International Journal of Surgery Case Reports 131 (2025) 111353

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Complex management of ectopia cordis complicated by pentalogy of cantrell: Report of two cases and review of current evidence

Abarham Martadiansyah [a,*], Nuswil Bernolian [a], Putri Mirani [a], Peby Maulina Lestari [a], Aswin Nugraha [b], Bella Stevanny [c]

[a] Division of Fetomaternal Medicine, Department of Obstetrics and Gynecology, Faculty of Medicine, Sriwijaya University, Moh. Hoesin General Hospital, Palembang, South Sumatra, Indonesia
[b] Division of Thoracic, Cardiac, and Vascular Surgery, Department of Surgery, Faculty of Medicine, Sriwijaya University, Moh. Hoesin General Hospital, Palembang, South Sumatra, Indonesia
[c] Department of Obstetrics and Gynecology, Faculty of Medicine, Sriwijaya University, Moh. Hoesin General Hospital, Palembang, South Sumatra, Indonesia

ARTICLE INFO

Keywords:
Bogota bag
Bovine pericardium
Cantrell's pentalogy
Congenital anomaly
Ectopic cordis

ABSTRACT

Introduction and importance: Ectopia cordis is a rare congenital anomaly characterized by the abnormal positioning of the heart outside the thoracic cavity, frequently associated with additional congenital defects such as those seen in Pentalogy of Cantrell. Due to the rarity and severity of these anomalies, timely diagnosis and appropriate management remain clinically challenging but critical for patient outcomes.
Case presentation: We report two distinct cases of ectopia cordis managed at our institution. The first case presented with complete ectopia cordis and absence of the sternum, requiring initial management with a Bogota bag followed by placement of bovine pericardium; unfortunately, the patient succumbed to complications of sepsis and heart failure at 12 days of life. The second case involved partial ectopia cordis accompanied by features of Pentalogy of Cantrell, managed conservatively with a Bogota bag and spontaneous epithelialization, resulting in successful discharge with stable clinical status.
Clinical discussion: Management of ectopia cordis, especially when complicated by associated anomalies, presents significant surgical and clinical challenges. Survival rates remain low due to complications such as sepsis, cardiac instability, and limited thoracic cavity space. Utilization of biomaterials like bovine pericardium has shown promise in providing necessary cardiac protection when primary closure is not feasible. Early multidisciplinary intervention, involving neonatologists, pediatric cardiologists, and surgeons, is crucial for optimizing outcomes.
Conclusion: While prognosis for ectopia cordis remains poor, careful and individualized management strategies, including timely intervention and infection prevention, can lead to favorable outcomes as demonstrated in our second case. This underscores the importance of a multidisciplinary approach and early prenatal diagnosis in enhancing survival rates for this rare condition.

1. Introduction

Ectopic cordis (EC) is an rare congenital abnormality of the heart characterized by a malformation in the chest and abdominal wall, causing the heart to be located outside the thoracic cavity. This syndrome also encompasses irregularities in the pericardium, diaphragm, sternum, and frequently incorporates cardiac anomalies. Ectopic cordis is characterized by the abnormal displacement of the heart from its usual position within the thoracic cavity. The Pentalogy of Cantrell is an uncommon congenital syndrome marked by anomalies in the lower sternum, anterior diaphragm, abdominal wall, ectopia cordis, and congenital heart disease. The etiology of this condition, first described by Cantrell in 1958, remains unknown. However, it is frequently linked to aneuploidy and the failure of lateral mesodermal folds during the early stages of embryonic development. The incidence of this condition is believed to be between 1 in 65,000 and 1 in 200,000 births, with fewer than 100 cases reported worldwide [1,2].

Despite advancements in medical interventions over the past 60 years, mortality rates associated with EC and POC remain high. Definitive reconstruction of the cardiac and thoracic cage poses significant

* Corresponding author at: Dokter Muhammad Ali, Sekip Jaya, Palembang, South Sumatra 30114, Indonesia.
E-mail address: abarhammartadiansyah@fk.unsri.ac.id (A. Martadiansyah).

https://doi.org/10.1016/j.ijscr.2025.111353
Received 5 February 2025; Received in revised form 25 March 2025; Accepted 21 April 2025
Available online 23 April 2025
2210-2612/© 2025 Published by Elsevier Ltd on behalf of IJS Publishing Group Limited. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

challenges, particularly in cases where the ectopia heart cannot be fully enclosed within a hypoplastic thoracic cavity, as seen in thoracic-type anomalies [3,4]. Some researchers have suggested that any necessary corrective cardiac surgeries should be performed before resorting to palliative closure. The complexity of achieving definitive thoracic reconstruction surgery often leads to the selection of palliative closure as the preferred treatment option in cases of ectopia cordis. The primary goal of palliative surgery in patients with ectopia cordis is to shield the heart from environmental exposure. The pioneering staged repair of ectopia cordis was accomplished by Koop in 1975. Early cases have demonstrated the long-term survival of individuals with thoracoabdominal ectopia cordis despite the abnormal positioning and heart protection by intact skin coverage. Utilizing the patient's skin for primary closure is the preferred approach, with biomaterials being utilized when skin coverage is insufficient [5].

