CASE REPORT

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Surgical Treatment of Severe *Pectus Excavatum* in an Adolescent: A Case Report

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ABSTRACT

BACKGROUND: *Pectus excavatum* is a malformation characterized by deformities of the cartilaginous parts of the ribs and sternum of varying severity and accounts for 75%–91% of all chest wall deformities. Surgical treatment of *pectus excavatum* in children remains an urgent issue, despite the considerable number of existing surgical correction techniques. These techniques are not universally applicable, especially for asymmetric and rigid forms, requiring ongoing modification and improvement.

CASE DESCRIPTION: A 17-year-old patient underwent surgical correction of a severe *pectus excavatum* complicated by a history of cardiac surgery, namely, median sternotomy and mitral valve prosthesis placement. The surgical procedure involved the release of major vital anatomical structures, the use of an external fixation device providing high corrective force during surgery, and gradual intraoperative correction under cardiac function monitoring.

DISCUSSION: Correction of severe *pectus excavatum* following prior median sternotomy is associated with a high risk of complications, including fatal events such as asystole or massive hemorrhage. In such cases, a minimally invasive thoracoplasty with an additional subxiphoid approach and sternal elevation using an external fixation device is recommended. Conventional sternal elevation techniques (e.g., bone hooks, sutures, and clamps) have been found to be not advisable for pronounced rigid deformities owing to the risk of sternal injury. Moreover, isolated thoracoscopy using the standard MIRPE technique does not ensure the integrity of intrathoracic structures, and traditional bar-flipping maneuver for immediate and forced correction of *pectus excavatum* is inappropriate.

CONCLUSION: Patients with *pectus excavatum* exceeding the threshold for severe deformity, particularly those with previous cardiac surgery, require a treatment different from standard thoracoplasty. The presented approach, involving the release of major vital anatomical structures and gradual intraoperative correction under cardiac monitoring with an external fixation device, is recommended for managing such complex clinical cases.

Keywords: *pectus excavatum*; minimally invasive thoracoplasty; median sternotomy; congenital heart defect; children; Grand Canyon deformity.

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КЛИНИЧЕСКИЙ СЛУЧАЙ

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Хирургическое лечение воронкообразной деформации грудной клетки крайне тяжелой степени у пациента подросткового возраста (клиническое наблюдение)

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АННОТАЦИЯ

Обоснование. Воронкообразная деформация грудной клетки — это мальформация, проявляющаяся деформациями хрящевых участков ребер и грудины различной степени выраженности и, согласно литературным данным, составляет от 75 до 91% общего числа деформаций грудной клетки. Оперативное лечение детей с воронкообразной деформацией грудной клетки на данный момент — актуальная проблема, требующая решения, несмотря на значимое количество способов хирургической коррекции. Существующие методики не универсальны, особенно в отношении асимметричных, ригидных форм данного порока развития, и поэтому их непрерывно модифицируют и совершенствуют.

Клиническое наблюдение. Пациенту 17 лет проведено хирургическое вмешательство в связи с крайне тяжелой степенью воронкообразной деформации грудной клетки, отягощенной ранее проведенным кардиохирургическим лечением порока сердца с использованием продольной стернотомии и протезом митрального клапана. Оперативное лечение включало релиз магистральных, жизненно важных анатомических образований, использование аппарата внешней фиксации с высоким корригирующим моментом в ходе операции, постепенную интраоперационную коррекцию под мониторингом функции сердца.

Обсуждение. Коррекция воронкообразной деформации грудной клетки тяжелой степени на фоне ранее проведенной продольной стернотомии — операция повышенного риска осложнений, в числе которых фатальные (асистолия, массивное кровотечение). Авторы данного наблюдения считают уместной следующую хирургическую технику для данных клинических случаев: малоинвазивную торакопластику с дополнительным субмечевидным доступом и элевацией грудины с применением аппарата внешней фиксации. Как считают авторы, в данном случае нецелесообразно применять стандартные методы элевации грудины (штопор, шовный материал, зажимы), которые могут привести к повреждению тела грудины при выраженной ригидной деформации, не рекомендовано использовать изолированную торакоскопию стандартной MIRPE, не гарантирующую сохранения целостности органов грудной клетки, неуместен традиционный переворот пластины для одномоментного (форсированного) достижения коррекции воронкообразной деформации грудной клетки.

