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# Congenital sternal cleft — a case report

## Wrodzony rozszczep mostka — opis przypadku

### Abstract

Sternal cleft is a very rare congenital defect of the sternum, reported only in a limited number of publications. Surgical treatment in the neonatal period is preferred. A case of a child with congenital sternal cleft operated on at 17 months of age with a successful outcome of repair was reported.

**Key words:** sternal cleft, surgery, congenital anomaly

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### Introduction

Congenital sternal cleft is a malformation of superior celosomy type and affects the anterior side of the chest wall [1]. It may be an isolated defect or associated with other malformations of the chest and internal organs. Isolated congenital sternal cleft is a very rare defect, which is a risk factor of cardiac injury, herniation of mediastinal organs and leads to the instability of the chest, but usually does not disrupt the mechanics of breathing. This is an evident cosmetic defect. The operative treatment is recommended at an early age because of high flexibility of the chest.

### Case report

The boy was born on 6 June 2009, II pregnancy at 38 weeks, II delivery (Reg. No.: 141/2008,8860/2009). The parents were healthy, without addictions: the mother was 27 years old, the fa-

ther was 34 years old. The mother had an infection of the respiratory tract in the 5<sup>th</sup> and 8<sup>th</sup> month of pregnancy which was treated symptomatically. She was under gynaecological supervision during the pregnancy and the prenatal tests were not performed. The 7-year-old brother of the child is healthy. The boy was born in good condition (APGAR 9/9), with competent circulatory and respiratory systems, body mass of 3220 g, body length of 56 cm. During the first physical examination of the newborn, there was seen a several-centimetre-long longitudinal concavity of the chest in the anterior midline, evidently visible during inspiration. The pulsation of the heart was felt on palpation of the chest wall in this area. The findings suggested a congenital defect in the anterior chest wall in the sternal area. That was the reason for admitting the child into the Department of Neonatal Intensive Care and Neonatal Pathology at Medical University of Silesia in Zabrze. The chest radiogram revealed

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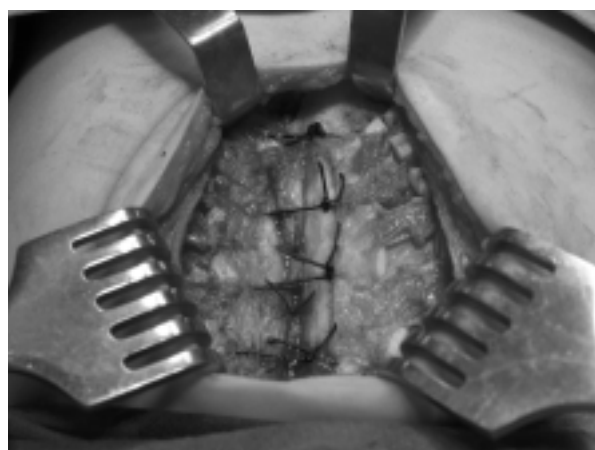


**Figure 1.** Chest CT scan – 3D reconstruction. Angular position of clavicles, no ossification of sternum

incorrect, oblique position of clavicles and no sternal ossification nuclei. The definitive diagnosis of congenital sternal cleft was established on the basis of thoracic computed tomography (Fig. 1). The defects of the heart and great vessels were excluded in echocardiography. Ultrasound examinations of the brain and abdominal cavity in this newborn were within normal limits. Because of good general condition of the newborn and isolated character of the defect, the surgery was postponed. After discharge from the Department, the child was referred to the outpatient surgical clinic where it was systematically controlled. The boy was treated ambulatory during his first year of life for respiratory tract infections, including bronchitis. His psychomotor development was normal. The parents were concerned about the risk of a direct heart injury as a result of the defect, so they decided for surgery when the child was 17 months old. The surgery according to the Ravitch technique modified by Ribas was performed in the Department of Malformation Surgery and Traumatology at Medical University of Silesia in Zabrze. The intraoperatively dissected sternum was similar to the V letter, consisted of 2 cartilaginous plates connected together in the distal part (Fig. 2). The sheets of perichondrium were dissected, the lower part which connected the cleft ledges was removed and the cartilaginous ledges were sutured together in the median line to approximate the cartilaginous plates (corresponding in the development to the body and manubrium of sternum). The parasternal cartilaginous fragments of ribs