Bovine pericardium has emerged as a favoured biomaterial over synthetic options like surgical mesh for use as patches in surgery due to its compatibility, reduced complications, reliable suture retention, minimal suture line bleeding, optimal handling characteristics, and lower risk of inflammation associated with foreign body implantation. Processed bovine pericardium patches are rendered acellular to prevent host reactions to bovine proteins and subsequent inflammatory responses [6].

We provide two case reports of ectopia cordis. The first case involves a full ectopia cordis, which was treated with a Bogota bag and Bovine pericardium. Unfortunately, the patient passed away at the age of 12 days. The second case involved partial ectopia cordis, which underwent spontaneous epithelialization. The newborn was treated with a Bogota bag and subsequently discharged in a healthy state. These cases report aim to capture the attention of neonatologists and other stakeholders involved in the treatment of such patients by emphasizing that, despite their rarity, the prognosis after surgery is unfavorable. Documenting these cases is crucial for enhancing clinical understanding and developing effective management strategies. The patient's parents granted permission for the publication of this case in a scientific journal. This case is documented in compliance with the SCARE criteria [7].

2. Methods

This study is a retrospective case series involving non-consecutive cases, conducted at a our institution. We retrospectively reviewed two cases of ectopia cordis: one with complete heart displacement and another with partial displacement, accompanied by anomalies in the abdominal wall, anterior diaphragm, and lower sternum, consistent with Pentalogy of Cantrell. Written informed consent was obtained from the patients' guardian for publication of this case report and accompanying images and videos.

2.1. Case number 1

A 17-year-old primiparous woman at 35 weeks of gestation was referred from a remote hospital due to suspected fetal ectopia cordis. Ultrasound conducted by a maternal-fetal consultant confirmed ectopia cordis with a small thoracic cavity. The patient underwent a lower segment cesarean section, and a female neonate was delivered.

2.2. Case number 2

A 27-year-old primiparous woman at 35 weeks of gestation, with a suspected case of partial ectopia cordis, was referred from a remote hospital. An ultrasound examination, conducted by a maternal-fetal consultant, revealed multiple congenital anomalies, raising suspicion of partial pentalogy of Cantrell. The patient underwent an elective cesarean section at 38 weeks of gestation.

Written informed consent was obtained from the patient's parents for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

3. Results

3.1. Case number 1

The cesarean section resulted in the birth of a live female neonate with Apgar scores of 8 and 9 at the 1st and 5th minutes, respectively, and a birth weight of 1955 g. The neonate exhibited vigorous crying, a normal heart rate, and respiratory rate with minimal retractions. Physical examination revealed an ectopia cordis situated completely outside the chest, without the presence of pericardium (Fig. 1).

The supplementary file (Video 1) demonstrates the extrathoracic motion of the heartbeat. Following delivery, the newborn's heart was enveloped in a sterile dressing soaked in warm saline solution, which was then covered with sterilized plastic that is not meant to be permanent. The infant was administered vitamin K and chloramphenicol eye ointment. Subsequently, the newborn was promptly transferred to the specialized neonatal critical care unit. The thoracoabdominal x-ray revealed that the heart is positioned outside the thoracic cavity. The broncho-vascular patterns appeared normal, with no indications of intestinal obstruction or hepatomegaly (Fig. 2A). The echocardiogram revealed a balanced condition in all four chambers of the heart, along with a patent ductus arteriosus and an atrial septal defect of 7-8 mm (Fig. 2B and C).

At 2 days old, the infant underwent a surgery to implant a modified Bogota bag. A sterile urine bag was used and sewed continually using a monofilament 3.0 non-absorbable cutting thread between the skin around the ectopia cordis (Fig. 3). After 6 days of the Bogota bag insertion, a surgical procedure was carried out to protect the exposed heart by using bovine pericardium, a routinely used material for reconstructive cardiac surgery. During the procedure, it was observed that the heart was completely located outside the thoracic cavity, and there was a lack of a sternum. Next, the heart was rinsed using a saline solution and then the bovine pericardium was stitched onto the freshened skin edge using a continuous suture made of 5-0 polypropylene. There were no notable changes in the blood flow or pressure during or after the treatment. The surgical specifics can be found in Fig. 4.