Заключение. Пациенты с воронкообразной деформацией грудной клетки, значительно превосходящей показатели тяжелой степени, а также с последствиями ранее проведенных кардиохирургических вмешательств нуждаются в объеме лечения, который отличается от стандартной торакопластики при воронкообразной деформации грудной клетки. Рассмотренный вариант лечения, включающий релиз магистральных, жизненно важных анатомических образований, использование аппарата внешней фиксации для постепенной интраоперационной коррекции под мониторингом функции сердца, может быть рекомендован в сложных клинических ситуациях.

Ключевые слова: воронкообразная деформация; торакопластика малоинвазивная; стернотомия продольная; порок сердца; дети; гранд-каньон.

Как цитировать

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BACKGROUND

Pectus excavatum (PE) is a common orthopedic condition marked by a chest wall deformity, which in more severe cases, can lead to reduced respiratory and cardiac function [1]. A notable decrease in the anterior mediastinal volume results in the heart shifting leftward, placing additional stress on the right chambers. Reduced pleural cavity space also impairs breathing and limits physical performance in children. These issues are often most noticeable during emotional stress [2].

In the last 25 years, two main approaches have emerged for correcting PE. The first is conservative treatment for children with mild, flexible chest wall deformities. The second involves surgical intervention, which has evolved to include minimally invasive techniques for addressing moderate to severe PE. This group of procedures is now known as MIRPE (minimally invasive repair of pectus excavatum) [3].

In recent years, the surgical correction of moderate and severe PE with a typical, localized, symmetrical form has become a widely accepted method in clinical practice. However, it is important to note that even when these procedures are performed with appropriate indications and technique, there remains a risk of complications in cases with extremely severe deformities or certain anatomical variations [4, 5].

Several factors increase the surgical risk for this patient group, including the severity and extent of the deformity [6], as well as the significant asymmetry and rigidity of the chest wall curvature [7]. Additionally, a history of prior surgery involving the sternocostal complex notably raises the likelihood of unfavorable outcomes in patients undergoing treatment for chest wall deformities [8].

Existing classifications of PE pathology do not account for deformity grades that exceed severe cases by a considerable margin (e.g., four or five times). However, such extreme cases are encountered by surgeons treating these patients. These cases require special attention and a customized approach during preoperative planning, along with precise surgical techniques, continuous monitoring, and active rehabilitative care during the early postoperative period [9]. Rigid, asymmetrical chest wall deformities complicate treatment due to the need for more complex and invasive reconstruction of the anatomical contour in the affected area.

In cases of previous surgeries involving the sternocostal complex, the most common scenario involve patients who underwent median sternotomy in early childhood for cardiac reconstruction. These patients often have scar tissue in the anterior mediastinum, which increases the risk of complications such as bleeding or cardiac arrhythmias resulting from cardiac injury.

A key factor in severe chest wall deformities is the position of the heart. For example, in cases of significant leftward displacement of the heart (residual space), procedures aimed at restoring the retrosternal space carry not only standard surgical risks but also the risk of the heart shifting back toward the midline, potentially leading to rotation and cardiac arrhythmias. This condition may result from compromised patency of the major vessels due to their displacement and deformation, causing myocardial ischemia or impaired sympathetic innervation [10]. However, in some clinical scenarios, the formation of scar tissue in the retrosternal space after previous cardiac surgery can result in a fixed pericardium that keeps the heart in a central position, while progressive PE leads to a cardiogenic syndrome. In these cases, pericardial release may be necessary but is often not feasible during the thoracoscopic stage of MIRPE due to the increased risk of cardiac injury. Some authors highlight the importance of preparing for significant bleeding and the potential need for cardiac reconstruction by a cardiac surgeon in the event of intraoperative heart injury in this patient group [11].

The combination of these factors, along with underlying conditions (such as genetic disorders or chronic comorbidities), raises the likelihood of an atypical and complex clinical scenario in patients with PE. These cases require careful consideration in developing an individualized surgical plan and its precise execution, including preventive measures to avoid complications.

Given these factors, the clinical case presented here represents a complex, rare, and atypical situation that demands a personalized treatment approach and careful execution of the surgical plan.

