CASE REPORT

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Clinical outcomes and surgical intervention using Bogota bag and bovine pericardium in patient diagnosed with ectopia cordis: a case series

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ABSTRACT

Introduction: Ectopia cordis is a rare congenital anomaly characterized by the heart's partial or complete displacement, often associated with ventral wall defects. This condition poses significant challenges due to potential cardiac and respiratory issues. Effective management typically involves reconstructive surgery using techniques like the Bogota bag and bovine pericardium. This case series aims to describe the clinical manifestations, associated disorders, surgical interventions, and outcomes of patients diagnosed with ectopia cordis.

Case description: Case 1 involved a premature baby with complete thoracal ectopia cordis, significant atrial septal defect (ASD), patent ductus arteriosus (PDA), moderate tricuspid regurgitation (TR), and sepsis. The patient received two surgeries, including an initial cover with a modified Bogota bag and a subsequent replacement with bovine pericardium. Unfortunately, the patient died from sepsis. Case 2 involved a full-term baby with partial thoracoabdominal ectopia cordis, tetralogy of Fallot (TOF), significant PDA, ASD, and omphalocele. This patient was managed with a continuously replaced sterile stoma bag and had a positive outcome.

Conclusion: Ectopia cordis requires prompt and multidisciplinary management to improve outcomes. The prognosis varies based on the extent of the anomaly, associated defects, and the effectiveness of surgical treatments. Prenatal screening is crucial, but timely postnatal care is also essential. In cases with poor prognoses, pregnancy termination might be considered.

Keywords: Bogota bag, bovine pericardium, congenital anomaly, ectopia cordis, reconstructive surgery.
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INTRODUCTION

Ectopia cordis is a very rare congenital anomaly. This congenital anomaly can be detected at 18 to 23 weeks of gestation. Ectopia cordis can be detected routinely at gestational age above 18 weeks through a four-chamber view examination.[1,2] Congenital anomalies in cases of ectopia cordis are characterized by isolated malformations or malformations associated with other types of extensive ventral wall abnormalities, including the thoracic, abdominal, or both areas.[2,3] This abnormality can occur due to failure of fusion of the ventral body wall in the midline line during development in the womb.[3]

Epidemiologically, only around 5.5-7.9 babies out of 1 million babies experience ectopia cordis. Most common in baby girls. The majority of them are stillborn or die within three days after birth.[3,4] In 1997, ectopia cordis was classified into five types based on the position of the heart, namely 1) Cervical, where the heart is located in the neck with the sternum usually still intact; 2) Thoracocervical, where the heart is partially located in the cervical region, but the upper part of the sternum is split; 3) Thorax, where the sternum is split or absent altogether, and the heart is located partially or entirely outside the chest wall; 4) Thoraco-abdominal, this disorder is usually followed by Cantrell's syndrome (Pentalogy of Cantrell); and 5) Abdominal, where the heart passes through the defect in the diaphragm and enters the abdominal cavity.[3]

Clinical manifestations that arise in patients suffering from ectopia cordis include difficulty breathing due to disruption of the heart's pumping mechanism, which is related to intrinsic anomalies of the heart. In some cases, other clinical manifestations can also be found, such as fever. Fever is caused by the absence of skin and pericardium, making it easy for bacterial contamination to occur. In some severe cases, infection in babies can cause sepsis. Therefore, as long as the heart's contact with the outside world (atmospheric contact) is not improved, it will be challenging to treat sepsis to produce good outcomes for the patient.[4]

The diagnosis of ectopia cordis during pregnancy depends on how far the malformation is formed and the type of ectopia cordis. Complete ectopia cordis,

where the entire heart is outside the chest wall, can be diagnosed by ultrasound examination during pregnancy, while thoracoabdominal ectopia cordis may be difficult to diagnose via ultrasound. Several studies report that the diagnosis of ectopia cordis can be made at the youngest age, namely 10 weeks of gestation. Meanwhile, other sources write that the diagnosis of ectopia cordis can be made at the age of 18 to 23 weeks of pregnancy.[2,5]