The patient was transferred to the NICU after surgery with stable hemodynamic. She was lightly sedated during the first post-operative day. Unfortunately, increased of WBC count was documented during the first and second preoperative day. It was also followed by the increasing of the CRP and procalcitonin level. Antibiotics were

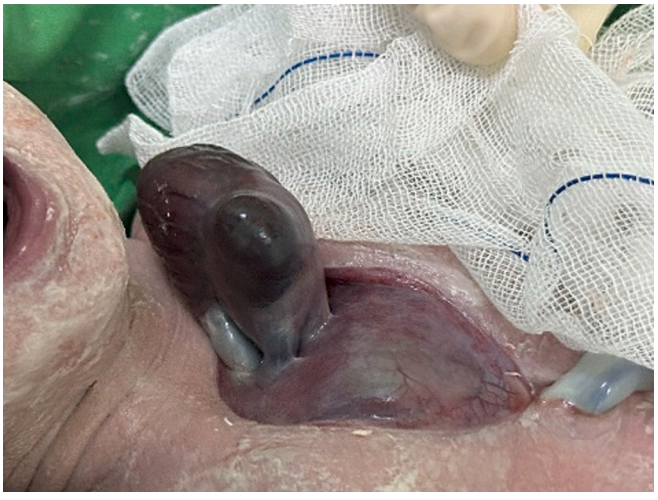

Fig. 1. External heart located entirely outside the chest cavity without pericardium.

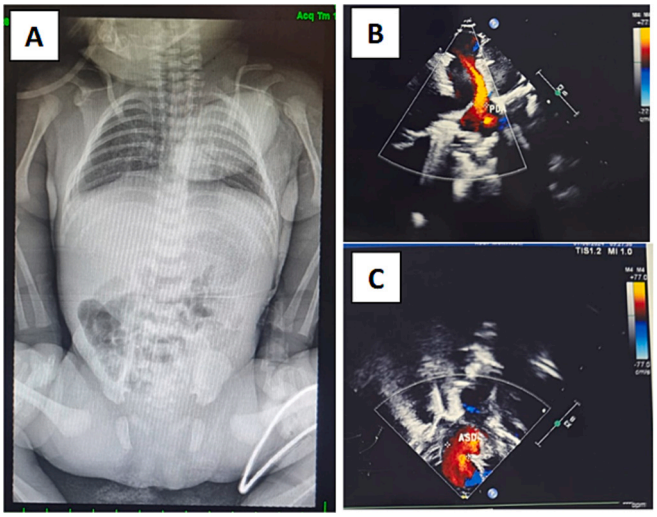

Fig. 2. (A) Thoracoabdominal x-ray resulted heart is located outside the thoracic cavity, (B) echocardiography resulted balance four chambers with result patent ductus arteriosus, (C) atrial septal defect measuring 7-8 mm.

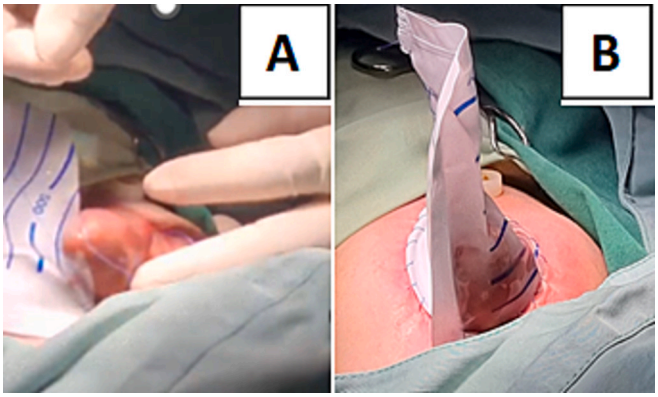

Fig. 3. (A) The process of Bogota bag insertion (B) final results of Bogota bag insertion.

continued according to the infection control team. On the fifth postoperative day, her blood pressure decreased gradually. Inotropic supports were administered and increased accordingly, but the patient eventually died on postoperative day five due to sepsis and heart failure.

3.2. Case number 2

The cesarean section resulted in the birth of a live female neonate with Apgar scores of 7 and 8 at the 1st and 5th minutes, respectively, and a birth weight of 2800 g. The neonate exhibited vigorous crying, a normal heart rate, and respiratory rate but showed signs of cyanosis due to low oxygen saturation. Physical examination revealed partial ectopia cordis located outside the thoracic cavity without the presence of a pericardium. Additionally, a portion of the intestine appeared to be enclosed within a 2 cm membrane extending from the paraumbilical area (Fig. 5).

The dynamic movement of the heartbeat outside the thoracic cavity is shown in the supplementary file (Video 2). An X-ray of the thoracoabdominal region revealed a radiopaque mass in the midline between the thorax and abdomen, raising suspicion of an obstruction, while broncho-vascular patterns appeared normal (Fig. 6A). Echocardiography findings revealed balanced four cardiac chambers, a large atrial septal defect measuring 13.1 mm, a large ventricular septal defect measuring 10.6 mm, a large patent ductus arteriosus measuring 3.8 mm,