CASE DESCRIPTION

In 2024, a 17-year-old male patient, B., sought consultation at the H. Turner National Medical Research Center for Children's Orthopedics and Trauma Surgery, presenting with complaints of chest wall deformity, a noticeable cosmetic defect, shortness of breath with minimal physical activity, dizziness, palpitations, and occasional near-syncope.

Since birth, the patient had been under nephrological care for right-sided hydronephrosis and chronic pyelonephritis, for which he had surgery at 3 months of age. PE and mild spinal deformity were diagnosed during the first year of life, along with benign intracranial hypertension. These conditions were managed conservatively and monitored until the age of 3. At 1 year, the patient was diagnosed with grade II mitral valve prolapse with myxomatous degeneration of the leaflets and stage I chronic heart failure. Follow-up with a cardiologist was advised, including regular ECG and echocardiographic evaluation. At 12 years old, genetic testing revealed a pathogenic heterozygous nucleotide substitution in exon 45

of the *FBN1* gene: NM_000138.4:c.5470T. A diagnosis of Marfan syndrome with autosomal dominant inheritance was confirmed, and the patient was classified as disabled. At 13 years old, due to worsening mitral valve prolapse, which included grade III mitral insufficiency and dilation of the aortic root at the sinus of Valsalva, the patient underwent cardiac surgery via median sternotomy. The surgery involved complex mitral valve repair, including quadrangular resection of the posterior leaflet, suturing of the anterior leaflet cleft, and ring annuloplasty.

Over the next 2 years, during periods of rapid skeletal growth, progression of PE and scoliosis was noted, along with worsening cardiac function, including increased hemodynamic stress on the repaired mitral valve. The patient continued to be monitored and received conservative treatment from various specialists, including a pediatrician, gastroenterologist, cardiologist, orthopedic surgeon, and ophthalmologist, who noted moderate mixed astigmatism consistent with the underlying syndrome.

Despite ongoing conservative treatment, the next 4 years were characterized by significant progression of chest wall and spinal deformities, a decline in overall health, and reduced physical activity. The patient was repeatedly referred to specialized medical centers by cardiac surgeons, where surgical correction of the chest wall deformity was recommended. However, surgery was declined due to the high risk of irreversible and potentially life-threatening intraoperative complications.

In 2024, after a consultation with a specialist at our institution, the patient was referred for elective surgery to correct the chest wall deformity, based on the confirmed signs of decompensated PE.

Physical Examination

Upon admission for inpatient treatment at the H. Turner National Medical Research Center for Children's Orthopedics and Trauma Surgery, the patient underwent a thorough evaluation to assess the feasibility of elective orthopedic surgery.

The patient had a proportionate asthenic body type, with a height of 193.5 cm and an arm span matching his height. His body weight was 49.9 kg. He was fully oriented to time and place and communicative but showed a negative attitude toward his condition and the need for treatment. The initial assessment (using institutional questionnaires developed at H. Turner National Medical Research Center for Children's Orthopedics and Trauma Surgery to evaluate baseline status and postoperative satisfaction after thoracoplasty) revealed significant psychoemotional stress and a cardiorespiratory syndrome. The epidemiological history was unremarkable, with a past history of varicella at age 4. The skin appeared clear, pale, and normally pigmented. The patient was able to walk independently, without the use of assistive devices or limping. His head was centered along the body's axis and in a neutral position. Cervical spine mobility was full and painless. Notable shoulder asymmetry (up to 20 mm) and unequal height of the inferior scapular angles (up to 20 mm) were observed, as well as waist triangle asymmetry. The trunk alignment was decompensated, with a leftward shift of up to 15 mm, accompanied by a pronounced left lumbar curve, torsional component, and a compensatory right thoracic curve. The traction test showed limited spinal deformity mobility.

The chest wall exhibited severe deformity (see supplementary material; DOI: 10.17816/PTORS654520-4252264), with a right-sided PE. The deformity area was rigid, with a sternal depression depth of up to 92 mm (measured in the standing position during inspiration at the deformity's apex). The transverse diameter of the anterior chest wall measured 300 mm. The deformity was extensive, asymmetrical, and eccentric, with the apex located low at the level of the fifth intercostal space. The cranial extent of the PE involved the first rib. A prominent apical cardiac impulse was observed on the left side. The patient reported chest pain on palpation in the cardiac triangle area, although it was not well localized. A postoperative scar, approximately 14 cm long, was present over the sternum, extending from the inferior border of the manubrium to beyond the base of the xiphoid process. The overlying skin was adhered to the underlying tissues; the scar was immobile and showed no signs of inflammation.

