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SUBTOTAL CONGENITAL STERNAL CLEFT

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СУБТОТАЛЬНАЯ ВРОЖДЕННАЯ РАСЩЕЛИНА ГРУДИНЫ

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Sternal cleft (SC) is a severe and rare congenital anterior chest wall defect associated with impaired sternal embryogenesis. Complete and incomplete forms of SC are distinguished depending on the degree of cleavage. The clinical significance of the defect is that along with a cosmetic defect of the sternum, the heart, and great vessels are protected only by the skin. The article presents a clinical observation of patient K., who was born with a subtotal SC. The child underwent radical surgery at four months. He was examined long-term (8 years after the operation). The boy is developing harmoniously according to his age. An invisible normotrophic scar has formed in the area of the postoperative wound. No chest deformation is detected.

Keywords: congenital sternal cleft, subtotal cleft, case report, children

Расщелина грудины (РГ) представляет собой редкий и серьезный врожденный дефект передней стенки грудной клетки, связанный с нарушением эмбриогенеза грудины. В зависимости от степени расщепления выделяют полную и неполную форму РГ. Клиническое значение порока состоит в том, что, наряду с косметическим дефектом грудины, сердце и магистральные сосуды защищены лишь кожей, что представляет опасность для жизни ребенка. В статье представлено клиническое наблюдение пациента К., который родился с субтотальной РГ. Ребенок был радикально оперирован в 4 месяца. Осмотрен в отдаленном периоде (через 8 лет с момента операции). Мальчик гармонично развивается согласно возрасту. В области послеоперационной раны сформировался незаметный нормотрофический рубец. Деформации грудной клетки не определяется.

Ключевые слова: врожденный порок, субтотальная расщелина грудины, клинический случай, дети

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CCWD – congenital chest wall deformities
ECG – electrocardiogram
EchoCG – echocardiography
SC – sternal cleft
US – ultrasonography

The clinical significance of the sternal cleft (SC) is that the cosmetic defect, heart, and trunk vessels are protected only by the skin, which is a danger to the child's life [1, 2]. SC is often associated with congenital disabilities such as craniofacial hemangioma, omphalocele. Heart defects are generally absent in patients, and pericardium and skin integuments intact [3].

Clinical case. Patient K., 09.09.2015, was born with a body weight of 3400 g from 2 pregnancies and two births. The first child, the boy – healthy. After being discharged from the maternity hospital, the child was referred for consultation to the Republican Scientific and Practical Center of Pediatric Surgery. The examination revealed a V-shaped subtitle SC (Fig. 1). The pulsation of the heart and large vessels was visible under the skin of the breast defect.

Fig. 1. Patient with SC at first consultation. Age 12 days

An open oval window was found during the EchoCG. No other pathology was found. The patient is diagnosed with congenital chest wall deformities. The newborn was placed under the supervision of a pediatrician and a pediatric surgeon in the community. The patient was recommended to report for surgery within 4–5 months.

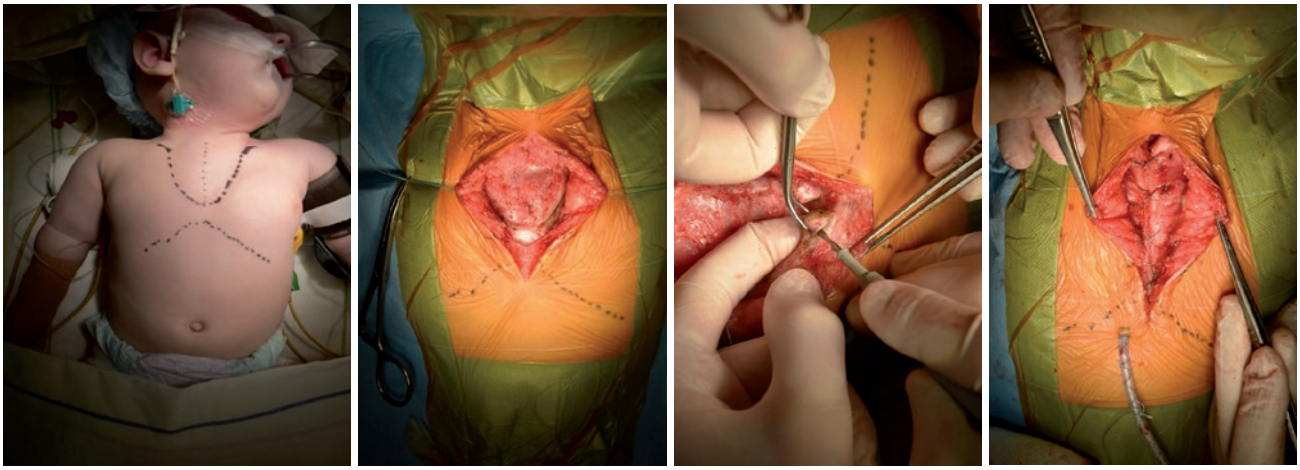
At the age of 4 months (weight 7200 g), he was hospitalized. At the time of admission, the child's condition was compensated. Physical development and body mass are consistent with age norms. Local status: A V-shaped defect in the bone of the sternum with preservation of integrity (contact) only in the region of the xiphoid process. There is a visible pulsation of the heart.