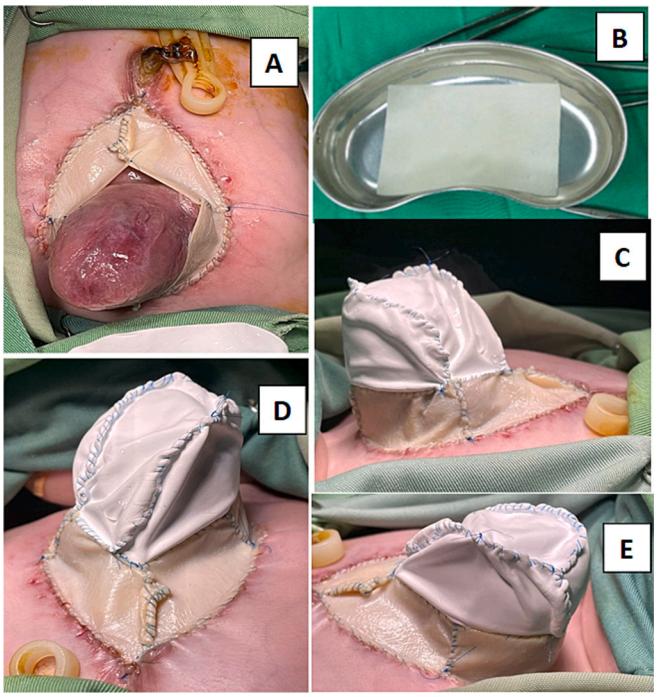

Fig. 4. (A) The heart is completely outside the thoracic cavity without pericardium covering, (B) Bovine pericardium, (C) The heart is covered with the new pericardium (right view), (D) new pericardium (bottom view), (E) new pericardium (left view).

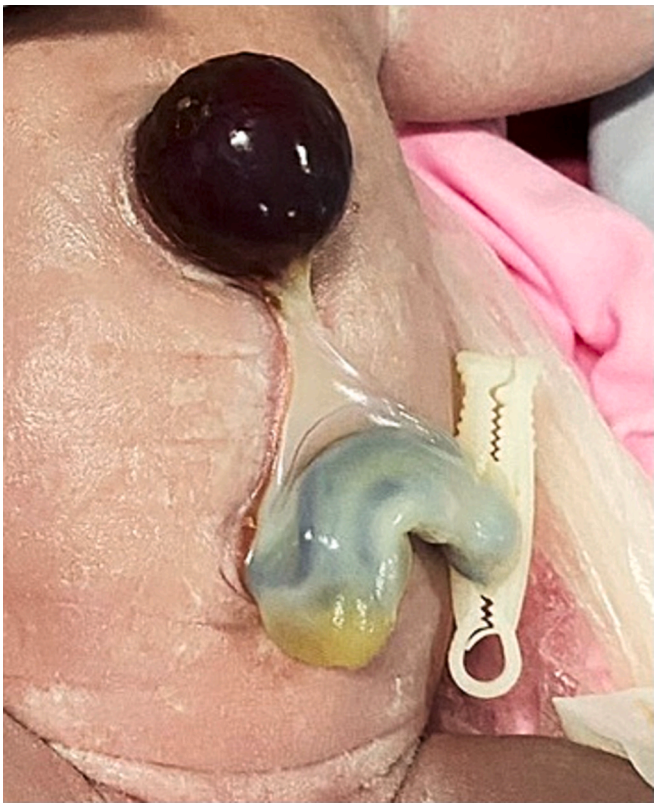

Fig. 5. Partial ectopia cordis without pericardium and part of intestine is wrapped in a 2 cm long membrane.

right ventricular hypertrophy, mild pulmonary stenosis, an overriding aorta, and no pericardial effusion, leading to a diagnosis of Tetralogy of Fallot with a large PDA and large ASD (Fig. 6B and C).

The newborn received a combination of ampicillin and gentamicin to prevent infection, and was scheduled for surgery to protect the exposed heart using bovine pericardium and removal of the omphalocele. Nevertheless, the Bovine pericardium surgery may only be performed on newborns that are precisely 16 days old, as a result of the scarcity of tools and supplies. Curiously, just before the procedure was performed, self-epithelialization was observed in the pericardium cordis (Fig. 7A), so the only action that was carried out was the installation of a Bogota bag and resection of the omphalocele (Fig. 7B). The patient was moved to the Neonatal Intensive Care Unit (NICU) following the surgical procedure, exhibiting stable hemodynamic conditions. A reduction in the white blood cell (WBC) count was observed both the day before surgery and the day of surgery. The administration of antibiotics was continued in accordance with the directives of the infection control team. After several days of monitoring, the newborn was discharged in stable condition and had a Bogota bag installed.

4. Discussion

Ectopic cordis is a rare congenital anomaly in which the heart is partially or entirely located outside the thoracic cavity. This condition may occur as an isolated defect or in conjunction with other abnormalities involving the thoracic or abdominal walls. It is characterized by malformations of the anterior chest wall, resulting in the heart being positioned externally to the thoracic cavity. Ectopic cordis may be linked to chromosomal abnormalities such as Turner syndrome (XO), trisomy 21 and Edwards syndrome (trisomy 18). Intracardiac defects may coexist with ectopia cordis, including atrial septal defect (ASD), ventricular septal defect (VSD), Tetralogy of Fallot (TOF), tricuspid atresia (TA), and double outlet right ventricle (DORV) [7–9].

