

Surgical Correction of Chest Deformity in Currarino - Silverman Syndrome (Clinical Case)

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Abstract

Currarino-Silverman syndrome (combined chest deformity - DHA) belongs to the red kim forms of DHA. Often this type of deformity is combined with malformations of the heart (coarctation aorta, prolapse, mitral stenosis). The article identifies two types of combined DHA, which allows you to determine the tactics of surgical treatment. Most promising in terms of the possibility of surgical correction type 2 deformity with protrusion of the manubriosternal synovial laxness, but without retraction of the sternum. The paper presents a description of successfully completed operations in 3 patients with combined DHA, in one of whom the pathology was combined with Poland's syndrome.

Keywords: *Currarino-Silverman Syndrome; Thoracoplasty*

Introduction

Currarino-Silverman syndrome, also known in the literature as a combined deformation thorax (DHA) or a special type of keeled noah DHA (pigeon breast, rectus sarinatum type 2 deformity or upper pectus carinatum), refers to ed- to these forms of DHA and is characterized by early syn- stasis of parts of the sternum, stopping the growth of the latter and, as a consequence, secondary DHA. It manifests itself protrusion of the manubriosternal articulation and the fall of the middle and lower third of the sternum. More often there are symmetrical, less often - asymmetric forms of combined DHA. May be present other concomitant developmental disorders musculoskeletal system such as scoliosis, and in rare cases Poland's syndrome [1-4].

Case Report

The first to describe the combined form of DHA and proposed a technique for her surgical by M. Ravitch in 1952 [4,5]. He considered this deformation within the keeled DHA, you are dividing it into a separate species and calling it "chest naked bya" (pigeon breast). The operation consisted of sub- perichondral resection of 2-7 costal cartilages on both sides, cutting off the xiphoid process and anterior wedge-shaped sternotomy at the level the greatest standing of the sternum. Subsequently the sternum was sutured at the level of the superior sternotomy, thereby eliminating its arched deformation, lowered to a normal level and to the front the surface of the lower third of the body of the sternum is fixed drew the xiphoid process on a muscle knife ke. Additionally, the perichondral of resected ribs. No fixes tori of the sternocostal complex (HRC) while were not used.

In 1958 G. Currarino and F. Silverman [6] for the first time noticed the premature closure zones of growth of the sternum. They also pointed to a large the percentage of combined with a combined type of DHA malformations of the heart (coarctation of the aorta, laps, mitral valve stenosis) and the predominance in this category of female patients. S.S. Rudakov, *et al.* [1,7] consider com- binned DHA as a

manifestation of malformation or true malformation. This is indicated by manifestation of the disease immediately after birth and the absence of rapid progression, i.e. the deformity progresses as the child grows. The main indication for surgery, according to auto- moat, there is a cosmetic defect.

In the literature, the description of this pathology is single messages are sacred. So, in the database PubMed found only three sources of literature with a description of the combined DHA. At- than in two of them [3,8], the emphasis was placed on treatment diagnostics of heart defects in Currarino syndrome- Silverman without mentioning DHA correction. Articles R. Shamberger and K. Welch [9] and A.A. Pe- Chetova., *et al.* [10] described 5 and 3 patients, respectively. naturally, those operated on for Currarino syndrome- Silverman using the Ravitch method. Remote the results of operations are not shown.

Given the traumatic nature of the operative chastity, high frequency of concomitant defects development of the heart, kidneys and other internal organs new, surgery to correct DHA in patients with rarely accepted. Indication for surgery in pain In most cases, there is a cosmetic defect. In this study, two types of combined DHA: type 1 - with protrusion of manubriosternal articulation and retraction of the middle her and the lower third of the sternum and type 2 - with protruding zia of the manubriosternal articulation, without a sternum. Based on the type of deformation, lined up surgical tactics of treatment. At the 1st type of combined DHA performed the operation according to the Ravitch method. More promising for chirurgical correction is considered the 2nd type, since ku in this case, it is possible to perform less traumatic surgical intervention. Opera- the walkie-talkie is limited only to the subperiosteal removal of a deformed fragment of the sternum with its replacement with a graft or stitching fragments of the sternum.

