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Thoracic Wall Defect with Ectopia Cordis: A Case Report

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Abstract
Introduction: *Ectopia cordis is a rare congenital condition that is defined by the abnormal position of the heart outside the thorax associated with defects in the parietal pericardium, diaphragm, sternum, and, in most cases, cardiac malformations.*
Case Report: *A 28 year old primigravida delivered a female life baby with thoracic wall defect with pericardium and heart lying outside the thoracic cavity, in the labour room of Obstretic & Gynaecology Department in Regional Institute of Medical Sciences, Imphal. Mother did not come for ante natal checkup so the condition was not diagnosed prenatally. We report this case because of its rarity.*
Conclusion: *Surgical closure of the thoracic wall defects and proper management of other associated anomalies are the overall objectives in the management of ectopia cordis. Prenatal diagnosis can be done by using ultrasound as early as 9 weeks of gestation.*
Key words: *Ectopia cordis, thoracic cavity, thoracoabdominal, anomalies, Cantrell`s pentology.*

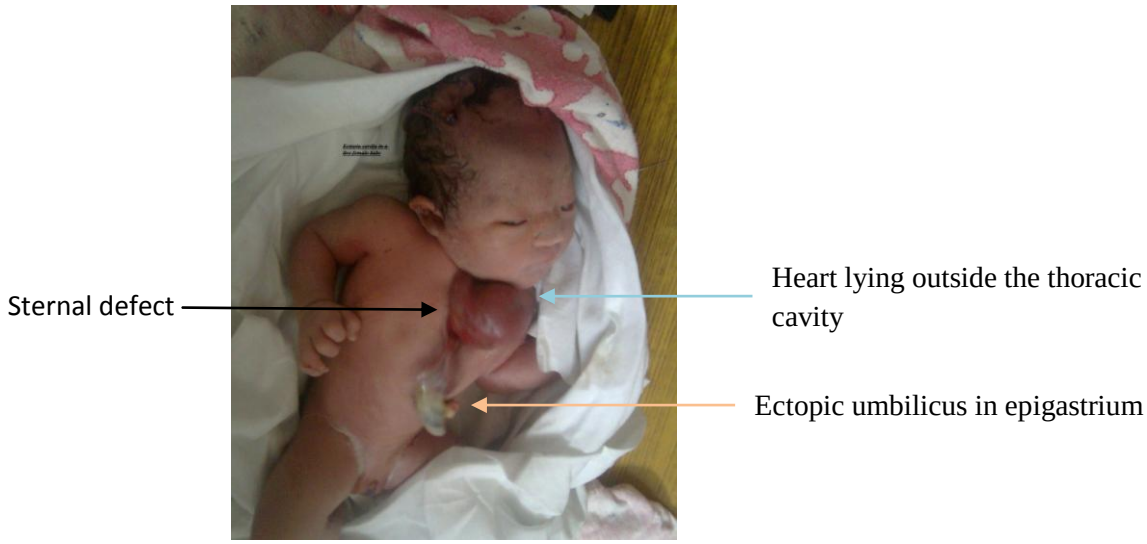
Introduction

Ectopia cordis is a rare congenital condition that is defined by the abnormal position of the heart outside the thorax associated with defects in the parietal pericardium, diaphragm, sternum, and in most cases, cardiac malformations. The reported prevalence is 5 to 8 per million live births.[1] The cause of ectopia cordis is currently unknown and most cases are sporadic. Byron classified ectopia cordis into four types: cervical, thoracic, thoracoabdominal and abdominal. The prognosis is poor and most infants are still born or die within the first few hours or days of life. [2] We present a case report because of its rarity.

A Case Report

A 28 year old primigravida delivered a female life baby with thoracic wall defect with heart lying outside the thoracic cavity, in the labour room of Obstretic & Gynaecology Department, Regional Institute of Medical Sciences, Imphal. Baby also shows an ectopic umbilicus in the epigastric region. Mother did not come for antenatal checkup so it was not diagnosed prenatally. Baby was immediately shifted to Neonatal intensive care unit for further management. Condition of the baby was explained to the parents and relatives of the baby. Unfortunately after 48 hours of delivery parents leave the hospital along with the baby

against medical advice. Baby expired at home after 60 hours of postnatal period probably because of respiratory distress and infection.



Discussion

Because of its rarity and other associated abnormalities ectopia cortis is a challenging congenital anomaly. Ravitch,[3] classified ectopia cordis into four types according to the location of heart, as thoracic (60%), abdominal (30%), thoracoabdominal (7%), and cervical (3%). We have reported a case of thoracic type of ectopia cordis with heart lying outside the thoracic cavity. Thoracoabdominal type is regarded as a distinct type as a Cantrell`s pentology, which includes five associated anomalies i.e. distal sternal defect, midline supraumbilical abdominal wall defect, ventral diaphragmatic hernia, defect in the apical pericardium with free communication with peritoneal cavity, and congenital intracardiac defect.[4] Ectopia cordis is frequently associated with other congenital defects involving multiple organ systems. Ventricular septal defects and Fallot's tetralogy are the most common associated intracardiac defects, while omphalocele is the most common associated abdominal wall defect.[5] The prenatal diagnosis of ectopia cordis is carried out using ultrasound, which allows visualization of the heart outside the thoracic cavity. Diagnosis has been reported by Bick et al[6] and Tongsong et al.[7] at 11 and 9 weeks of gestation, respectively. The overall objectives of ectopia cordis management are: closure of the chest wall defect, including the sternal defect, repair of the associated omphalocele, placement of the heart into the thorax, and repair of the intracardiac defects. [8,9] Unfortunately in most of the cases the thoracic cavity is small and mediastenum offer too little space for the heart. Attempts to close the chest wall after replacing the heart into the thoracic cavity often results in intolerable haemodynamic embarrassment secondary to kinking of the great vessels on compression of the heart muscles.[9,10] If the diagnosis of ectopia cordis is confirmed during pregnancy an early plan should be made for elective atraumatic caesarian delivery.[11] Immediately after birth the newborn should be stabilized and the lesion should be covered with saline-soaked gauze pads and wrapped to prevent desiccation and heat loss of the exposed visceras. [12]The prognosis of the ectopia cordis depends on its classification and associated anomalies. [10]Reports of successful repair of ectopia cordis are rare, cervical type is universally fatal, and the thoracoabdominal type has a higher rate of

successful repair but also has high mortality rate. [13]Etiology remains unknown but failure of closure of the ventral wall in the developing embryo is the leading explanatory hypothesis. [14]

Conclusion

We report this case for its rarity and 100% fatality without surgical intervention. Surgical closure of the thoracic wall defects and proper management of other associated anomalies are the overall objectives in the management of ectopia cordis. Prenatal diagnosis can be done by using ultrasound as early as 9 weeks of gestation.

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