Ectopic cordis can be classified into five types based on the location of the heart: cervical (5 %), cervicothoracic and thoracic (65 %), thoracoabdominal (20 %), and abdominal (10 %). The condition occurs in approximately 8 to 9 cases per million births. As of 2001, a total of 267 cases had been reported, with 102 (39.2 %) identified as thoracic and 99 (38 %) as thoracoabdominal. The prognosis is generally poor, with 90 % of affected newborns not surviving beyond the first year of life. Prenatal diagnosis of ectopia cordis can be achieved through ultrasound (USG), which allows visualization of the heart outside the thoracic cavity. Early prenatal detection is crucial for identifying ectopia cordis and associated anomalies, typically during the second trimester [10,11].

The Pentalogy of Cantrell (POC) is a constellation of five congenital anomalies that manifest along the midline of the body. These anomalies involve the heart, pericardium, diaphragm, sternum, and abdominal wall. POC is categorized into two types: complete and partial. A complete POC is defined by the presence of all five abnormalities, whereas a partial POC includes fewer than five defects. This syndrome is often referred to as thoracoabdominal ectopia cordis, where the heart is encased in a membrane resembling an omphalocele. Ectopia cordis (EC) frequently occurs in fetuses with POC. Newborns with POC often present with various cardiac anomalies, with ventricular septal defects and Tetralogy of Fallot being the most common. POC can also be associated with chromosomal disorders such as trisomies 13, 18, 21, and Turner syndrome. Initial management focuses on assessing the condition of the skin in areas where the heart and abdominal cavities are exposed [12,13].

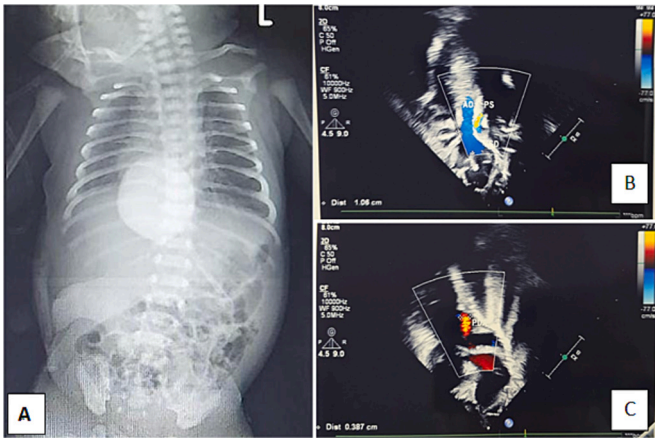

Fig. 6. (A) Thoracoabdominal x-ray resulted radiopaque mass in the midline. (B and C) Echocardiography conclude Tetralogy of Fallot, Large PDA, Large ASD.

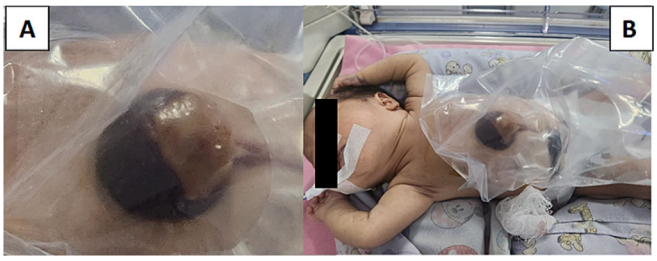

Fig. 7. (A) Self-epithelialization was found in the pericardium cordis. (B) Installation of a Bogota bag and resection of the omphalocele.

Surgical correction of EC poses significant challenges due to associated structural abnormalities and hemodynamic instability. Early surgical interventions aim to reposition the heart within the thoracic cavity and reconstruct associated defects. However, the mortality rate remains high, with many patients not surviving the immediate postoperative period [14]. Araujo Júnior et al. (2023) conducted an international multicenter study across Brazil, Germany, Italy, and Poland, analyzing 31 cases of ectopia cordis (EC) to compare surgical and conservative management outcomes. Of the seven neonates born alive, three underwent surgery and all survived, with follow-up ages of 5 months, 13 years, and 29 years. In contrast, conservative management mostly results in termination of pregnancy (48 %) and spontaneous fetal demise (22.5 %) accounted for most cases, while all four liveborn neonates who did not receive surgery died shortly after birth. The study concluded that surgical correction is the only viable approach for survival, whereas conservative management results in 100 % mortality [15].

Zhang et al. (2014) conducted a study in China analyzing eight cases of ectopia cordis (EC) with pentalogy of Cantrell, comparing staged vs. early one-stage repair. One-stage repair was performed in seven patients (87.5 %), achieving complete anatomical correction but carrying risks of hemodynamic instability due to increased intrathoracic pressure. One patient (12.5 %) required staged repair due to insufficient thoracic space, which reduced the risk of low cardiac output syndrome but prolonged hospitalization and increased infection risk. The study concluded that one-stage repair is preferable for stable neonates with adequate thoracic space, while staged repair is necessary for severe defects where immediate closure is not feasible [16].