We present our own experience in surgical treatment with combined DHA in 3 patients. Patient B., 33 years old, was admitted in July 2014 in the 10th department of the CITO them. N.N. Priorov with a sting bami for the presence of coarse DHA and the resulting cosmetic defect (Figure 1a). DHA appeared from birth and progressed in proportion the growth of the body. Heredity is not burdened.

When viewed, starting from the level of the 2nd rib, on- an arched standing of the sternum and cartilage was observed cheek divisions of the 2nd to 4th ribs. Middle and lower these sternums are positioned correctly, without a tendency to the sinking. The costal arches will not stand up. The patient underwent a complex preoperative examination. Mitral prolapse was detected. 1st degree leg valve, no regurgitation.

The operation was performed on 07/08/14. In the middle line the sternum was accessed 10 cm long bilized the pectoral muscles within the zone of de- formations. Subperchondral resection was performed 2 - 4 costal cartilage on both sides with bone part, the sub- periosteal resection was the bathroom of the sternum for 6 cm. In the bed the removed fragment of the sternum was placed fragments of bone tissue. The price was restored flatness of the anterior plate of the sternum by separate seams. Carried out knocking down of the perichondrium rese- cited costal cartilage by Ravitch. Was before- the correct configuration of the sternum is stiffened. Breast- the cage was fixed in a corset. Postoperative the operation period was uneventful, on the 9th the patient was discharged for an outpatient treatment (See figure 1b).

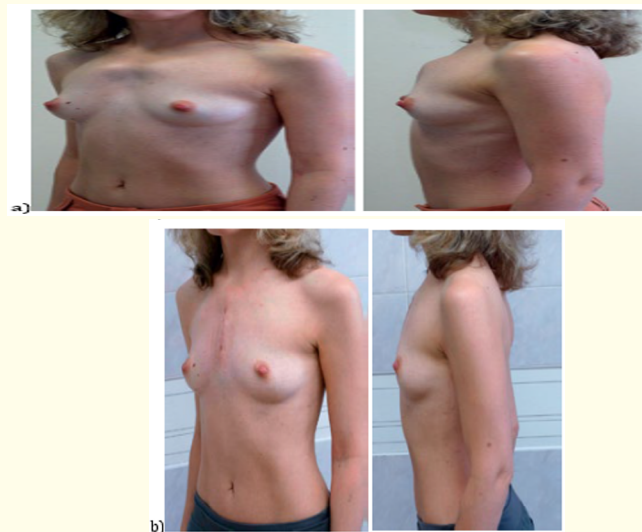


Figure 1: Appearance of patient B., 33 years old, with combined DHA of symmetrical form (type 2) before (a) and 8 days after (b) operations.

Patient M., 17 years old, was admitted in June 2017 with foreheads for the presence of coarse DHA and the resulting cosmetic defect (Figure 2a). DHA from birth. When viewed, starting from the level of the 3rd rib, on- an arched standing of the sternum and cartilage was observed cheek divisions of the 3 - 5th costal cartilage. Bottom depression of the sternum without a tendency to retraction (Figure 2b).

The examination was diagnosed with pro mitral valve laps of the 1st degree with moderate noisy mitral regurgitation. The operation was carried out on 06/30/17. Average the sternum line was accessed with a length of 10 cm. The pectoral muscles were mobilized within lax of the deformation zone. With technical difficulties sub perichondral resection was performed 3 - 5 costal cartilage on both sides with bone part, the subperiosteal resection was bathroom part of the sternum for 6 cm. Fragment the cops of the sternum were sewn together with wire. Was re- the integrity of the anterior plate of the din, and also achieved the correct configurations of the sternum.

The postoperative period passed without complications. neny, on the 10th day the patient was discharged for a bulat treatment. When viewed after 3 months: forehead no. The correct configuration was stated thorax and consolidation of the sternum in the area osteotomy (Figure 2c).

