

Case Report

Ectopia Cordis - A Rare Case

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ABSTRACT

Ectopia cordis is a rare congenital anomaly characterised by partial or complete displacement of the heart outside the thoracic cavity. It has a low survival rate, despite the various surgical techniques reported, because of the magnitude of the deformity and the associated intracardiac anomalies. With the development of the antenatal diagnosis and the tendency of parents to choose therapeutic abortion as a solution; this malformation is becoming rarer in the clinical practice. The present case was a 3 hour old male neonate weighing 3.2 kg, delivered by Cesarean section of a 20 year old primigravida in a tribal area, presenting with ectopia cordis.

After investigations, first stage surgery was performed but the neonate died within 24 hours.

Key Words: Ectopia Cordis, Turner's Syndrome, Trisomy 18.

INTRODUCTION

Haller first described the term "ectopia cordis" in 1706. The first attempted repair was done in 1925 by Cutler and Wilens; however it was Koop who achieved his first successful repair in two stages in 1975.

Christopher Wall (19 August 1975) holds the Guinness World Record for the oldest living person with this condition.

Ectopia Cordis, also known as Exocardia or Ectocardia, is a rare congenital anomaly having a prevalence of 5.5 – 7.9 per million live births.^[1] The term was derived from Greek word *ektopos* meaning away from a place. Ectopia cordis is defined as complete or partial displacement of the heart outside the thoracic cavity.

It occurs due to a defect in fusion of the anterior chest wall.

It is classified into five types: Cervical, Cervico-thoracic, Thoracic, Abdominal, and Thoraco-abdominal.^[2] The two most common forms of ectopia cordis are the thoracic and thoraco-abdominal type.^[3,4]

The latter is frequently associated with Cantrell's pentalogy, which include bifid sternum, deficiency of the diaphragm, defect of diaphragmatic pericardium, defect of the anterior abdominal wall, and intracardiac defects.^[5]

CASE HISTORY

A 3 hour old male neonate weighing 3.2 kg, delivered by Cesarean section of a 20 year old primigravida in a tribal area,

presented with a pulsatile mass projecting out of the thoracic cavity through a defect of 5 – 6 cm in the sternal area (Figure 1).

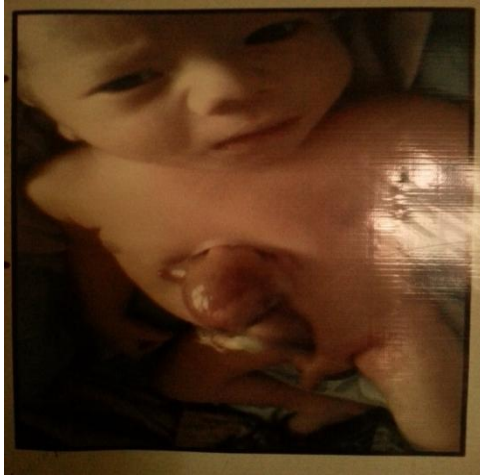


Figure 1: Ectopia cordis.

On Examination:

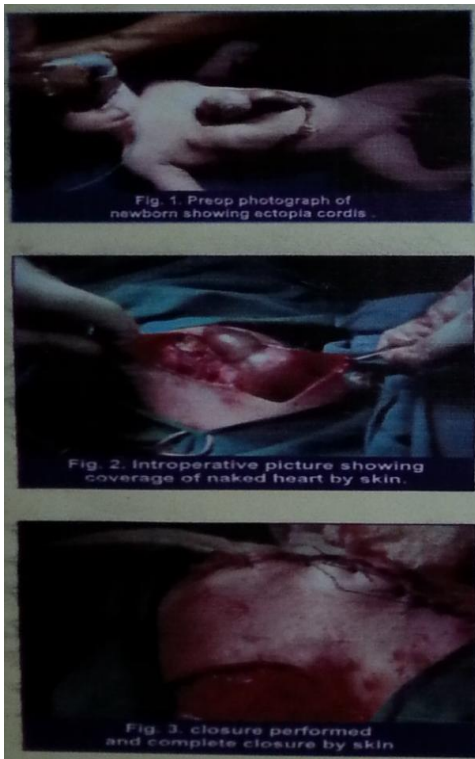


Figure 2: Intraoperative pictures.

