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CASE IN IMAGES

Ectopia cordis totalis: case in images

Jayce V Estrera,[1] Delbrynth P Mitchao,[1] Kathleen Rose Descallar-Mata,[1,2]

[1]Department of Surgery, Southern Philippines Medical Center, JP Laurel Ave, Davao City, Philippines

[2]Thoracic and Cardiovascular Surgery Section, Department of Surgery, Southern Philippines Medical Center, JP Laurel Ave, Davao City, Philippines

Correspondence
Jayce V Estrera
ace_jayce_estrera@gmail.com

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Ectopia cordis (EC) is a rare congenital malformation characterized by a complete or partial extrathoracic presentation of the heart,[1 2] with an incidence of only 6 to 8 in a million live births globally.[3 4] It occurs due to failure of maturation of the midline mesoderm and improper formation of the chest and abdomen during embryonic development.[5 6] Depending on the heart's ectopic location, EC is classified into four types—i.e., thoracic (65% of cases), thoracoabdominal (20%), abdominal (10%), and cervical (5%).[1 2 4 7] Only two cases of EC, one thoracic and one thoracoabdominal, from the Philippines had been reported in literature.[8] Cantrell's pentalogy—findings of bifid sternum, absence of the diaphragm, defect of anterior diaphragmatic pericardium, defect of the anterior abdominal wall, and intracardiac defects — usually accompanies the thoracoabdominal type of EC.[1 2]

EC is usually diagnosed prenatally through ultrasonography as early as the first trimester or in the beginning of the second trimester.[2 4] Magnetic resonance imaging, in combination with fetal echocardiography, is used to identify coexistent cardiac anomalies and to monitor fetal development throughout the pregnancy.[2 9]

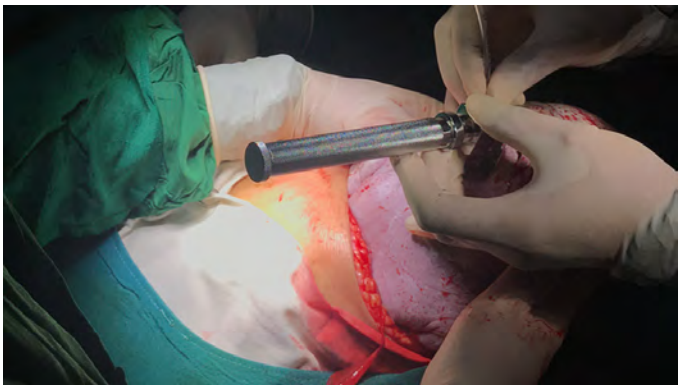
Postnatal surgical management of EC has two stages. The first stage involves soft tissue coverage of the exposed heart using cadaveric skin grafts or prosthetic materials, while the second stage involves relocation of the heart down into the thoracic cavity, reconstruction of the chest wall, and repair of any coexistent intracardiac defect—e.g., Tetralogy of Fallot, pulmonary hypoplasia, ventricular septal defect, and atrial septal defect.[3] The second stage can commence at a later time, when the hemodynamic effects of the first stage have already stabilized.[10] EC is lethal when accompanied by other cardiac malformations,[11] with patient demise occurring within just a few hours or days after birth.[7] The abdominal type of EC holds a better prognosis, since it is less associated with cardiac defects.[12]

The thoracic type of ectopia cordis is usually associated with a poor prognosis, especially when associated with intracardiac defects and extracardiac anomalies. Treatment of this type presents a formidable surgical challenge. However, there have been several reported cases of successful corrective or palliative surgery for EC performed during the neonatal period, infancy, and childhood.[5 13 14] Such success is only possible through early detection and accurate diagnosis of this condition during prenatal consultations, with a multidisciplinary team implementing an optimal delivery and postnatal management strategy.

An 18 year-old primigravid mother at 31 weeks age of gestation was admitted in the Obstetrics ward of our institution due to an ultrasound finding of ectopia cordis during routine prenatal check-up. She and her family were apprised of her condition and the guarded prognosis of the fetus, and she was referred to the Pediatric Surgery Service and Neonatology Service for evaluation and co-management. However, she decided to go home against medical advice before she was fully evaluated. Prior to going home, she received two out of three planned doses of dexamethasone for fetal lung maturation. She denied any comorbidities, smoking and alcoholic beverage drinking history, exposure to environmental risk factors, and history of any infectious illnesses during the whole course of her pregnancy. There was no family history of any such or related congenital abnormality. At 37 4/7 weeks age of gestation, the pregnant mother returned due to a ruptured bag of water and was subsequently readmitted. A multidisciplinary team—composed of Obstetrics, Pediatric Cardiology, Pediatric Thoracic and Cardiovascular Surgery (PTCVS), and Pediatric and Cardiovascular Anesthesiology services—was immediately convened to manage the mother and the baby. Our team, the PTCVS Service, planned and implemented the surgical intervention for the baby.