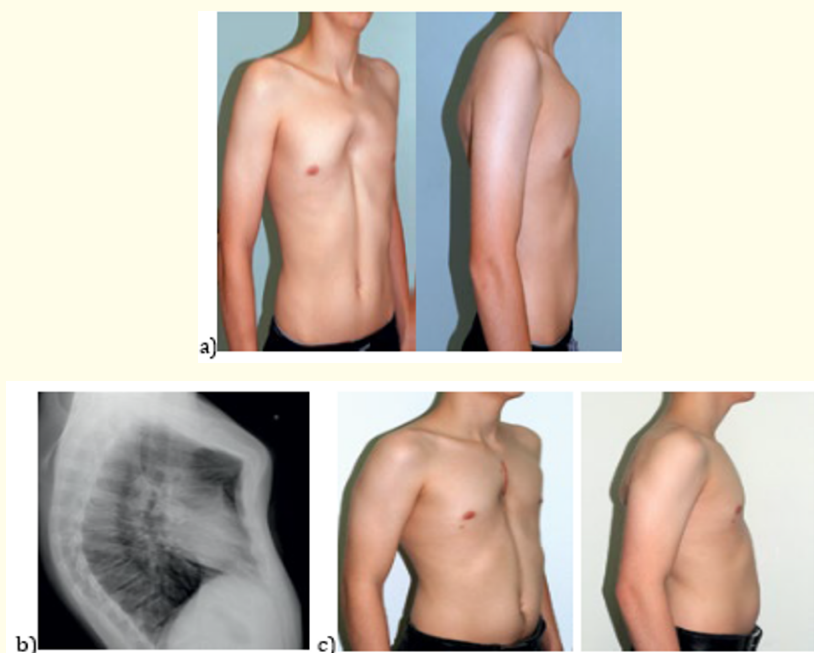


Figure 2: Patient M., 17 years old, with combined symmetric DHA (type 2). a - appearance before surgery; b - chest x-ray in lateral projection; c - appearance 3 months after operations.

Patient K., 16 years old, in March 2018 was admitted to the 10th Department of the Central Institute of Traumatology and orthopedics. N.N. Priorov with complaints about the presence of rough DHA and the resulting cosmetology defect (Figure 3a). Deformation from birth. On examination: the left half of the chest is not fully developed. Large and small pectoral muscles on the left aplastic. Starting from the level of the 2nd rib, observe were given an arched sternum with a helical deformation and endurance of cartilaginous divisions 2 - 5 costal cartilages, more on the right. Lower from- affairs of the sternum without a tendency to retraction (Figure 3b). The costal arches will not stand up. The upper limbs are the whites are symmetrical, their function is not disturbed.

Discussion

When examining concomitant pathology has not been identified. Was diagnosed with a combination need DHA, asymmetric right-sided the form. Poland's syndrome on the left (large aplasia and pectoralis minor). They began to prepare the patient to the operation. The

operation was performed on 03/22/18. Wednesday her sternum line was accessed 10 cm. The pectoral muscles were mobilized in the pre- affairs of the deformation zone. Conducted sub perichondrium lateral resection of the 2nd to 7th costal cartilage on the right and 3 - 5th - on the left. The ribs from 2nd to 5th on the right were resected with the bone part. Carried out under- periosteal planar resection of deformity sternum, knocking of the perichondrium resected costal cartilage. Was achieved chickpea the correct configuration of the bone framework chest. According to the marking lines, a sub- skin pocket-bed, in which the pectoral implant (POLYTECH) with a volume of 231 ml. The ribcage has acquired the correct configuration. walkie-talkie. The postoperative period was uneventful. Neni, on the 8th day the patient was discharged to the ambulance late treatment. When viewed after 4 months: female forehead no. Chest configuration is correct form (Figure 3c).



Figure 3: Patient K., 16 years old, with combined DHA of an asymmetric right-sided form and Poland's syndrome on the left. a - appearance before surgery; b - CT data of the chest: there is an arcuate, helical deformity of the sternum, pathologically wide, ossified sternum, aplasia of the pectoral muscles on the left; c - appearance 4 months after surgery.

Conclusion

Thus, given in our standing article clinical observations of a surgeon for the treatment of a rare congenital pathology chest are of clinical interest res and deserve attention. Correct understanding obsession with the essence of the pathological process underlying at the heart of the development of combined DHA, allowing allows the use of adequate surgical Sobie. High risk perceptions of complications and the traumatic nature of the operation in this group of patients tov are exaggerated.

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