Conservative management, focusing on supportive and palliative care, is often considered when surgical options are deemed unfeasible. Berry et al. (2016) explored a novel palliative care approach for neonates with complete ectopia cordis (EC), emphasizing the challenges and benefits of conservative management. The major advantage of this approach is that it allows nonviable cases to avoid the risks of aggressive surgical intervention, offering a dignified, comfort-focused end-of-life care in a community setting rather than a prolonged NICU stay. Their case report describes a neonate with severe intracardiac defects, deemed inoperable, who was discharged home on day 8 and survived 15 days with palliative care, enabling parental bonding. However, the primary limitation of conservative management is its inevitably poor prognosis,

as EC without surgical correction leads to near-universal neonatal mortality due to cardiac failure, infection, or vascular accidents. Despite meticulous wound care to prevent desiccation and infection, the exteriorized heart remains highly vulnerable. Additionally, Berry et al. highlight the psychosocial burden on families, requiring extensive counseling and support. Ultimately, while palliative management offers a humane alternative for nonviable EC cases, it does not improve survival and is best suited for infants where surgical intervention is not feasible [17]. Each case of EC requires individualized assessment to determine the most appropriate management strategy based on the patient's condition and associated anomalies [17].

In our case, the patient presents with abnormal excellent vessel angulation alongside a hypoplastic thoracic cavity, precluding the feasibility of definitive surgical management. Attempts to reposition the heart would lead to constriction and distortion of the great vessels, resulting in compromised cardiac function and rapid hemodynamic deterioration. Consequently, palliative surgery was performed to minimize contamination by limiting the heart's direct exposure to the external environment. Bovine pericardium was used over synthetic mesh due to the former's superior compatibility and durability, especially in the absence of adequate midline skin for direct closure. The procedure was executed successfully without significant hemodynamic disruptions.

Management for the first case, a multidisciplinary approach plays a vital role to handling the complex congenital anomaly. Consultation with expert such as a perinatologist, pediatric cardiologist, and pediatric cardiac surgeon is necessary to plan appropriate interventions. In the second case, pregnancy underwent a cesarean section to safely deliver the newborn. Considering the poor of prognosis, termination immediately may be considered if the diagnosis is made before the viability period. After birth, repair of sternal, diaphragmatic, and pericardial defects can be attempted simultaneously. Surgical correction is often challenging due to thoracic cavity hypoplasia and the inability to cover the ectopia heart. Early detection and management of any intracardiac anomalies are crucial because congenital heart disease is a major cause of morbidity in neonates period. Due to the rarity of ectopia cordis cases, there is no standardized technique established. However, the general surgical approach typically involves closing the chest wall and sternal defect, repairing the omphalocele, relocating the heart into the thorax, and addressing any intracardiac defects [18].

In 2021, Yıldız et al. in Turkey reported four cases of ectopia cordis including abdominal, thoracoabdominal, partial thoracoabdominal, and sternal midline defects who underwent surgery. They concluded that the level and degree of heart exposure will inhibit survival rate in the early life, and complex repair of intracardiac defects may be challenging in cases requiring shunting for long-term palliation. The same year in Iraq, Aboud et al. reported a case of complete thoracic ectopia cordis, but due to poor condition, the newborn passed at 36 h before any intervention performed [19,20].

In the first case, newborn immediately provided in the NICU after birth with only ectopia cordis. A Bogota bag was temporarily placed as protection for the cardiac. Echocardiography shows a large ASD and large PDA. At 5 days of age, the newborn having the symptoms and signs of sepsis. On day 9, surgery was performed to implant bovine pericardium on the newborn 's cardiac. The newborn 's condition worsened with multiple complications including sepsis, hyponatremia, hyperkalaemia, acute kidney injury (AKI), hypoalbuminemia, and metabolic acidosis. The newborn was unable to survive, succumbing to sepsis on day 14. Infants with external cardiac conditions are particularly susceptible to infections. Long-term prognosis of these cases relies on the success of surgery and the medical team's ability to manage the newborn complications [21,22].

In the second case, the newborn underwent immediate care in the NICU after birth. Soon after birth, the newborn was diagnosed with partial ectopia cordis, large PDA, large ASD and omphalocele suggesting Pentalogy of Cantrell. Intensive care was provided in the NICU including ventilator support, fluid therapy, and antibiotics (ampicillin and gentamicin). The pediatric surgeon managed the omphalocele by closing the defect with Burnazin ointment under moist and dry dressings. Management plans were made to implant bovine pericardium as the protection for the newborn 's cardiac. But the procedure has been postponed because epithelialization of the pericardial tissue has occurred. The newborn's condition remains stable at one month follow-up.

Ectopia cordis exposes the heart to environmental pathogens, significantly increasing the risk of infection and sepsis. Early postnatal interventions, including sterile dressing and systemic antibiotics, are essential strategies to prevent neonatal infection and sepsis. Zhang et al. (2014) in China reported lower post-surgical infections with early antibiotic therapy, while Araujo Júnior et al. (2023) in their multicenter study found successful infection control in surgical survivors. Post-operative wound care and sterile techniques remain crucial, along with a multidisciplinary approach to long-term infection management. Integrating these preventive and therapeutic strategies improves neonatal outcomes in EC cases [15,16]. In the second case, the newborna received a combination of ampicillin and gentamicin to prevent infection and have better outcome.