At this point, our plan was to perform coverage of the exposed heart and to run a series of diagnostic imaging studies to determine the presence of intracardiac defects and other concomitant abnormalities immediately after birth. The results of the diagnostic workup would provide useful information for further surgical planning and management of the patient.

The Obstetrics Service performed an emergency caesarean section due to a non-reassuring fetal status. Upon delivery of the head and upper torso of the neonate, the anesthesiologist immediately performed intubation (Figure 1). The obstetrician delivered a live female neonate



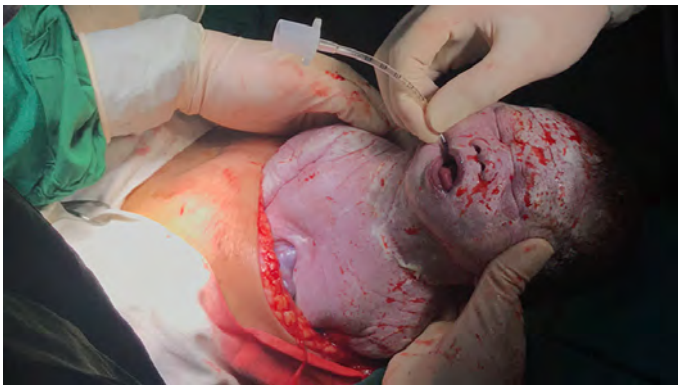


Figure 1 Intubation of the neonate upon delivery of the head and upper torso, prior to discontinuation of the maternal-fetal circulation.

