To cite this article: Yuvaci AU, Gencel E, Benli O. Staged reconstruction of thoracic ectopia cordis with complete sternal aplasia: a case report. Curr Thorac Surg 2026;11(2):203-207.

Case Report

Staged reconstruction of thoracic ectopia cordis with complete sternal aplasia: a case report

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ABSTRACT

Ectopia cordis is a rare and frequently fatal congenital anomaly characterized by partial or complete displacement of the heart outside the thoracic cavity due to defective midline fusion during embryogenesis. Surgical management remains challenging because of the associated cardiac and chest wall defects. A 37-week gestational age neonate presented with thoracic ectopia cordis, atrial septal defect, pericardial defect, and complete sternal aplasia. On day 29 of life, after hemodynamic stabilization and multidisciplinary operative planning, the patient underwent combined cardiovascular and reconstructive surgery. Following cardiac repositioning into the thoracic cavity and pericardial reconstruction using a bovine pericardial patch, chest wall reconstruction was performed with bipedicled fasciocutaneous flaps harvested from the pectoral region. Initial postoperative recovery was complicated by suture line necrosis, which required surgical debridement and Z-plasty reconstruction. Subsequent patch exposure was successfully managed with patch excision, flap release, medial advancement, repeat Z-plasty reconstruction, and wound dressings. Wound cultures demonstrated no significant bacterial growth. Complete secondary healing was achieved without additional complications. Thoracic ectopia cordis requires individualized multidisciplinary management and careful staged reconstruction. Bipedicled fasciocutaneous flaps provided reliable soft tissue coverage and allowed successful reconstruction following postoperative wound complications. Flexible reconstructive strategies and close postoperative monitoring may improve outcomes in these complex patients.

Keywords: ectopia cordis, chest wall reconstruction, surgical flaps, sternal aplasia

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DOI: 10.26663/cts.2026.029
Received 12.05.2026 Accepted 14.07.2026

Introduction

Ectopia cordis is a rare and often fatal congenital anomaly characterized by the partial or complete displacement of the heart outside the thoracic cavity, posing significant neonatal challenges [1]. This condition results from incomplete fusion of the anterior chest wall during embryogenesis, typically between the sixth and eighth weeks of gestation, leading to aberrant extra-thoracic positioning of the heart [2]. With an incidence of 5.5 to 7.9 per million live births, ectopia cordis remains an uncommon but critical congenital defect.

The severity of this condition varies; it is classified into cervical, cervicothoracic, thoracoabdominal, and abdominal subtypes, each requiring distinct management strategies. Ectopia cordis is frequently associated with additional anomalies, notably Cantrell's pentalogy, which includes diaphragmatic hernia, omphalocele, sternal and pericardial defects, and congenital heart disease. The absence of pericardial protection further increases vulnerability to trauma and infection [2]. When the heart protrudes through the chest wall above the diaphragm, it is classified as extrathoracic ectopia cordis, one of the most severe forms of the anomaly. Prognosis is generally poor, particularly in thoracic ectopia cordis, emphasizing the need for immediate intervention [3].

Surgical reconstruction primarily involves repositioning the heart and covering the chest wall defect; however, because of the high mortality associated with this condition, successful surgical correction remains uncommon [4]. Single-stage thoracoabdominal repairs have demonstrated feasibility, reducing the need for multiple interventions. Given the complexity of ectopia cordis, surgical approaches must be tailored to the severity of the presentation and associated anomalies to optimize survival [3].

Although successful repairs have been reported, long-term outcomes and quality of life remain underexplored due to the rarity of this condition [3]. Large-scale studies and clinical trials are limited, posing challenges in developing standardized management guidelines. Future research should focus on refining surgical techniques and evaluating individualized approaches based on severity and associated anomalies [3].

Case Report

We present the case of a 37-week gestational age neonate diagnosed with ectopia cordis, characterized by the heart apex positioned outside the thoracic cavity (Figure 1). At birth, the infant exhibited bradycardia and respiratory distress, requiring positive pressure ventilation, endotracheal intubation, and admission to the neonatal intensive care unit (NICU) with synchronized intermittent mandatory ventilation. Intravenous ampicillin and gentamicin therapy was initiated, and the exposed cardiac tissue was covered with saline-soaked dressings. No other congenital anomalies were immediately noted.