Ectopia cordis (EC) is primarily associated with midline fusion defects, arising due to disruptions in ventral folding morphogenesis, a process regulated by Bone Morphogenetic Protein 2 (BMP2), which is critical for heart tube formation and ventral body wall closure. Chromosomal abnormalities, particularly trisomy 18 and Turner syndrome, have been frequently reported in EC cases, suggesting a genetic predisposition [23]. Early genetic testing and fetal imaging techniques, such as fetal MRI and 3D ultrasound, can aid in the early identification of EC, allowing for informed parental counseling and pregnancy management [24].

In cases of sex chromosome aneuploidies, significant risk factors including living place, paternal chemical exposure, low parental education levels, paternal smoking and alcohol consumption, and a history of recurrent abortus. Screening for chromosomal abnormalities in fetuses aims to enhance awareness of risk factors, screening tests, diagnostic examinations, and family counseling. The ultimate goal is early detection in every pregnancy to prepare both parents. For future pregnancy prevention, genetic counseling plays a crucial role, especially for families with a history of congenital anomalies. Chromosomal microarray analysis and whole-exome sequencing are recommended to assess genetic predispositions. Prenatal screening, including first-trimester ultrasound, fetal echocardiography, and non-invasive prenatal testing (NIPT), enables early detection of chromosomal anomalies associated with EC. Additionally, preconception folic acid supplementation and nutritional optimization, particularly ensuring adequate levels of zinc and vitamin A, may support proper midline development and reduce congenital defect risks. Maternal exposure to teratogens, including alcohol, tobacco, high-dose retinoids (Vitamin A derivatives), and certain infections such as rubella and cytomegalovirus, has been linked to an increased risk of congenital heart defects. Avoidance of these environmental risk factors is recommended to improve fetal outcomes. In cases where EC is associated with genetic syndromes, preimplantation genetic testing (PGT) during in-vitro fertilization (IVF) allows for the selection of embryos without chromosomal abnormalities, reducing the likelihood of recurrence. While most cases of EC are sporadic, a combination of genetic counseling, prenatal screening, maternal nutritional support, and avoidance of teratogens can help reduce the risk of EC in future pregnancies. Continued research into genetic markers and environmental risk factors is essential to improve early detection and refine prevention strategies [23,24].

When planning future pregnancies for individuals with a history of congenital defects, the medical team must adopt a careful and coordinated approach. Comprehensive genetic counseling is essential to assess the risk of genetic abnormalities, along with the development of tailored treatment plans that include specific medication adjustments and close

monitoring throughout the pregnancy. Multidisciplinary consultations involving obstetricians, cardiologists, and geneticists are critical. Providing thorough information, emotional, and psychological support is also crucial to preparing for future pregnancies. The primary goal is to minimize potential complications and ensure the health and well-being of both the mother and the fetus.

5. Conclusion

The management depends on evaluation finding and involving corrective surgery or symptomatic management. Maternal prognosis was good, in pregnancy and after the delivery while fetus prognosis was bad. For the next pregnancy plan, complication risk assessment is required with the help of medical team and genetic counsellor.

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ijscr.2025.111353.

Consent statement

Written informed consent was obtained from the patient's parents for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical statement

Ethics approval was not required for this case report as it does not constitute research involving human participants under the guidelines of The Health Research Ethics Committee. This aligns with institutional guidelines, which exempt single case reports or case series focused on clinical observations from requiring ethical review. However, all necessary permissions and consents were obtained in accordance with institutional policies.

Funding

None.

Declaration of competing interest

None.