Management of ectopia cordis can be done with reconstructive surgery, namely reconstructive surgery of the thoracic cavity and heart. There are several considerations in the management of ectopia cordis, one of which is to consider cardiac correction surgery before palliative closure. Using materials or skin, palliative surgery focuses on closing the heart from contact with the outside world. Materials like biomaterials may be considered if skin closure is inadequate. The biomaterial that can be used is bovine pericardium because of its compatibility and causes fewer complications than other synthetic materials.[6] After reconstructive surgery, patients suffering from ectopia cordis with sufficient birth weight and full-term age show better outcomes than patients who were born prematurely and with a birth weight below normal.[5] This case series aims to describe the clinical manifestations, associated disorders, surgical interventions, and outcomes of patients diagnosed with ectopia cordis.

CASE DESCRIPTION

Case 1

In this first case, the patient was a 0-day-old preterm neonate who was born at OK COT at Dr. Hospital. Mohammad Hoesin Palembang (RSMH) from a G1P0A0 mother 36-37 weeks pregnant, has not labored yet with Parturition Premature Imminent (PPI) + Cervical Shortening (1.9 Cm) single life fetus cephalic presentation with congenital abnormalities (Ectopic cordis > 70%) + persistent bradycardia + Intrauterine Growth Restriction (IUGR). The patient's birth history: the patient was born by cesarean section assisted by a doctor with sufficient amniotic fluid, clear, and had no umbilical cord entanglement, and cephalic was born first. The baby was born crying immediately, with an

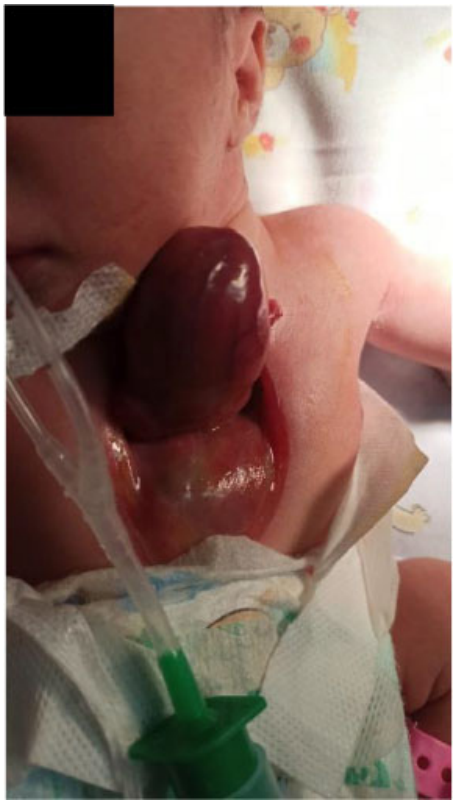

Figure 1. The location of the patient's heart is outside the chest wall.

Apgar score of 8/9—birth weight 1,955 grams, body length 40 cm, and head circumference 31 cm.

In this case, there were manifestations in the form of shortness of breath with a respiratory rate of 44 times per minute, and it was found that the heart was outside the chest wall and there was open skin around the heart accompanied by minimal intercostal retraction (Figure 1).

Supporting examinations revealed leukocytosis, prolonged prothrombin time (PT), and activated partial thromboplastin time (APTT). Leukocytosis in the examination results can indicate infection, while PT and APTT prolongation can be related to blood coagulation, which is influenced by several factors, such as disseminated intravascular coagulation (DIC) coagulopathy, liver function disorders, hypothermia, and vitamin K deficiency. Based on a chest X-ray examination, the impression of the heart outside the thoracic cavity is obtained. Bronchovascular patterns were normal. There were no signs of intestinal obstruction. Hepatomegaly. Meanwhile, based on an echocardiography