**Figure 2.** Congenital sternal cleft. Image after surgical thoracotomy



**Rycina 3.** Congenital sternal cleft. Image of a “new” sternum after surgical reconstruction

I–VI were resected according to the Ravitch technique to reduce the tension on the suture line [2]. The sheets of periosteum were used to reconstruct the posterior layer of the sternum. In the median line, between the sutured bone ledges, the resected cartilaginous rib fragments were placed to reinforce the restored sternum (Fig. 3). Pneumonia with respiratory failure, which occurred during the postoperative period, was treated in the intensive care unit. The boy was extubated after 8 days of therapy and in good condition; after a short stay on surgical ward he was discharged home. At present, the child is 2.5 years old, develops normally and is under supervision of outpatient surgical clinic. The mother reports that since the operation the boy has not developed bronchitis yet (as before the operation). The postoperative scar in the anterior median line healed with a good cosmetic result. On palpation, the reconstructed sternum is ossified.

## Discussion

The sternum develops in the 10<sup>th</sup> week of foetal life from consolidation of 2 bands of mesodermal cells [3].

The sternal cleft was previously known as sternal agenesis. After considering the embryogenesis of the sternum, this term is incorrect, because in the place of a “missing” sternum, there are always two unconsolidated laminae. The presence of these laminae practically determines the reconstructive surgery. The causes of this defect are unknown. In the experimental work of Ramirez-Solis et al. [4] the HoxB-4 gene was inactivated *in vitro* in a mouse during the disruption. On the basis of own observations, the authors of this report connected the manifestation of sternal cleft with the inactivation of HoxB-4 gene. The frequency of congenital sternal cleft has not been established yet. This is a very rare defect. The first case report was published in 1740 [5] and between 1800–2007 only 73 cases were reported [6]. In the work of Ribas, who described 8 patients aged from 18 months to 19 years, the congenital sternal cleft was more common among girls [7]. The isolated congenital sternal cleft may be classified as total or partial (superior or inferior). The more common superior sternal cleft is the V-shaped one (the cleft reaches the xiphoid process) or the U-shaped one, when a wide defect in the place of the sternum is closed with a distally located osseous fragment, which connects the ends of third or fourth rib on both sides [3]. In the case described in this paper, a V-shaped defect was found. The detection of congenital sternal cleft, which is already possible in prenatal ultrasound, is not difficult during observation and palpation of the newborn. The diagnosis of congenital sternal cleft as a benign isolated defect does not allow for resignation from searching for other defects. The congenital sternal cleft may be associated with chest deformities, for example chicken breast or is an element of other malformation syndromes such as pentalogy of Cantrell (defects involving skin, sternum, diaphragm, pericardium and heart) or the Leiber syndrome (angiomas and teleangiectasias) [8] or is associated with defects of great vessels (the subvalvular aortic stenosis). The concomitance with the pentalogy of Cantrell is a lethal type of defect. There was described one case of congenital sternal cleft as an element of the VACTERL syndrome (defects involving vertebrae, anus, heart, trachea, oesophagus, kidneys, limbs) [6].

Lannelongue was the first one who surgically corrected the defect in 1888 [7]. If the congenital sternal cleft is associated with other chest defects, an early surgical treatment is recommended (until 3 months of life, optimally during the first month)

due to high chest flexibility and good adaptation of vessels, heart and lungs to the compression of a closed scaffold of the chest wall. The first report of successful surgery on a newborn, a few days old, comes from India [9].

In the described patient, surgery was postponed on the basis of the isolated character of the defect. The choice of the time of surgery during the period of increasing motor activity of the child appears to be justified by the possibility of an injury of the relatively “opened” precordial space. The decisions about surgery in teenagers were based on cosmetic reasons. In the work of Ribas, the mean age of operated patients was 7.2 years [7]. The authors stress the fact that surgery performed also at a later age is not particularly difficult [3]. The authors of this article think that postponement of sternal reconstruction until adolescence is not justified. The influence of a major defect in the chest wall on the mechanics of breathing should be considered. It is known from observations that congenital sternal cleft causes changes in the route of breathing. It appears that the late treatment of major congenital sternal cleft can have a negative influence on the efficiency of lungs ventilation and may promote atelectasis formation. But a very rare occurrence of this defect and lack of reports which take into consideration mechanics of breathing do not allow for unequivocal conclusions. Additionally, the interesting surgical technique deserves attention. It is based on the use of the cut out cartilaginous elements of parasternal ribs to restore the sternum, after their placement into the space between 2 approximated plates of the cleft sternum. This operation can be treated as a transplantation of own cartilage to obtain additional nuclei of ossification of the “new” sternum.

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