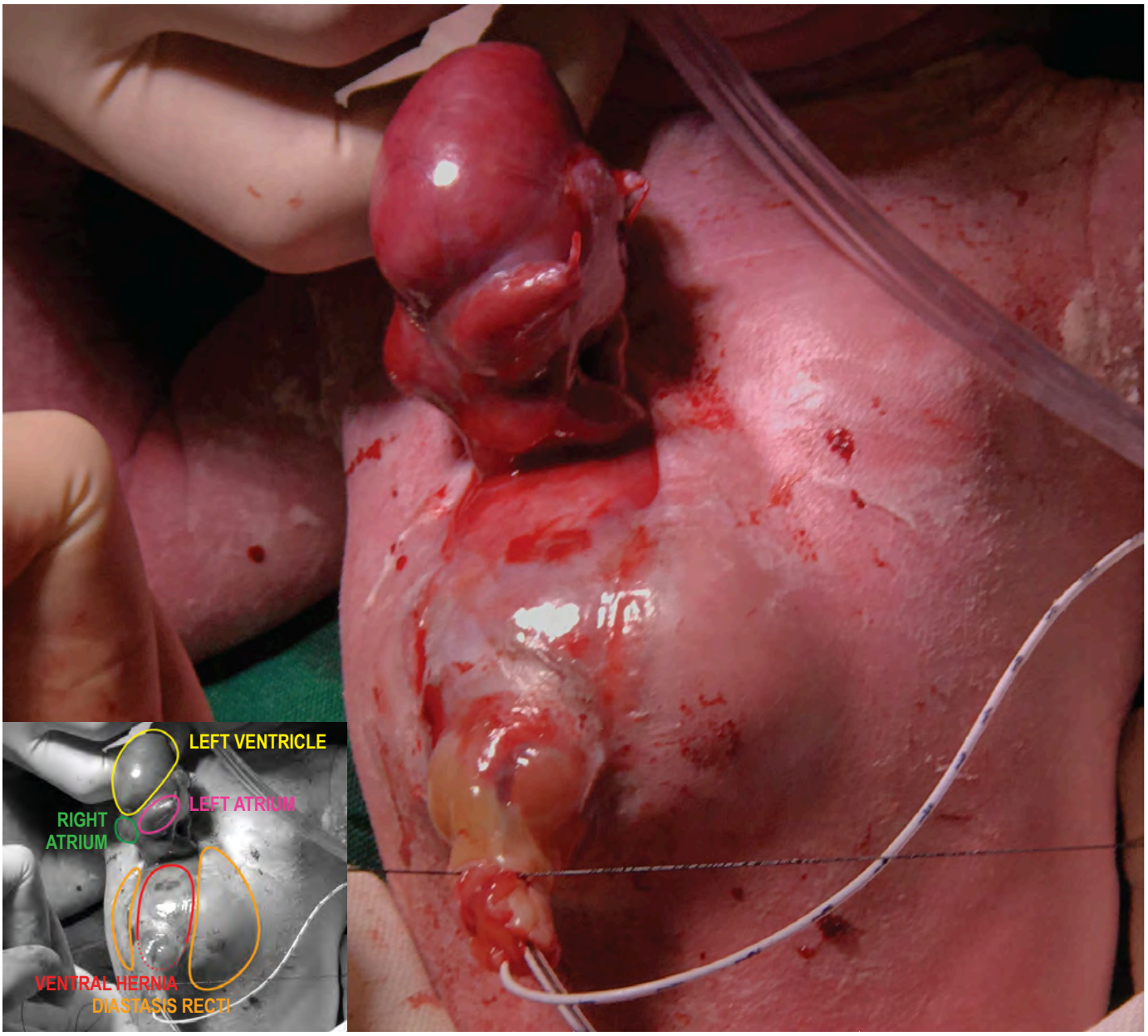


Figure 2 Extrathoracic, upturned heart without pericardium or skin cover. Both the right and left ventricles were directed anterosuperiorly. The right atrium was posteroinferior and to the right of the right ventricle, and the left atrium was posteroinferior and to the left of the left ventricle. Diastasis recti and a ventral hernia are present in the abdominal area.

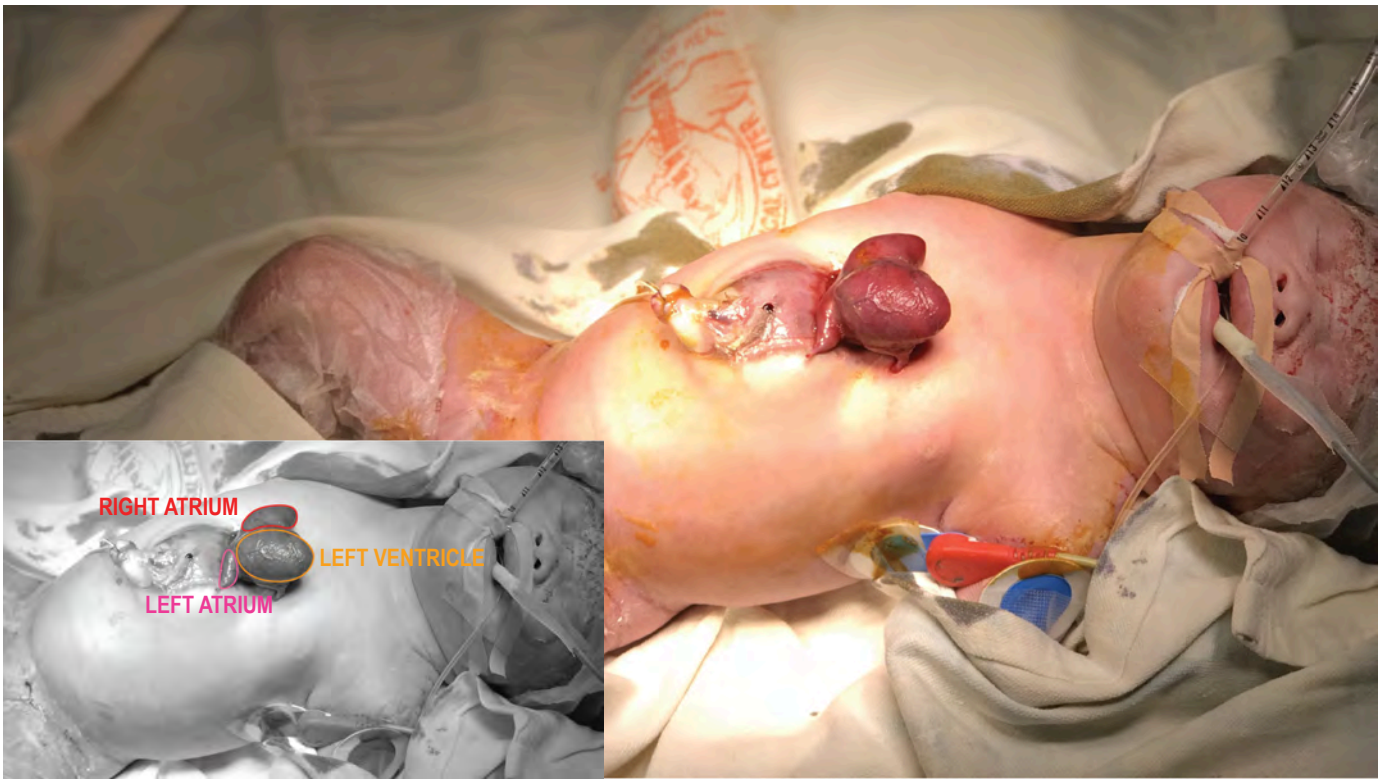


Figure 3 Anterior wall defect extending from the level of the nipples to the umbilicus along the midline, exposing the cardiac chambers. The width of the defect measures 5 cm on the thoracic surface at the level of the nipples, and 6 cm on the abdominal surface at the level of the umbilicus.