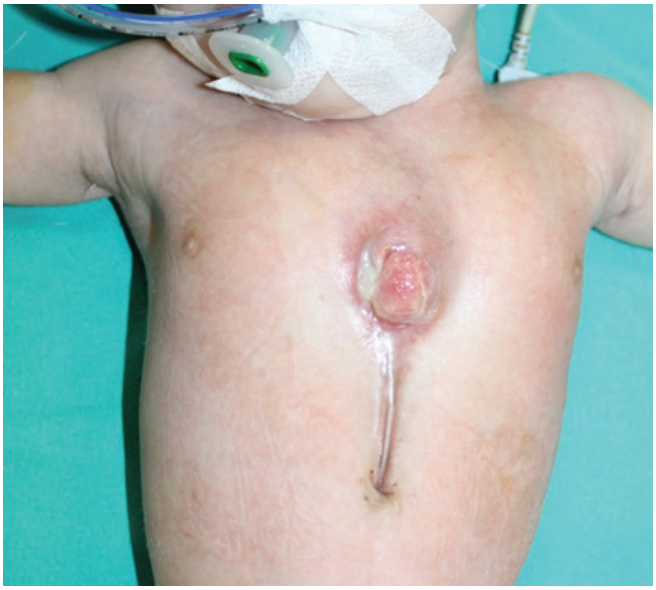

Figure 1. Preoperative clinical presentation of the neonate with thoracic ectopia cordis. The apex of the heart is exposed through a midline anterior chest wall defect, without sternal coverage.

Echocardiography revealed an atrial septal defect with a left-to-right shunt, a 2 mm patent ductus arteriosus, and an open ductus venosus. Additional findings included aplasia cutis, retrognathia, and a high-arched palate. Thoracic computed tomography (CT) demonstrated complete sternal aplasia with intact bilateral clavicles and ribs, together with a 2 cm anterior thoracic wall defect through which the cardiac apex herniated. Dextrocardia was also noted.

On day 29 of life, after hemodynamic stabilization, completion of diagnostic evaluations, and multidisciplinary operative planning, the patient underwent a combined cardiovascular and plastic surgery procedure. During the preoperative period, the exposed cardiac tissue was managed with saline-soaked dressings to maintain tissue hydration. Antibiotic ointments and advanced wound coverings were intentionally avoided because of concerns regarding possible local reactions and irritation over the exposed cardiac structures. For infection prophylaxis, intravenous ampicillin and gentamicin therapy was initiated and continued throughout the surgical procedures

and postoperative period. The cardiovascular surgery team repaired the pericardial defect with a bovine pericardial patch (4 x 6 cm, 0.1-0.5 mm in thickness) and repositioned the heart apex within the thoracic cavity. Cardiopulmonary bypass was not required during the procedure. Simultaneously, chest wall reconstruction was performed using 15 × 15 cm bipedicled fasciocutaneous flaps harvested from the pectoral region (Figure 2). Flap donor sites were closed primarily, and two surgical drains were placed (Figure 3).

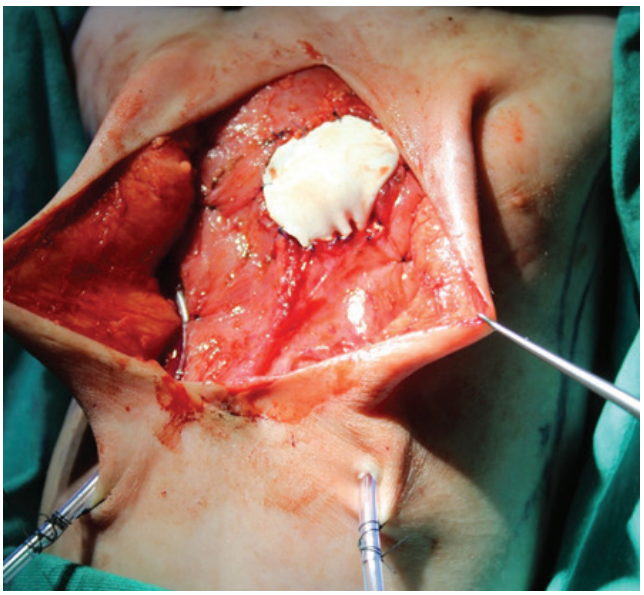

Figure 2. The heart was repositioned within the thoracic cavity, and the pericardial defect was reconstructed using a bovine pericardial patch. Bipedicled fasciocutaneous flaps, harvested from the pectoral region, were advanced to cover the anterior thoracic defect.