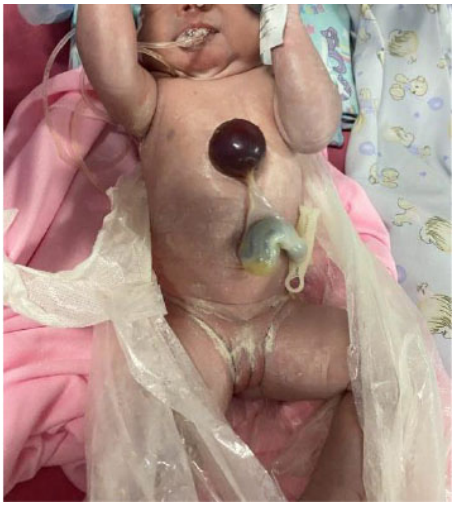

Figure 2. The location of the patient's heart is outside the chest wall and part of the intestine is wrapped in a membrane 2 cm long from the paraumbilical.

examination, it was found that there was complete ectopia cordis, there was an atrial septal defect (ASD), and a ventricular septal defect (VSD).

In this case, the patient was diagnosed as a preterm neonate according to the gestational age (36 weeks), low birth weight with respiratory distress down score 2 et causa transient tachypnea of the newborn with a differential diagnosis of hyaline membrane disease accompanied by complete ectopia cordis. In this patient, reconstructive surgery was performed to cover the exposed heart with Bogota bag material. In the patient, the location of the heart was found to be entirely outside the chest cavity, and large ASD and large patent ductus arteriosus (PDA) were also found. During the operation, there was no hemodynamic deterioration. The patient was then treated in the Neonatal Intensive Care Unit (NICU) with clinical monitoring, leukocyte count, C-reactive protein (CRP), and echocardiography. Post-operative echocardiography examination revealed large ASD, large patent ductus arteriosus (PDA), and moderate tricuspid regurgitation. Antibiotics are still given to prevent infection.

On the fifth day after surgery, another follow-up was carried out on the patient, and it was found that there was an improvement in respiratory distress, decompensation cordis Ross score 5,

complete ectopic cordis, significant atrial septal defect, large patent ductus arteriosus, moderate tricuspid regurgitation, sepsis.

Case 2

In the second case, the patient was a 0-day-old baby girl born at Dr. Hospital. Mohammad Hoesin Palembang Central Operation Theatre to a G1P0A0 mother 38-39 weeks pregnant, has not labored yet with multiple congenital abnormalities (suspect Dextrocordis with ectopic cordis + VSD + hypotelorism) + fetus tended to be small + suspect Partial pentalogy of Cantrell + cervical shortening (1.0 cm). The patient was born at term with a birth weight of 2800 grams, a body length of 45 cm, and a head circumference of 32 cm. The patient was born without crying immediately with an Apgar score of 7/8.

From the results of the physical examination, it was found that there was shortness of breath with a respiratory rate of 52 times per minute with SpO2 82% using NIV FiO2 40% PIP 18 PEEP 5, and the heart appeared to be outside the chest cavity (Figure 2), intercostal retraction was present, part of the intestine was visible wrapped in membrane for 2 cm from paraumbilical. From the results of the laboratory examination the results were within normal limits. From the patient's thoracoabdominal x-ray examination, it was revealed that there was a radiopaque mass in the midline between the thorax and the abdomen. In contrast, the results of the echocardiography examination showed that there was tetralogy of Fallot, significant PDA, and ASD.

The patient was diagnosed as a term neonate according to gestational age with respiratory distress down score 2 et causa TTN with partial ectopia cordis + tetralogy of Fallot + large PDA + large ASD + omphalocele. The patient suffers from Pentalogy of Cantrell (PC), a congenital abnormality or malformation involving the heart, pericardium, diaphragm, sternum, and abdominal wall.

The patient underwent cross-professional consultations with pediatric cardiology consultants, thoracic and vascular surgery consultants, and pediatric surgery. The pediatric cardiology consultant recommended that the protruding heart be closed with sterile gauze moistened with 0.9% NaCl dripped every 6 hours and then covered with a colostomy bag from the consultant, it was recommended to install a Bogota bag in the operating room, and from the pediatric surgery consultant, it was recommended to give silver sulfadiazine cream the omphalocele area and cover with sterile gauze moistened with 0.9% NaCl.