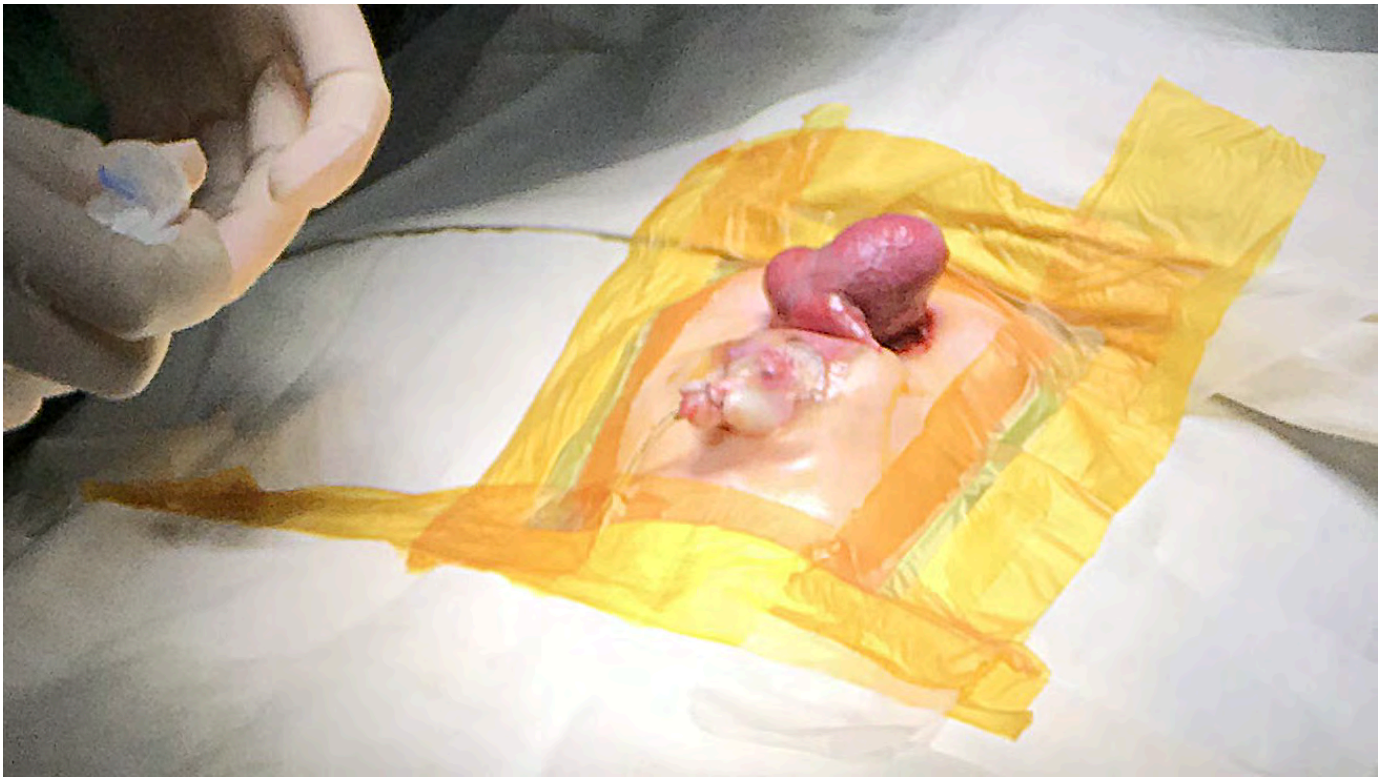


Figure 4 The ectopic heart prior to chest coverage with polytetrafluoroethylene.