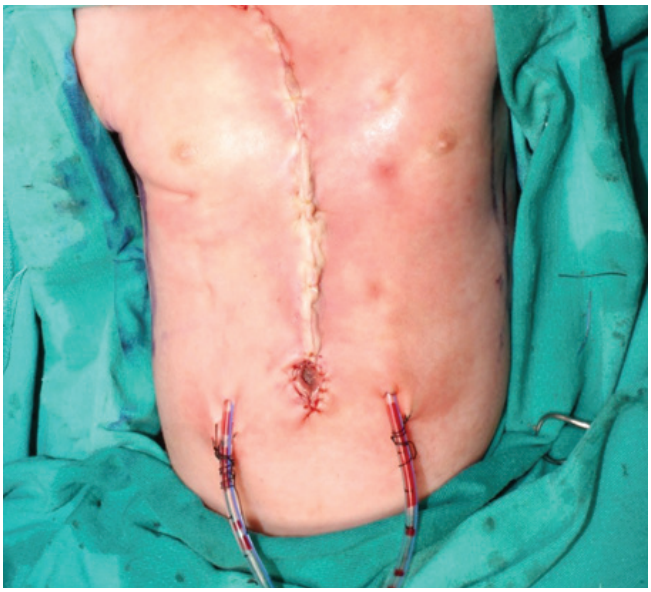

Figure 3. Immediate postoperative appearance showing complete soft-tissue coverage of the anterior chest wall and bilateral closed-suction drain placement.

On day 35 of life, necrosis along the inferior suture line was noted, requiring a second surgical intervention. The necrotic tissue was debrided, and Z-plasty was performed for wound closure. On day 49 of life, erythema developed at the flap junction, and further evaluation revealed patch exposure localized to the flap suture line (Figure 4). Wound culture demonstrated no significant bacterial growth. The bovine pericardial patch was excised, and the inflamed area was trimmed. To improve wound closure, previously elevated bipedicled fasciocutaneous flaps were released via lateral thoracic incisions, advanced medially, and approximated at the midline using new Z-plasty flaps. Donor sites were treated with epidermal growth factor-based wound dressings (Figure 5).

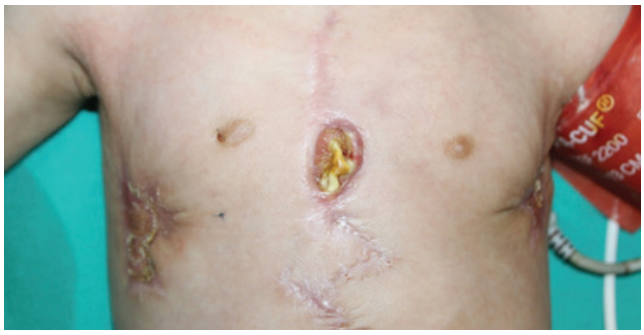

Figure 4. On day 49 of life, erythema appeared at the flap junction, and further evaluation revealed exposure of the bovine pericardial patch.