Pathogenesis: Embrologically ectopia cordis is due to defect in the closure of lateral body wall folds to form ventral body

Heart rate = 120/min, BP = 60/48 mm Hg, RR = 40/min,

O₂ saturation = 99% with O₂ supplementation.

All pulses well felt, umbilical cord shifted cranially, peripheral cyanosis present, respiratory distress present.

X-ray chest:

Defect in the sternal area with irregular cardiac shadow.

Echocardiography could not be done because of unavailability of paediatric cardiologist.

Diagnosis of Ectopia Cordis was made.

It was decided to do first stage repair by covering the heart with skin (Figure 2).

Patient underwent operative procedure, but died within 24 hours.

wall. At the end of third week of embryo the lateral plate mesoderm splits into outer somatic and inner splanchnic layers. The space created between the layers of lateral plate mesoderm constitutes the primitive body cavity (Figure 3).

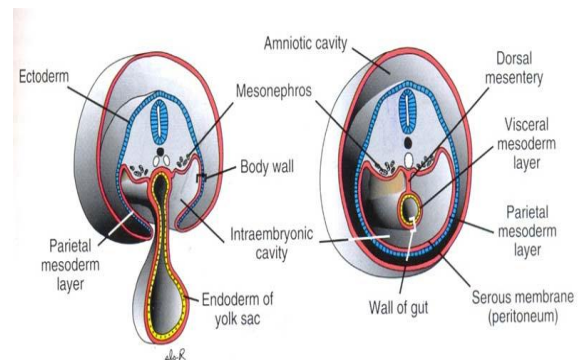


Figure 3: Lateral folds of embryo.

During 4th week, the sides of embryo begin to grow ventrally forming two lateral body wall folds. By the end of 4th week, the lateral body wall folds meet in the mid line and fuse to close the ventral body wall. This closure is aided by head and tail folds. The

developed heart reaches the original position by the end of 4th week. Complete or incomplete failure of midline fusion and defect in mesoderm fusion in embryonic state can result in a variety of disorders ranging from isolated ectopia cordis to complete ventral evisceration.

The other organs generally involved in ectopia cordis are:

1. CNS - Cranial cleft, Hydrocephalus, Cephalocele.
2. Face - Cleft lip and cleft palate.
3. Cardiac - AVSD, VSD, Tetralogy of Fallot, pulmonary stenosis, mitral atresia, tricuspid stenosis, coarctation of the aorta.
4. Pulmonary - Hypoplastic lung.
5. Skeletal – Scoliosis, limb hypoplasia, syndactyly, sternal cleft.
6. Abdominal - Divarication of recti muscle, Omphalocele, diaphragmatic hernia.

Ectopia cordis is frequently associated with Turner's syndrome and Trisomy 18. Carmi^[6] *et al* 1993 also supported that these ventral midline defects should be linked up by an X-linked mutation.

DISCUSSION

If the diagnosis of ectopia cordis is confirmed during the pregnancy, an early plan should be made for elective atraumatic cesarean delivery.^[7] Immediately after birth, the newborn should be stabilized and the lesion should be covered with saline-soaked gauze pads and wrapping to prevent desiccation and heat loss of the exposed viscera.

After completing the preoperative evaluation, the patient should be taken promptly to the operation room for surgical repair of the defects.

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 defect.^[8,9]

CONCLUSION

Prognosis of ectopia cordis is usually very poor. Review of literature reported only three survivors out of 29 cases.^[10]

Successful outcome requires a multidisciplinary approach.

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