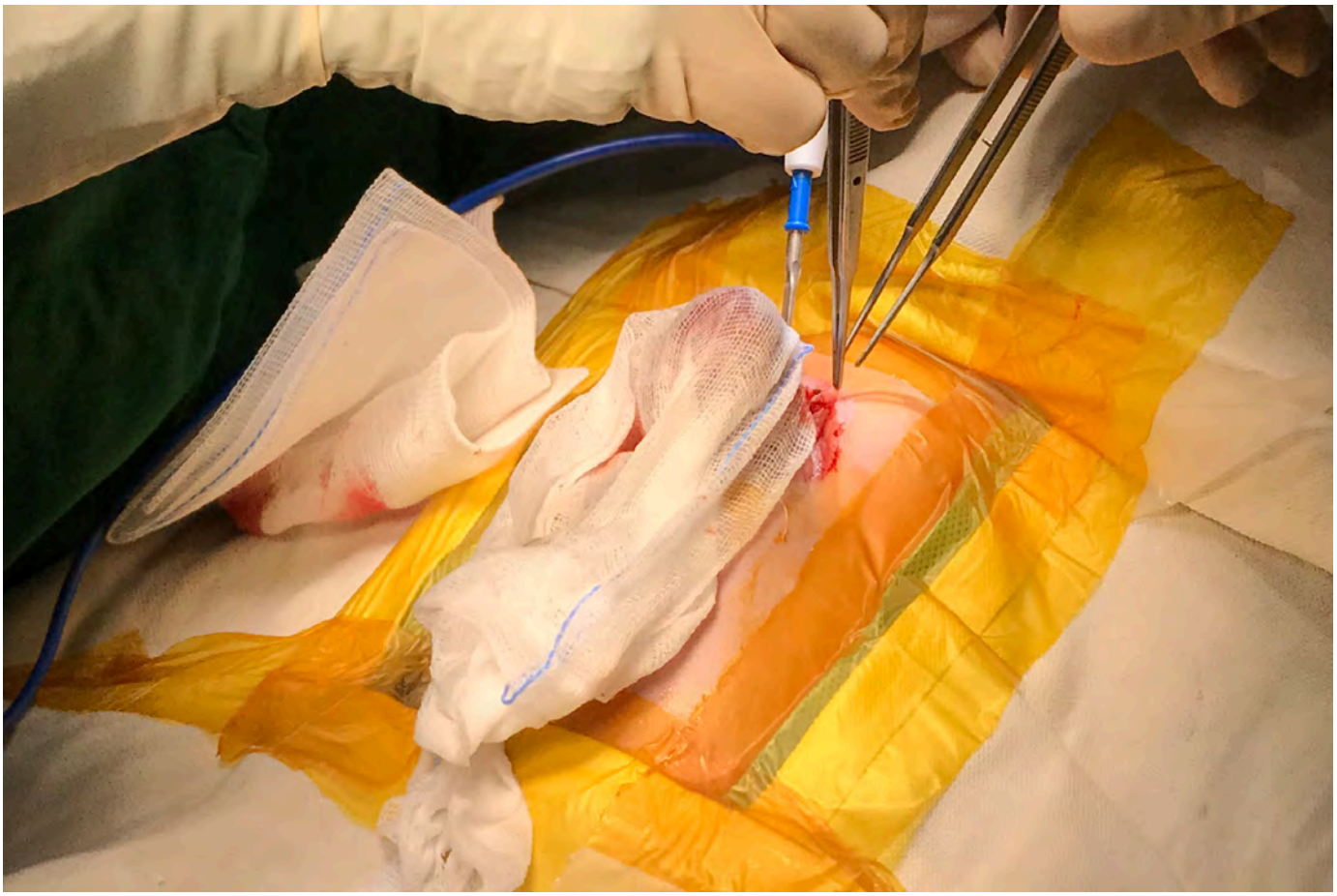


Figure 5 Mobilization of skin and subcutaneous tissue surrounding the ectopic heart. In this picture, we used a sterile gauze to protect and lightly retract the heart during dissection.