Examination of the upper and lower limbs revealed symmetrical absolute and relative lengths on both sides. The range of motion in the upper limb joints showed no functionally significant limitations. No neurological or hemodynamic disturbances were noted in the distal segments of the extremities.

Diagnostic Assessment

Echocardiography: Left ventricular end-diastolic diameter, 43 mm; end-systolic diameter, 28 mm; ejection fraction, 63%; fractional shortening, 34%; interventricular septum thickness in diastole, 7 mm; posterior wall thickness of the left ventricle in diastole, 7 mm; relative left ventricle wall thickness, 0.33%; indexed end-diastolic dimension, 25.29 mm/m^2. Left atrial index, 19.41; right atrial index, 17.65. Mitral valve: Leaflet thickening with regurgitation; peak flow velocity, 1.3 m/s; diastolic function E/A ratio, 1.2. Aorta: Sinus of Valsalva diameter, 35 mm; ascending aorta, 28 mm. Aortic valve: Mild regurgitation; peak flow velocity, 1.0 m/s. Tricuspid valve: Mild regurgitation; tricuspid regurgitation pressure gradient, 25 mmHg. Pulmonary artery: Trunk diameter, 22 mm; valve leaflets normal; peak velocity up to 0.8 m/s. Pericardium: No effusion. Conclusion: Status post-mitral valve repair; mitral insufficiency and fibrosis; aortic root dilation.

Electrocardiography: Marked left-axis deviation; heart rate, 71 bpm; alpha angle, 77°; incomplete right bundle branch block. Emergency ECG monitoring may be necessary.

Pulmonary function testing: Severe restrictive ventilatory impairment.

Abdominal ultrasonography: Ultrasound findings suggest gallbladder shape anomaly and dilation of the right renal pelvicalyceal system.

Stabilometry: Leftward and posterior displacement of the body's center of mass; grade III dysfunction of the statokinetic system with signs of subcompensation.

Chest computed tomography: Chest wall deformity with an asymmetric PE component. Haller index, 15.170; compression index, 0.154; right-sided sternocostal angle, 34°. Evidence of prior sternotomy with cerclage wire at six levels. The distal edge of the sternum is closely positioned to the ventral surface of the spine and major vessels (aorta, inferior vena cava). A mitral valve support ring (post-mitral valve repair) was observed.

Chest radiography: No additional findings compared to the chest CT. Spinal deformity confirmed, showing a primary left-sided lumbar curve peaking at the second lumbar vertebra and measuring up to 78°, and a compensatory right-sided thoracic curve of up to 34°, observed in the standing position (Fig. 1).

Additional specialist evaluations:

- Ophthalmologist: Moderate mixed astigmatism
- Endocrinologist: Protein-energy deficiency, constitutional tall stature
- Cardiologist: Marfan syndrome, grade III mitral valve prolapse with myxomatous degeneration; grade II–III mitral regurgitation; dilation of the aortic root at the sinus of Valsalva; previous multicomponent mitral valve repair with support ring

Following a comprehensive evaluation in the specialized department, the decision was made to proceed with elective surgical correction of the severe PE. The goal of the procedure was to restore the retrosternal space, alleviate right heart overload (including strain on the previously repaired mitral valve), reduce the severity of the cardiorespiratory syndrome linked to the underlying condition, and prevent further progression of the deformity.

Interventions

After standard preoperative preparation, the patient underwent surgery under endotracheal anesthesia combined with continuous regional paravertebral block. The procedure was performed with the patient in a supine position, arms abducted to 90° and elbows flexed to 45°. The surgery included minimally invasive thoracoplasty with an additional subxiphoid approach and osteosynthesis of the sternocostal complex using a fixation system developed at the H. Turner National Medical Research Center for Children's Orthopedics and Trauma Surgery (Patent RU No. 2828751C1; Registration Certificate RU No. РЗН 2024/22790, "Sternocostal fixation system for surgical correction of asymmetrical chest wall deformity," manufactured by Anatomica Innovation Center, Kazan).