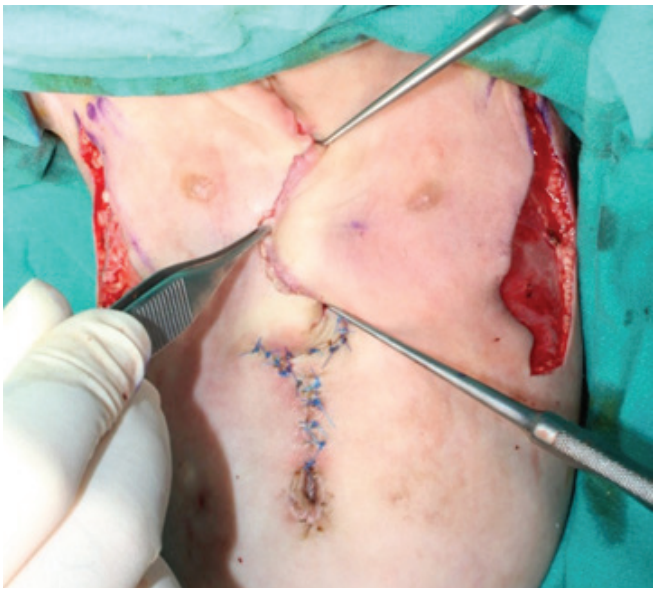

Figure 5. Final reconstructive procedure. The exposed bovine pericardial patch was excised, and the inflamed tissue was debrided. Previously elevated bipedicled fasciocutaneous flaps were released from the lateral thoracic region, advanced toward the midline, and reapproximated using new Z-plasty flaps to reinforce closure. Donor sites were treated with epidermal growth factor-based wound dressings.

By day 64 of life, the dressings were removed, demonstrating complete secondary wound healing without additional complications (Figure 6). The patient remained hospitalized for an additional 10 days and was discharged in stable condition. Postoperative follow-up continued for 2 months, during which no recurrent wound complications, signs of infection, hemodynamic instability, or additional cardiac complications were observed. After the second postoperative month, the patient did not return for further follow-up at our institution. To the best of our knowledge, no mortality was reported during the available follow-up period.

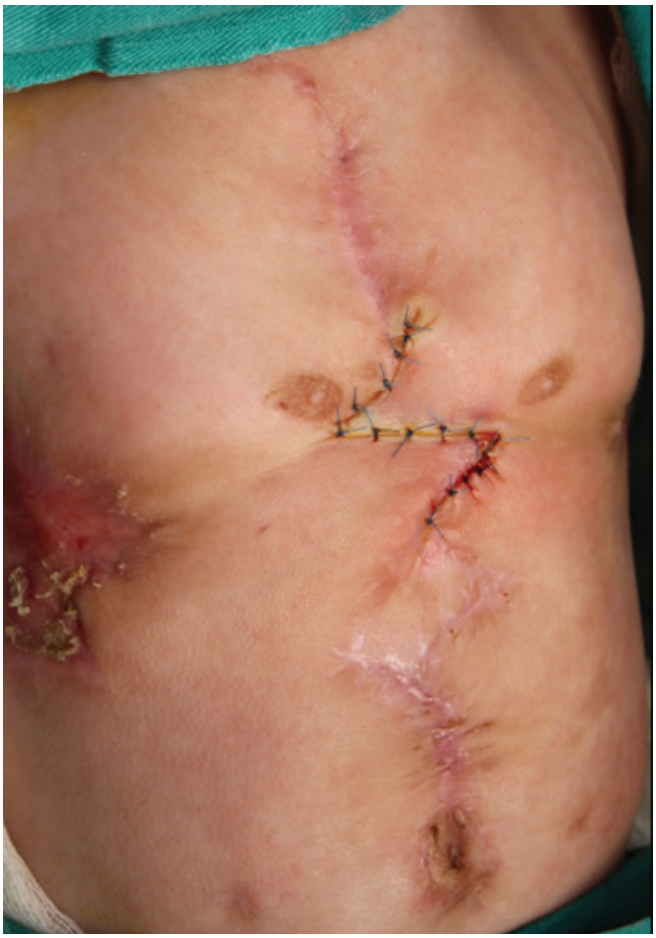

Figure 6. On day 64 of life, the dressings at the flap donor sites were removed, showing complete secondary healing with no further complications.

Discussion

Managing ectopia cordis requires a multidisciplinary approach involving neonatology, cardiothoracic surgery, and reconstructive surgery to optimize outcomes. Both our case and the existing literature demonstrate that successful cardiac and chest wall repair may be achieved in infancy, particularly in thoracic and thoracoabdominal forms of ectopia cordis [3]. Initial management focuses on protecting the exposed heart from trauma, dehydration, and infection while addressing associated anomalies such as omphalocele [5]. When immediate surgical correction is not feasible, sterile dressings and protective barriers remain essential for temporary management.