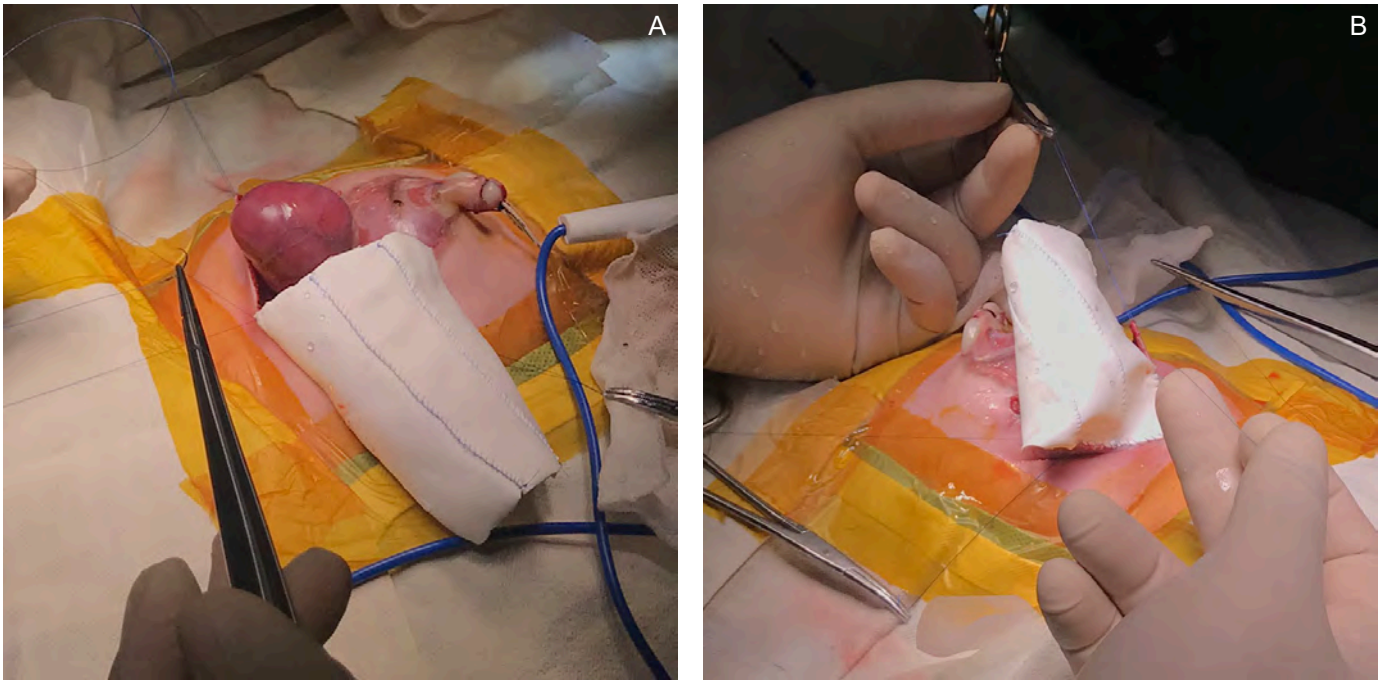


Figure 6 Suturing of a polytetrafluoroethylene patch to the mobilized skin using 5-0 polypropylene.

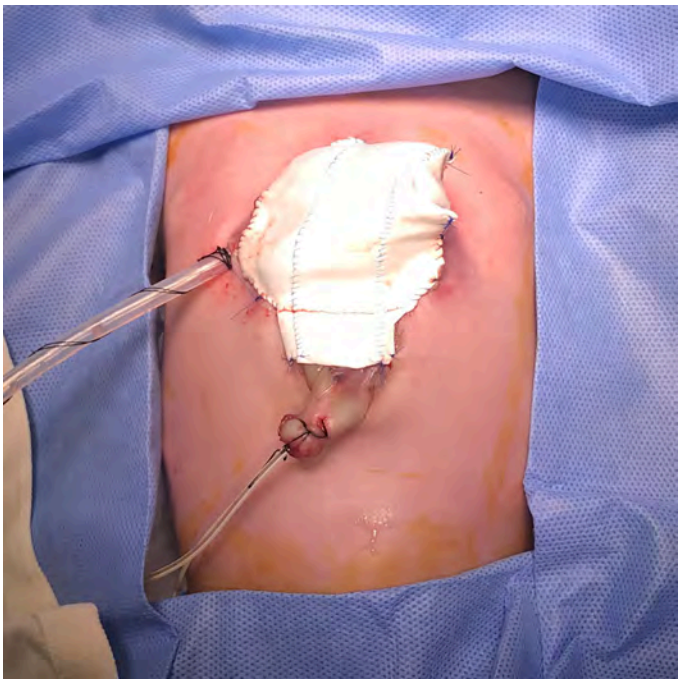


Figure 7 Polytetrafluoroethylene patch fully covering the thoracic wall defect.

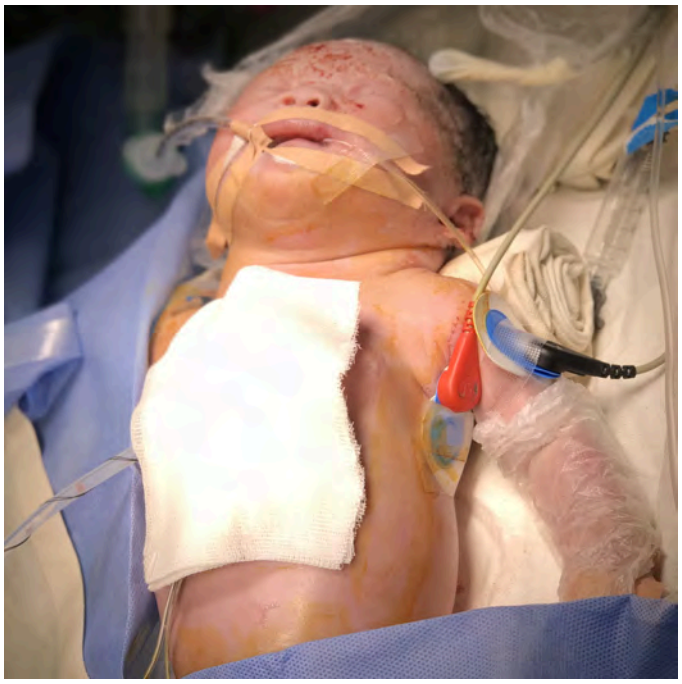


Figure 8 Minimal dressing over the chest and abdomen to prevent restriction of heart movement.