References

[1] F. Mraihi, J. Basly, A. Mezni, Z. Ghali, M. Hafsi, D. Chelli, The pentalogy of Cantrell: a rare and challenging prenatal diagnosis, Int. J. Surg. Case Rep. 112 (2) (2023) 1–8941.
[2] E. Dosedla, D. Fric, P. Calda, Prenatal diagnosis of pentalogy of Cantrell in the third trimester, Prague Med. Rep. 110 (1) (2009) 85–90.
[3] E. Araujo Júnior, M.C. Carrilho, B.R. Toneto, J.C. Guilhen, Pentalogy of Cantrell: prenatal diagnosis, delivery, and immediate postnatal surgical repair, J Neonatal Surg. 6 (2) (2017) 32.
[4] M. Fazea, M. Alhameli, F. Ahmed, M.R. Askarpour, W. Murshed, A. Jarwsh, et al., Pentalogy of Cantrell associated with ectopic Cordis: a case report, Pediatr Heal Med Ther. 13 (8) (2022) 283–287.
[5] L. Yeo, S. Luewan, D. Markush, N. Gill, R. Romero, Prenatal diagnosis of dextrocardia with complex congenital heart disease using fetal intelligent navigation echocardiography (FINE) and a literature review, Fetal Diagn. Ther. 43 (4) (2018) 304–316.
[6] P.S. Rao, N.S. Rao, Diagnosis of dextrocardia with a pictorial rendition of terminology and diagnosis, Children 9 (12) (2022) 1–6.
[7] C. Sohrabi, G. Mathew, N. Maria, A. Kerwan, T. Franchi, R.A. Agha, The SCARE 2023 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int J Surg Lond Engl. 109 (5) (2023) 1136.
[8] P. Amitabh, R. Sharan, A. Talapatra, Ectopic cordis, Images Clin Neonatol. 1 (2) (2012) 166–167.
[9] A. Cabrera, D. Rodrigo, M.T. Luis, E. Pastor, J.M. Galdeano, S. Esteban, Ectopic cordis and cardiac anomalies, Rev. Esp. Cardiol. 55 (11) (2002) 1209–1212.
[10] S.M. Mohamed, H.M. Isa, A.K. Sandhu, Ectopic Cordis as a lethal neonatal condition: a case report from Bahrain and a literature review, Case Rep Pediatr. 8 (3) (2022) 1–5.
[11] J. Theola, N.M. Yakub, V.R. Yudianto, B.C. Sinaga, Defek Septum Ventrikel: Diagnosis dan Tata Laksana, Cermin Dunia Kedokt. 50 (3) (2023) 1–7.
[12] W. Dakkak, M.H. Alahmadi, T.I. Oliver, Ventricular septal defect. [Updated 2024 Apr 14], in: StatPearls [Internet], StatPearls Publishing, Treasure Island (FL), 2024 Jan. Available from: https://www.ncbi.nlm.nih.gov/books/NBK470330/.
[13] Society for Maternal-Fetal Medicine, B.R. Benacerraf, B. Bromley, A.C. Jelin, Hypotelorism, Am. J. Obstet. Gynecol. 221 (5) (2019) 1–7.
[14] Leonte L. Dordea, Fetology., Diagnosis and Management of the Fetal Patient, Acta Endocrinol. 9 (4) (2013) 1–7.
[15] E. Araujo Júnior, et al., Ectopia cordis: prenatal diagnosis, perinatal outcomes, and postnatal follow-up of an international multicenter cohort case series, J. Matern. Fetal Neonatal Med. (2023) 2203791, https://doi.org/10.1080/14767058.2023.2203791.
[16] X. Zhang, Q. Xing, J. Sun, X. Hou, M. Kuang, G. Zhang, Surgical treatment and outcomes of pentalogy of Cantrell in eight patients, J. Pediatr. Surg. 49 (8) (2014) 1335–1340, https://doi.org/10.1016/j.jpedsurg.2014.06.003.
[17] M.J. Berry, et al., Ectopia cordis: a novel palliative care technique, BMJ Support. Palliat. Care 7 (1) (2017) 102–104, https://doi.org/10.1136/bmjspcare-2015-000946;
M. Schipper, M.G. Slieker, P.H. Schoof, et al., Surgical repair of ventricular septal defect; Contemporary results and risk factors for a complicated course, Pediatr. Cardiol. 38 (2) (2017) 264–270.
[18] L.W. Irshaid, N. Elfky, B. Ahmed, Prenatal detection of critical congenital heart disease, Donald Sch J Ultrasound Obstet Gynecol. 10 (2) (2016) 131–135.
[19] V. Lack, C. Oetzmann Von Sochaczewski, K. Naidoo, J. Loveland, Pentalogy of Cantrell with thoracoabdominal ectopia cordis: attempted surgical correction and review of recent literature to aid prognostication prior to surgery, J Pediatr Surg Case Rep 3 (11) (2015) 476–480.
[20] K. Yıldız, M. Kır, Ş.B. Uğurlu, H.Z. Genç, N. Ünal, Four ectopia cordis cases surgically managed with different strategies, Turkish J Thorac Cardiovasc Surg. 29 (1) (2021) 114–118.
[21] M.J. Aboud, M.J. Aboud, M. Abudi, N, M Joudi H, M Joudi Z., Complete thoracic ectopic Cordis, dilemma of the outcome: a case report, Surg Case Reports. 3 (4) (2021) 1–4.
[22] G.C. Gabriel, M.W. Russell, G. Wang, et al., Congenital heart defects and genetic syndromes: a review of midline defects and ectopia cordis associations, Am. J. Med. Genet. C: Semin. Med. Genet. 166C (4) (2014) 310–326, https://doi.org/10.1002/ajmg.c.31406.
[23] L. Cripe, J. Cnota, M.D. Taylor, Ectopia cordis: advances in prenatal diagnosis, surgical management, and outcomes, J. Congenit. Cardiol. 6 (1) (2022) 6850305, https://doi.org/10.1155/2022/6850305.
[24] K.M. Alshamiri, A.Z. Albriek, T.W. Farrag, M.Q. Alshamiri, Ectopic cordis in an adult patient with COVID-19: a case report and literature review, Clin. Case Reports 10 (2) (2022) 1–5.