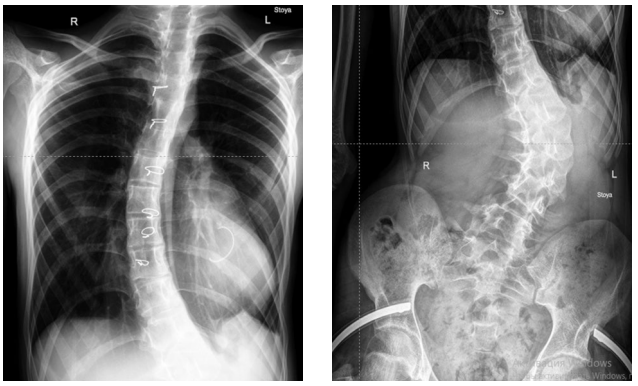

Fig. 1. Full-length standing anteroposterior radiograph of the spine showing left-sided lumbar scoliosis with an extended compensatory thoracic curve; sternal cerclage wire following median sternotomy; mitral valve support ring.

The surgery was performed through three access points: two lateral incisions, each 40 mm long, along the right and left anterior axillary lines at the level of the fifth rib, and one subxiphoid incision up to 65 mm in length. Due to near-complete fusion of the costal arches in this area, a 40-mm segment of the seventh rib from the left costal arch was partially resected through the subxiphoid access. Additionally, a portion of the right fourth costal cartilage was resected through the same access. A retrosternal tunnel was then created at the level of the resected costal cartilages. This space was filled with scar-altered tissues anchoring the pericardium of the right heart chambers, aorta, and inferior vena cava to the dorsal surface of the sternum. An external fixation device was assembled on the operating table, consisting of a support plate mounted on a threaded rod, allowing for gradual sternal elevation using a distraction screw mechanism (Fig. 2). With the external fixation device in place and under direct visualization using standard surgical instruments, the retrosternal space was carefully released to accommodate the support plate. Gradual release of the dorsal surface of the sternum was performed up to the second intercostal space (Fig. 2).

Sternal elevation was carried out gradually over a 40-min period, with the procedure intermittent pauses to address episodes of bradycardia. Additional release of scar-altered tissues in areas of tension within the retrosternal space was performed as necessary (Fig. 3).

Once optimal correction of the sternocostal complex was achieved, an introducer was inserted from left to right through the previously created lateral approaches under direct visual guidance. Two plates were then positioned within the retrosternal space to stabilize the correction. The plates

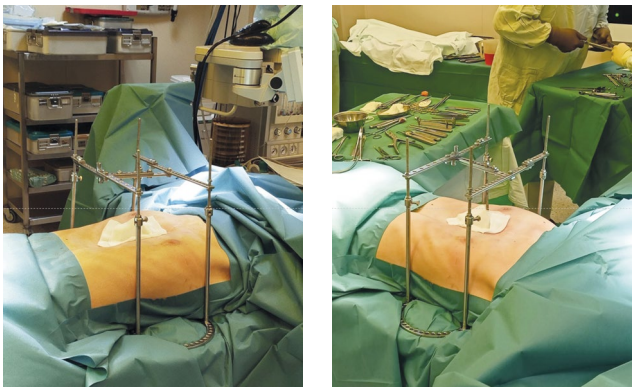

Fig. 2. Stage of retrosternal release and sternal elevation using the external fixation device (EFD) Release of the aorta, pericardium, and inferior vena cava. The EFD was used intraoperatively for 40 min.

were secured using a left-sided stabilizer and two M5 locking screws. The osteosynthesis of the sternocostal complex was stable. The external fixation device used for sternal elevation was removed, and drainage was placed in both pleural cavities and the retrosternal space along the ventral surface (Fig. 4).

Total operative time: 170 minutes. Intraoperative blood loss: 120 mL

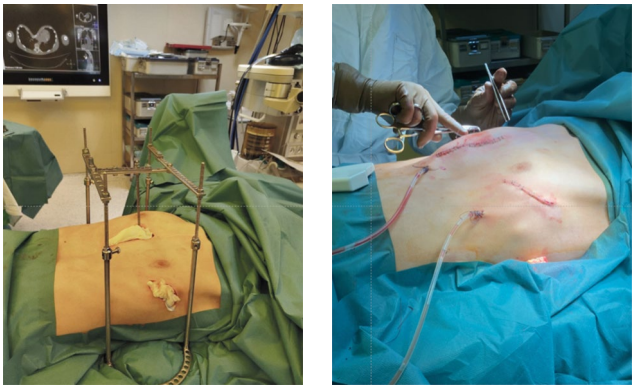

Fig. 3. Transition from elevation of the sternocostal complex using an external fixation device to osteosynthesis with two plates.