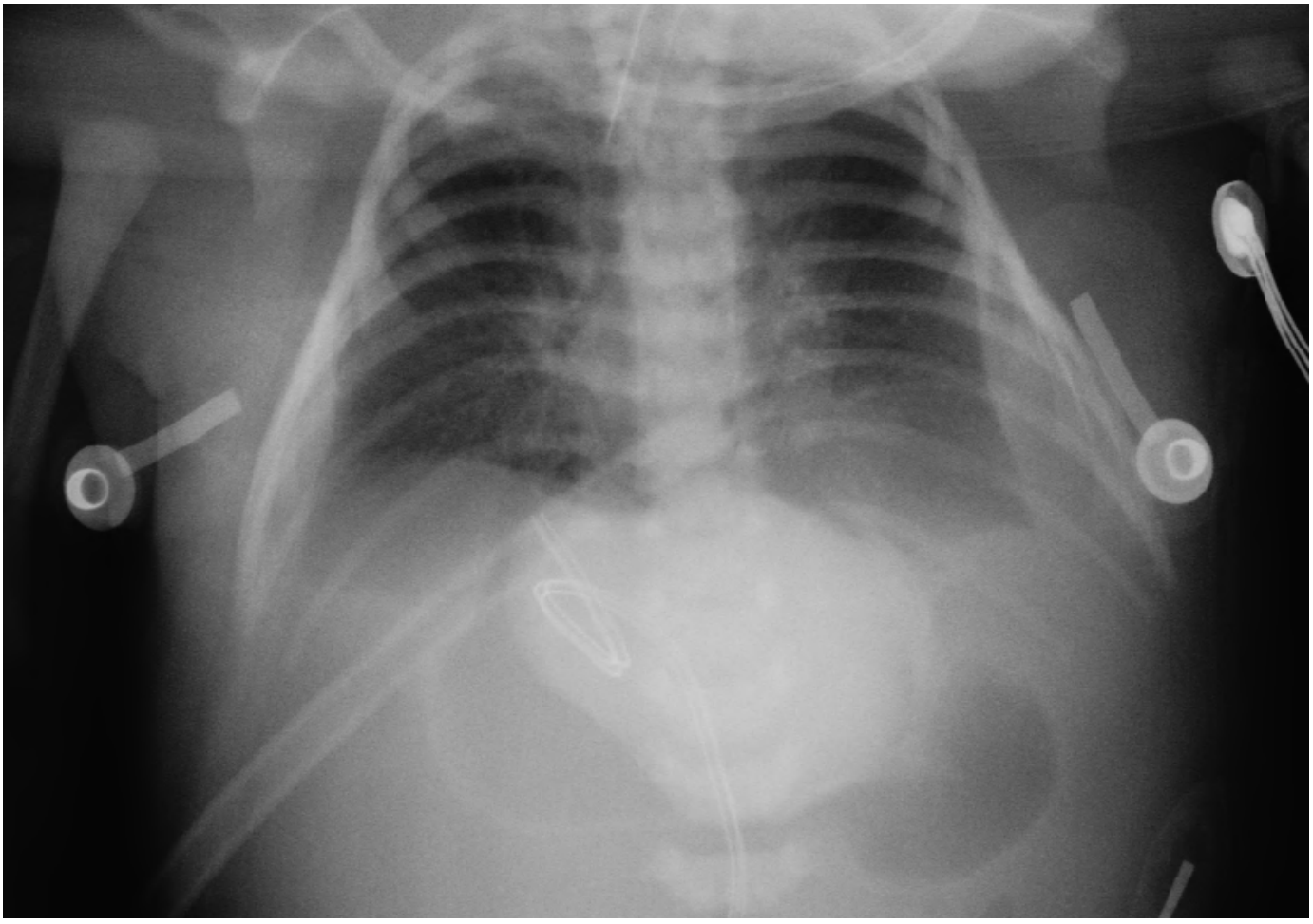


Figure 9 Anteroposterior chest x-ray taken post-coverage of the anterior thoracic wall defect, showing intact hemidiaphragms, and demonstrating loss of normal vascular pattern in the hilar areas of both lungs. The superior portion of the cardiac shadow is located at the level of the 12th thoracic vertebra.

weighing 2.7 kg, with a Ballard Score of 37 weeks. On physical examination, all four chambers of the heart were found to be extrathoracic, with neither pericardium nor skin cover (Figure 2). The heart was loose and unsteady, most especially pronounced when the neonate cried. Anatomically, the heart was upturned, with the two great arteries and four pulmonary veins serving as its base, and with the apex of the ventricles positioned directly anterior. Both the right and left ventricles were directed anterosuperiorly. The right atrium was posteroinferior and to the right of the right ventricle, and the left atrium was posteroinferior and to the left of the left ventricle. The anterior wall defect extended from the level of the nipples superiorly, and down to the umbilicus inferiorly (Figure 3). The widest diameter of the defect measured 5 cm, which was at the level of the T6-T8 vertebrae. From the thoracic defect, the anterior diaphragm was easily visible where the heart sat. Below the diaphragm, the abdominal defect extended 3 cm laterally on each side from the midline. The edges of the rectus muscles could be visualized from the lateral sides of the umbilicus, suggesting a diastasis of the rectus abdominis muscles (Figure 2). There was a ventral supraumbilical hernia ending just beneath the umbilicus, but there was no associated gastroschisis or omphalocele. Our subsequent management of the neonate's condition was based on the diagnosis of complete ectopia cordis, thoracic type.