DISCUSSION

Ectopia cordis presents a significant clinical challenge due to the complexity and variability of the associated anomaly. Early diagnosis, rapid stabilization, and a multidisciplinary approach are critical to improving patient outcomes. The prognosis depends mainly on the extent of the ectopia, associated abnormalities, and the feasibility of surgical correction.

In this neonate case, the patient was a premature baby with a gestational age of 36-37 weeks who was born via cesarean section at Dr. Hospital. Mohammad Hoesin Palembang. The birth was carried out with indications of premature rupture of membranes (PROM) and cervical shortening, accompanied by significant congenital abnormalities, namely ectopic cordis, where the baby's heart is located outside the thoracic cavity, as well as clinical manifestations in the form of persistent bradycardia and IUGR. The patient was born with a body weight of 1955 grams, a body length of 40 cm, and a head circumference of 31 cm. The Apgar score obtained was 8/9, indicating a relatively stable condition immediately after birth.

However, shortness of breath and an increased respiratory rate (44 times per minute) indicated respiratory distress in this baby. On physical examination, the heart appears outside the chest wall, with the skin covering the heart exposed and minimal intercostal retraction. Supporting examinations show leukocytosis, which indicates potential infection, and prolongation of PT and APTT, which can be related to coagulopathy, impaired liver function, or vitamin K deficiency. Chest x-ray results show the heart is located outside the thoracic cavity and hepatomegaly but without signs of intestinal obstruction. Echocardiography showed complete ectopia cordis, ASD, and VSD.

The primary diagnosis for this patient is neonatal respiratory distress with transient tachypnea of the newborn (TTN), with a differential diagnosis of hyaline membrane disease, as well as congenital abnormalities and complete ectopia cordis. Reconstructive surgery is carried out to cover the exposed heart using Bogota bag material. During surgery, there was no significant hemodynamic deterioration, although a large ASD and a large PDA were found.

Post-operatively, the patient was treated intensively in the NICU, closely monitoring clinical parameters, leukocytes, CRP, and echocardiography. Post-operative echocardiography showed large ASD, significant PDA, and moderate tricuspid regurgitation. Antibiotics were continued to prevent infection, and on the fifth post-operative day, there was an improvement in respiratory distress, but the patient still showed cordis decompensation with a Ross score of 5, as well as persistent congenital abnormalities.

In cases of neonates with ectopic cordis, the use of a Bogota bag in surgical intervention aims to protect the heart, which is located outside the thoracic cavity. Ectopic cordis is a rare congenital disorder in which the baby's heart is open and exposed, increasing the risk of infection and damage to this vital organ. As a surgical reconstruction material, the Bogota bag covers the exposed heart, reducing the risk of infection by providing a protective layer that protects the heart from direct exposure to the external environment that can cause contamination and infection. Also, closure with a Bogota bag maintains hemodynamic stability, avoiding the negative impact of temperature changes and mechanical trauma, which can affect heart function. This step is crucial to avoid a drop in blood pressure or circulation problems, which could worsen the baby's condition. The Bogota bag also allows the time needed for planning and carrying out more complex thoracic reconstructive surgery, including closure of heart defects and repair of congenital anomalies such as ASD and PDA. In addition, by paying attention to the results of laboratory examinations, which indicate coagulation disorders that may occur due to liver

disorders as indicated by hepatomegaly in patients, the Bogota bag helps minimize the risk of bleeding and infection, providing additional protection during and after surgery. In this way, using a Bogota bag not only physically protects the baby's heart but also allows the medical team to carry out long-term care effectively, ensuring optimal recovery and reducing the risk of further complications.[5,7,8]

In the second case, a 0-day-old newborn girl with a body weight of 2800 grams and a length of 45 cm had several congenital abnormalities, including suspected dextrocardia, ectopic cordis, and VSD, as well as hypotelorism and cervical shortening. The baby was born at term with an Apgar score of 7/8, indicating the need for respiratory support with SpO2 82% and use of NIV. Physical examination reveals shortness of breath, heart located outside the chest cavity, and intercostal retractions. Part of the intestine is seen covered in membrane in the paraumbilical area. Laboratory examination was within normal limits, but a thoracoabdominal X-ray showed a radioopaque mass in the thoracic-abdominal midline. Echocardiography identified tetralogy of Fallot, significant PDA, and large ASD.