Surgical decision-making depends on the type and severity of the defect, associated anomalies, and overall patient stability. Surgical correction remains the definitive treatment [2]. Although successful single-stage repairs have been reported, staged reconstruction may be necessary to allow adequate tissue adaptation and to prevent hemodynamic compromise [1,3,5]. Premature closure of the thoracic cavity may result in compression of the heart and great vessels, necessitating alternative reconstructive strategies such as subcutaneous pockets, tissue expansion, or staged soft tissue advancement [3]. In patients with complete or near-complete sternal aplasia, achieving durable chest wall coverage remains particularly challenging because of limited local tissue availability and increased wound tension. In our patient, postoperative wound complications were likely related to increased tension at the midline closure rather than prosthetic infection, as wound cultures demonstrated no significant bacterial growth.

In the present case, bipedicled fasciocutaneous flaps harvested from the pectoral region were preferred because they provided reliable vascularity, adequate soft tissue coverage, and preservation of thoracic wall compliance without muscle sacrifice. However, postoperative wound complications developed, including suture line necrosis followed by patch exposure. Management of prosthetic exposure in neonatal chest wall reconstruction remains challenging because wound closure and preservation of chest wall stability must be achieved simultaneously. Therefore, a staged reconstructive approach was preferred in our patient. Debridement, excision of the patch, flap release, medial advancement, and repeat Z-plasty reconstruction allowed preservation of chest wall integrity while achieving successful wound closure.

Recent surgical advances have improved the management of ectopia cordis. Extracorporeal membrane oxygenation (ECMO) has been used in infants developing cardiac failure following repair [3]. Additionally, three-dimensional printing technologies may facilitate preoperative planning and improve chest wall reconstruction accuracy. Alloplastic materials, including synthetic meshes and bioabsorbable plates, may provide structural support and improve long-term

reconstructive outcomes. Mohan et al. emphasized the importance of stable soft tissue coverage in pediatric chest wall reconstruction, particularly in patients requiring prosthetic support [4]. In our case, a soft tissue–based reconstructive approach was preferred because it provided adequate chest wall coverage while preserving thoracic compliance during the neonatal period.

Future research should focus on refining surgical techniques, exploring novel biomaterials, and developing individualized reconstructive strategies for these complex patients. Advanced imaging modalities, including fetal MRI, may improve prenatal diagnosis and facilitate earlier multidisciplinary planning. Long-term follow-up studies remain necessary to better evaluate cardiac function, respiratory mechanics, chest wall development, and quality of life in surviving patients [3]. In selected patients with significant intracardiac anomalies, additional palliative procedures such as Blalock-Taussig shunting or pulmonary artery banding may also be required [6].

Ectopia cordis remains one of the most challenging congenital thoracic anomalies and requires individualized surgical planning. The successful staged reconstruction of our patient with complete sternal aplasia highlights the importance of flexible reconstructive strategies and multidisciplinary management. Continued advances in neonatal care, biomaterials, and reconstructive techniques will be essential for improving survival and long-term outcomes in these patients.

This report has several limitations. The patient was lost to follow-up after two months, precluding assessment of long-term chest wall growth, respiratory mechanics, prosthetic durability, and developmental outcomes. Furthermore, because this is a single case report, conclusions regarding the superiority of this reconstructive strategy cannot be generalized.

In conclusion, ectopia cordis remains a rare and challenging congenital anomaly associated with high morbidity and mortality. Successful management requires individualized multidisciplinary planning involving neonatal intensive care, cardiovascular surgery, and reconstructive surgery teams. In our case, bipedicled fasciocutaneous flaps provided effective chest wall coverage following cardiac repositioning and allowed successful reconstruction after postoperative wound complications. Careful postoperative monitoring and staged reconstructive management are critical for preserving thoracic stability and optimizing clinical outcomes in these complex patients.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support.

Authors' contribution

AUY,EG: Surgical procedure, conceptualization, methodology, writing - original draft. OB: Cardiovascular surgery procedure, data curation, writing - review and editing. All authors read, critically revised, and approved the final version of the manuscript.

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