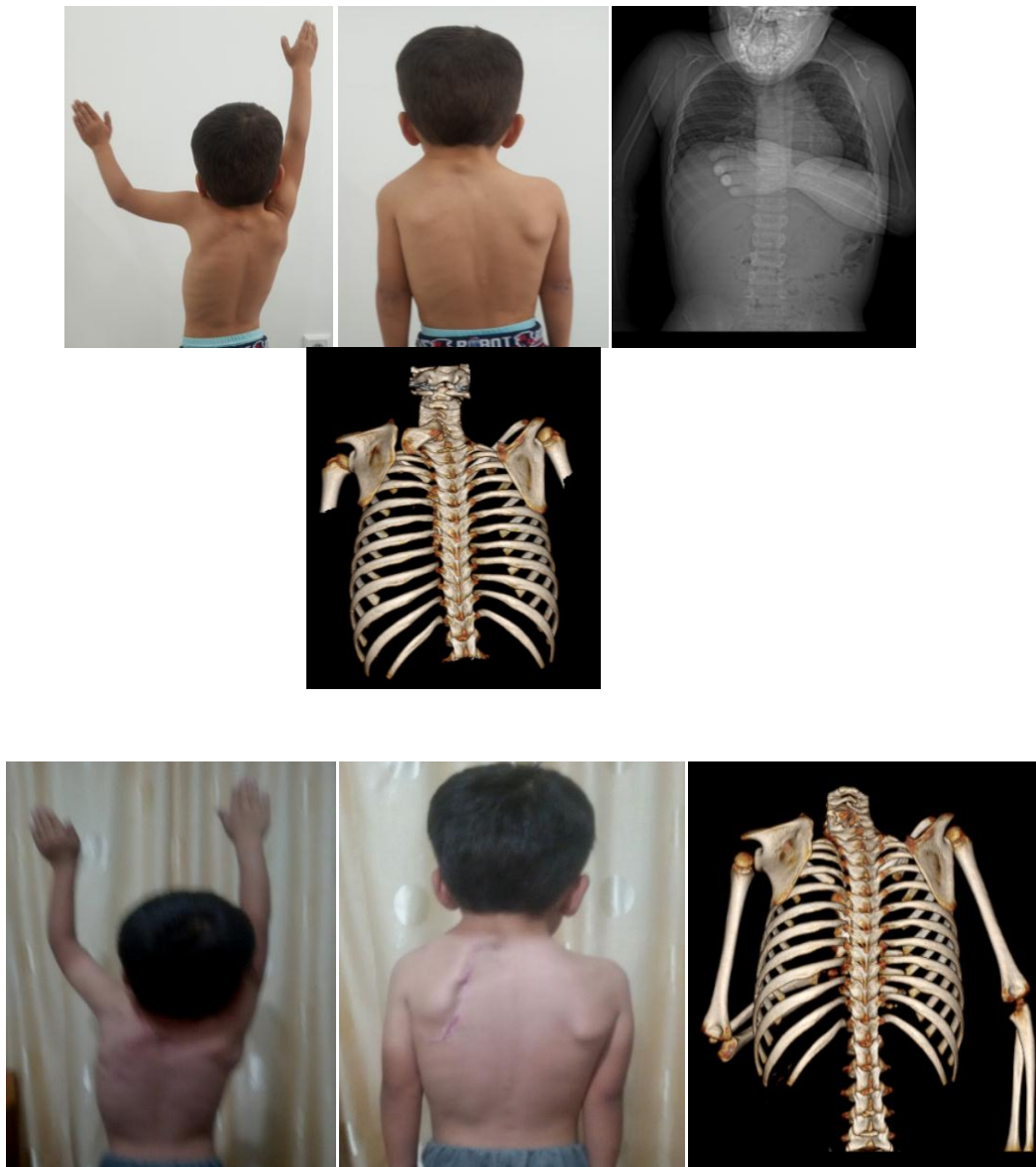


Fig. 5. A. Alokhonov 6 years old image and MSCT examination before and after surgery

Good treatment results have been achieved in children under 10 years of age and with soft tissue forms of the disease. Due to the presence of severe pathological changes in the scapula, bones of the chest and shoulder girdle with bone forms, the restoration of the function of the upper limbs may be difficult. In addition, as children growing, the secondary deformities of the spine, chest, scapula are added to the underlying disease, which

complicates the course of the disease. Consequently, it is advisable to perform surgical treatment of Sprengel disease in children under 7-10 years of age.

Thus, the surgical treatment of the congenital high standing of the scapula is a difficult task. The successful completion of this task requires a comprehensive examination of the child using modern methods for diagnosing the condition of the skeletal system, innervation, muscles of the shoulder girdle and shoulder joint. After a comprehensive assessment of the patient's condition, a plan of surgical intervention should be drawn up. Full mobilization from the surrounding tissues, strands and bringing down the scapula to a normal level is accompanied by significant tissue trauma and requires careful postoperative care for a sick child. Systematic postoperative rehabilitation treatment using the entire arsenal of physiotherapy and exercise therapy makes it possible to restore the functionality of the upper limb. To facilitate the uniform sliding of the scapula along the surface of the chest, the method of option is our proposed reconstructive surgical correction of the pathological shape of the scapula.

CONCLUSION:

1. The severity of anatomical and functional disorders in congenital high standing of the scapula is in direct depending on the changes in the shape of the scapula. In children with Sprengel disease, pathological changes in the shape of the scapula are observed in 95.1% of cases.

2. X-ray and MSCT studies revealed 4 types of changes in the shape of the scapula in Sprengel disease: without deformity, with a hook-shaped deformity of the supraspinatus, with concave deformity of the body and with mixed deformity of all parts of the scapula.

3. Surgical treatment of Sprengel disease is advisable to carry out differentiated, taking into account the form of the disease and pathological changes in the scapula. A prerequisite is the complete correction and adaptation to the surface of the thorax of the proximal region and the body of the scapula.

4. The proposed methods of correction of the pathologically altered shape of the scapula allowed to improve the results of surgical treatment of children with Sprengel disease. We achieved good anatomical results in 80.0% of observations and functional results in 85.0% of children.

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