The patient, a term neonate with complex congenital abnormalities, was diagnosed with respiratory distress, partial ectopia cordis, tetralogy of Fallot, significant PDA, ASD, and omphalocele. The primary diagnosis was the Pentalogy of Cantrell, which included malformations of the heart, pericardium, diaphragm, sternum, and abdominal wall. After cross-professional consultation, recommended treatment includes covering the exposed heart with wet sterile gauze and a colostomy bag by a pediatric cardiologist, installing a Bogota bag in the operating room by a thoracic and vascular surgeon, and treating omphalocele with silver sulfadiazine cream and wet sterile gauze by a pediatric surgeon. Management from various cross-specialty consultants aims to treat congenital abnormalities effectively and comprehensively.

In the case of this neonate, the management recommended by cross-specialty consultants reflects a holistic and integrated approach to managing the complex congenital abnormalities present in the case. The pediatric cardiology consultant recommended covering the exposed heart using sterile gauze soaked in 0.9% NaCl and closing with a colostomy bag. It aimed to protect the heart outside the chest cavity from the risk of infection and contamination, as well as helping to maintain moisture and prevent further tissue damage. Installation of a colostomy bag also provides additional protection by covering areas particularly susceptible to infection.[5,7,8]

Advice from consultant thoracic and vascular surgeons to use the Bogota bag in the operating room focuses on protection and temporary closure of the thoracic defect. The Bogota bag covers and protects the exposed heart, helps maintain hemodynamic stability during surgery, and facilitates the planning and implementation of further reconstructive treatment. Fitting a Bogota bag also contributes to the regulation of body temperature and protection from mechanical trauma.[5,7,8]

Meanwhile, the pediatric surgical consultant recommended administering silver sulfadiazine cream to the omphalocele area and covering it with wet sterile gauze. Omphalocele, in which part of the intestine is encased in a membrane, requires special attention to prevent infection and maintain a clean environment around the abdominal defect. Silver sulfadiazine cream aims to protect and prepare the area for further closure, while wet sterile gauze helps maintain moisture and protects the area from contamination.[5,7,8]

The involvement of the intracardiac malformation, associated malformations, and the degree of cardiac exposure of the chest wall determines the prognosis of the case. Even though surgical intervention is carried out, risk factors such as prematurity, low birth weight, and the degree of malformation and related malformations result in unfavorable outcomes in some cases.[7]

Therefore, prenatal diagnosis in cases of ectopia cordis needs to be carried out using an ultrasound examination by looking at the visualization of the heart located on the outside of the thoracic wall. It will help to make further decisions. In patients with a poor expected outcome or prognosis, termination of pregnancy may be considered. Our decision was based on limited research due to constraints in accessing information and resources and a short follow-up time after the therapy. Further evaluation of the effectiveness and safety of Bogota bag and bovine pericardium in patients diagnosed with ectopia cordis is warranted on a larger scale and over a more extended observation period. It would provide more comprehensive insights into the utility of Bogota bag and bovine pericardium in patients diagnosed with ectopia cordis.

CONCLUSION

This case series highlights the diverse presentations and outcomes of ectopia cordis. Although advances in surgical techniques and neonatal care have improved survival rates, this condition remains associated with significant morbidity and mortality. Morbidity and mortality are influenced mainly by risk factors, such as gestational age, birth weight, and the severity of the malformations that occur in the patient. It is also necessary to consider surgery carried out in stages to correct the malformations that occur. Continued cross-specialty research and collaboration is critical to optimizing care for these patients.

DISCLOSURES

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Conflict of Interest
None.

Author Contribution
AF, B, GS, AU, and AN contributed to the manuscript preparation, revising, and drafting. AH, IH, and RN contributed to the design of this study and supervision.

Ethical Consideration
The patient consented to the publication of this case for research purposes or in a journal.

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