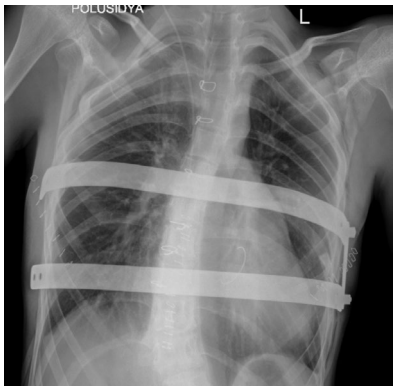

Fig. 4. Follow-up postoperative chest X-ray, anteroposterior view, 2 days after surgery.

After surgery, the patient was transferred to the intensive care unit in a supine position. On postoperative Day 1, mobilization in a seated position was initiated. Multimodal analgesia was used in the early postoperative period, combining the previously administered regional anesthesia with a microjet infusion of fentanyl. Starting from postoperative Day 2, as the patient progressed with mobilization, therapy was supplemented with nonsteroidal anti-inflammatory drugs for 2 days, and opioid analgesics were gradually reduced and discontinued. The patient was able to stand on postoperative Day 3.

Scheduled postoperative imaging included standard anteroposterior chest radiography on postoperative Days 1, 5, and 10 and chest CT on postoperative Day 3. After the chest CT and verticalization, two drains were removed; the right pleural drain was removed on Day 5 after additional radiographic control (Figs. 5 and 6).

Gradual mobilization continued for the patient for up to 10 days postoperatively until basic daily activities were fully regained. The patient was then discharged for outpatient follow-up. Over 6 months after surgery, physical activity (excluding strenuous physical activity and sports) has been fully restored, pain symptoms have resolved, and overall health is reported as good.

DISCUSSION

Correcting severe PE following a prior median sternotomy is a highly complex surgical procedure with risks of complications, including life-threatening events such as asystole and massive hemorrhage. Currently, there is no consensus on the best treatment approach for this patient group. Published data suggest several approaches can be considered for addressing this issue. One option is repeat sternotomy followed by release of the retrosternal space, allowing osteosynthesis of the sternum in the corrected position. This method is relatively safe in terms of dissecting mediastinal structures from scar-altered tissues, but it often requires osteosynthesis of the sternum in a corrected position, which can be challenging, especially when the sternum is already fixed during surgery. This approach also carries the risk of delayed union or nonunion of the sternocostal complex.

The second option is radical thoracoplasty, involving dissection and resection of the costal cartilage segments contributing to the deformity to achieve mobility of the sternum. After releasing the retrosternal space, the sternum is fixed in the corrected position. In this method, the rib cage's stability is restored along the preserved perichondrial tissues that were not resected. This technique is more traumatic and less frequently used than the alternative methods.

Standard minimally invasive thoracoplasty MIRPE was not deemed suitable for this clinical case due to the previously