The Anesthesia Service inducted the neonate under general anesthesia and placed her on mechanical ventilation. We mobilized the skin and subcutaneous tissue surrounding the defect (Figure 5), and sutured a 6 cm x 7 cm roughly oval patch of polytetrafluoroethylene (PTFE) graft using polypropylene 5-0 (Figures 6 and 7). We covered the graft with minimal dressing to avoid compression of the heart and the great vessels (Figure 8). The whole procedure lasted for an hour. At the post-anesthesia care unit, the neonate had several episodes of cyanosis, hypothermia, and bradycardia. We admitted the neonate to the intensive care unit (ICU) and placed her on cardiac monitoring. She had low urine output during the first 12 hours and had no urine output starting on the 24th hour of life. The oxygen saturation also fluctuated between 50% and 98% while at the ICU.

We obtained a postnatal chest radiograph, which revealed an abnormal cardiac configuration and location, absence of the normal splaying of blood vessels at the hilar region, and intact diaphragm (Figure 9).

Throughout the neonate's stay at the ICU, her arterial blood gases suggested metabolic acidosis, which persisted despite continuous correction. She had three episodes of cardiac arrest starting on the 19th hour of life and was resuscitated each time with epinephrine and cardiac massage. On the 4th cardiac arrest, at her 39th hour of life, she was not revived despite aggressive resuscitation. The primary cause of death was intractable metabolic acidosis.

The prognosis of EC, an uncommon condition resulting from an embryologic development defect, is worse when the heart is located completely outside the chest, without overlying pericardium or skin. On prenatal ultrasound done on the third trimester, our patient's heart was noted to be located outside of the chest. She was eventually diagnosed with ectopia cordis totalis, and she survived for a few days after birth. This clinical course is relatively common among patients with complete EC. The management of EC presents a challenge because it requires careful planning very early in the pregnancy, the full cooperation of the pregnant mother, and a proficient multidisciplinary management team. Since the neonate's mother presented at a very late stage in her pregnancy and had poor follow-up until delivery, we did not have ample time to prepare for our neonate's postnatal management. Despite this, the multidisciplinary team—which was convened just prior to delivery—was able to give immediate postnatal care and perform the first stage of EC correction within two hours after delivery. The combination of clinical foresight and detailed management planning by a competent clinical team may lead to better chances of survival of patients born with ectopia cordis.

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Patient consent

Obtained

Article source

Submitted

Peer review

External

Competing interests
None declared

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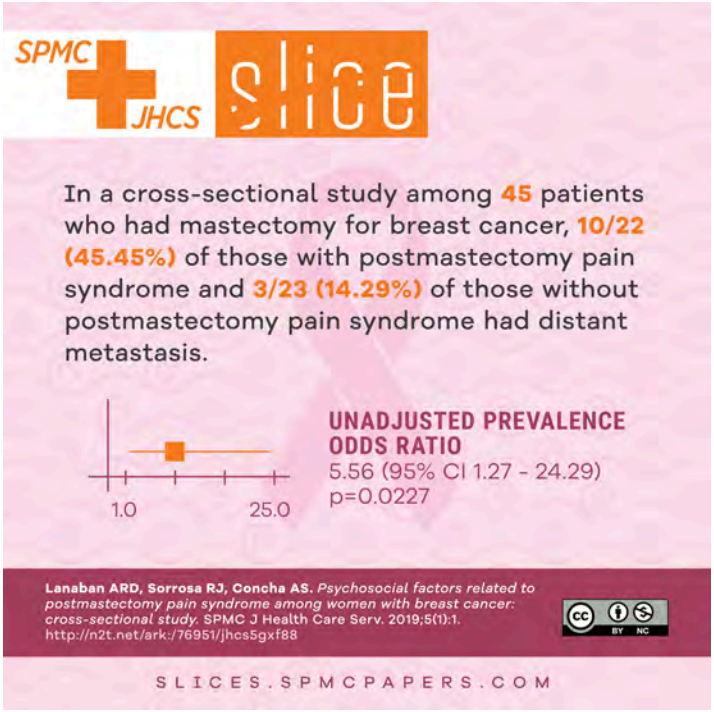

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