To eliminate the associated congenital abnormalities, the baby had an ultrasonic ultrasound of the abdominal cavity, EchoCG of the heart, ECG, and chest X-ray. An open oval window measuring 4.5 mm was noted, corresponding to the child's age. There were no indications of surgery.

The course of the operation. Under the endotracheal anesthesia, the thorax is marked for operational access (Fig. 2A). After that, median access to the sternum was made. Upon examination, the sternum was found to be almost entirely missing. Diastasis of edges is up to 6 cm. A small (1.0 cm) bridge connects the edges of the sternum in the area of the xiphoid process (Fig. 2B).

The edges of the sternum are removed bluntly and sharply. The lower edge of the sternum is partially resected (Fig. 2C). The edges of the defect are treated with a Volkman spoon. The sternum is stabilized with wire seams (Fig. 2D), and the front mediastinum is drained. There's an intradermal suture on Halstead.

The postoperative period went without features. Immediately after entering the intensive care unit, the child is extubated. There were no signs of «close mediastinum» syndrome. The drainage was removed 5 hours after the operation. Six hours later, the patient was transferred to the cardiac department. The child was discharged for five days after the surgical intervention and is in satisfactory condition.



A B C D

Fig. 2. Stages of the operation for surgical correction of SC:
A – marking chest landmarks for surgical access; B – appearance of the cleft sternum after mobilization;
C – resection of the lower edge of the sternum; D – bringing together the edges of the sternum with wire sutures

The patient was examined eight years latter after the operation. The child develops harmoniously according to age. In the postoperative wound area, the rupture is almost invisible. Palpating edges of the sternum are matched and motionless, and diastasis is not determined. There is no thorax deformation (Fig. 3).

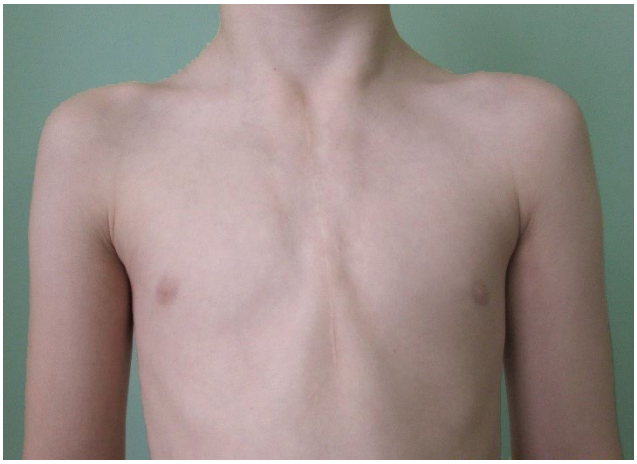


Fig. 3. The patient is eight years after surgery

The child performed a clinical tool study. The ECG shows a sinus rhythm of 80 beats per minute, the vertical position of the electric axis of the heart. According to EchoCG, a functioning oval window remains, hemodynamically insignificant, with no signs of hypervolemia of the small blood circulation.

Surgical treatment of SC is preferable in the chest due to its excellent flexibility. Surgical correction is represented by two main methods of treatment depending on the patient's age. If the defect is operated on in children for up to one year, the surgical correction consists of stitching the sternum along the middle line [4]. In children over one-year-old, a thoracoplasty is used. Furthermore, the sternum is reduced after partial excision and supplemented by a partial parasternal oblique chondrotomy to increase the thorax volume [5, 6]. However, both approaches partially reduce the chest volume to some extent. Therefore, the first postoperative days are critical and require the close attention of pediatric surgeons and intensive care physicians.

Conclusion. Thus, in the surgical treatment of SC, osteosynthesis should aim at a complete comparison of the edges with a minimum loss of the thorax volume. Especially pays attention to the prevention of heart and lung compression. Failure to do so may result in hemodynamic, respiratory failure, and death.

In our clinical case, we present successful treatment of the subtotal splinter of the sternum by treating the edges and partial resection of the body of the sternum with a complete comparison of the edges without complications. The remote period of observation showed not only a successful correction of the SC with the restoration of normal thorax anatomy but also allowed to ensure the child's good quality of life.

Disclosures: The authors declare no conflict of interest.

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