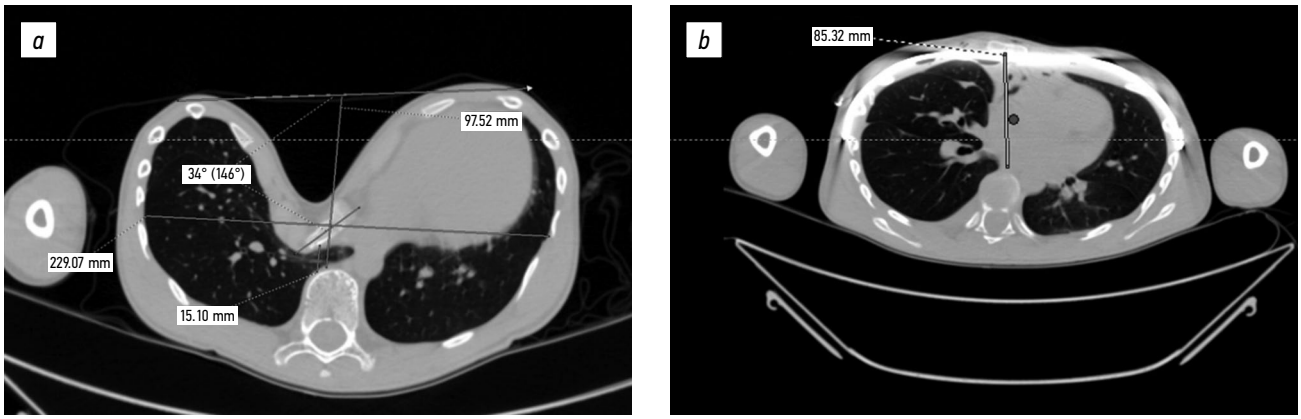

Fig. 5. Chest CT scans before and after surgical correction: *a*, initial retrosternal space size before surgery; *b*, control examination on postoperative Day 5.

mentioned risks of severe and potentially fatal complications. Although rare, these complications cannot be ignored because of their seriousness. A multicenter study of this patient group reported an accidental cardiac injury rate of up to 7% among other complications [12].

The surgical approach described in this article for this patient group involves minimally invasive thoracoplasty with an additional subxiphoid access. This technique minimizes surgical trauma, considering the prior cardiac surgery, while maintaining the stability of the sternocostal complex. It also allows access to scarred tissues at the deformity's apex and provides better control of the retrosternal space during instrument manipulation and implant placement during osteosynthesis. Importantly, this approach addresses two key challenges: ensuring optimal visualization of the retrosternal space (both visually and manually) and correcting the sternum, which can be particularly rigid in cases of extreme, eccentric deformity following sternotomy.

Given the intraoperative challenges in this case, we successfully corrected the retrosternal space without causing significant cardiac arrhythmias. This was achieved through very gradual distraction using an external fixation device, which allowed adequate visual and manual control from two access points: the subxiphoid entry into the retrosternal space and the site of the resected costal cartilage on the right side. We believe that conventional sternal elevation techniques (such as bone hooks, sutures, or clamps) should be avoided in such cases due to the risk of damaging the sternum from its marked rigidity. Similarly, isolated thoracoscopy with the standard MIRPE technique is not recommended due to the risk of internal organ injury. The traditional bar-flipping maneuver for a single, forceful correction of PE is also inappropriate in this situation. It is crucial to note that the controlled, time-based correction of the chest wall deformity created favorable conditions to prevent cardiac rhythm disturbances during the procedure. We consider this correction technique

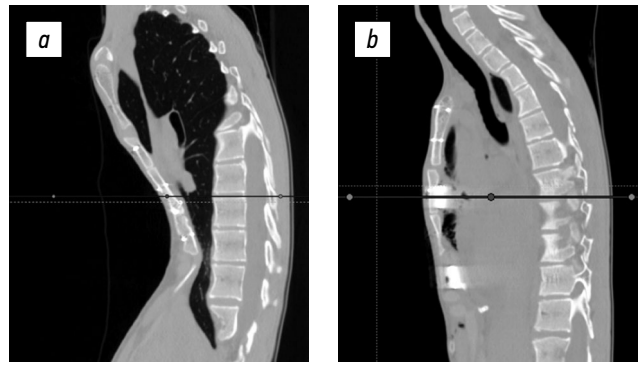

Fig. 6. Chest CT scans before and after surgical correction: *a*, preoperative; *b*, postoperative Day 5.

a feasible option for managing asymmetric forms of PE that significantly exceed the typical threshold for severe deformity, especially in patients with a history of cardiac surgery.

CONCLUSION

Patients with PE that significantly surpasses the threshold for severe deformity, especially those with a history of cardiac surgery, require an alternative treatment strategy to standard thoracoplasty protocols. The approach outlined, which involves releasing major vital anatomical structures, using an external fixation device for high corrective force, and performing gradual intraoperative correction while monitoring cardiac function, may be suitable for managing complex clinical cases.

ADDITIONAL INFORMATION

Author contributions: *D.V. Ryzhikov:* conceptualization, data analysis, writing – original draft; *B.H. Dolgiev:* data collection, writing – original draft; *S.V. Vissarionov:* study design, writing – review & editing; *J.O. Zhukova, I.A. Boroznyak:* data collection, writing – review & editing. All authors approved the version of the manuscript

to be published and agreed to be accountable for all aspects of the work, ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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