# Thoracic Ectopia Cordis

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## Abstract

Ectopia Cordis is a rare congenital anomaly characterized by partial or complete displacement of the heart outside the thoracic cavity with an estimated incidence of 5 to 8 per million live births We report a case of a 22-year-old primigravida, unbooked and immunized woman, with no relevant family or personal history, in which the prenatal fetal ultrasound, performed at 34 weeks of gestation, revealed a defect of the anterior chest wall with exteriorization of the heart. Baby was Pre-term 34 week by date but term by modified Ballard scoring, male weighing 2.4 kg delivered by lower segment caesarean section. Baby had complete thoracic ectopia cordis with large defect in anterior wall of chest and sternum was absent.


**Key words:** Ectopia cordis, developmental anomaly, ventral body wall developmental defects

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## Introduction

Hallerfirst described the term Ectopia cordis in 1706 which is derived from Greek word *ektopos* meaning away from a place. Ectopia cordis is complete or partial displacement of the heart outside the thoracic cavity. It may occur as an isolated malformation or it may be associated with a large category of ventral wall defects that affect the thorax, abdomen or both. Kim[1] *et al*, in 1997 classified the Ectopia cordis into 5 types. Depending upon the position of the heart they are; Thoracic, Abdominal, Thoraco-abdominal, Cervical and Cervico-thoracic. Thoracic type is more common and cervical type is the rarest.

The thoraco-abdominl Ectopia cordis associated with sterna cleft, diaphragmatic hernia, omphalocele and an intracardiac defect is known as Cantrell's pentalogy. The prognosis is poor and most infants are still born or die within the first few hours or days of life.

## The Case

A Pre-term, 34 week of gestation by date but term by Ballard scoring, male baby was born to a 22 years primigravida mother by lower segment caesarean section at BP Koirala Institute of Health Sciences (BPKIHS), Dharan, Nepal.. Baby had a beating heart without pericardium in the sternal area of anterior thoracic wall. Sternum was absent and there was large defect of 5-3 square cm in anterior wall of chest. Umbilical cord was normal. At the time of birth APGAR at 1 minute, 5 minute and 10 minute was 8, 8 and 9

respectively, weight was 2.4 kg. Heart rate was 160/minute; respiratory rate was 54/minute: Spo2 was 92%. Cry was good and there was no organomegaly. Mother had undergone antenatal check-ups abroad in Dubai and at local doctor during the 3rd trimester. There was no history of consanguineous marriage, family history of congenital anomalies, history of intake of teratogens, or of exposure to radiation, mother was not an alcoholic. Two doses of tetanus toxoid vaccine were given. Antenatal ultrasonography had been done at 29th week of gestation but malformation was not detected. Repeat ultrasound examinations at 34th week of gestation by the same radiologist showed a normal growing foetus with a protruding heart outside the chest. So, she had come to this hospital for termination of pregnancy.

The baby was immediately attended by a paediatrician. The heart was covered with Vaseline soaked gauze immediately after birth and initial management was done and admitted to the nursery. The baby was then referred to Gangalal National Heart Centre, Kathmandu for surgery. Surgery was performed on 4th day of life but baby expired during surgery.

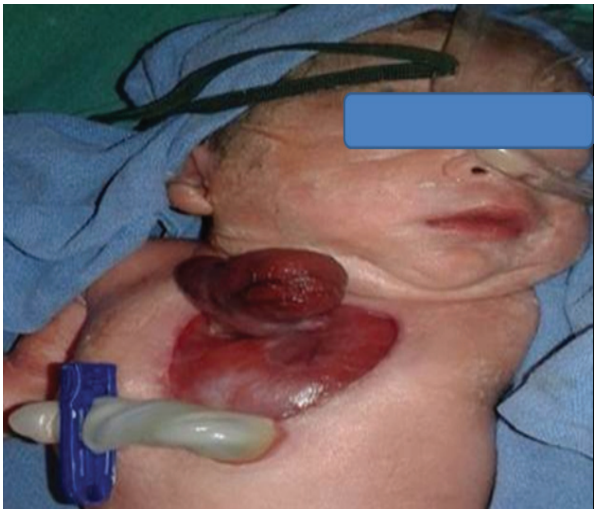

**Fig 1:** Showing the baby with ectopic heart.

## Discussion

The term ectopia cordis is defined as mal-position of heart, partially or completely outside the thorax. It is a rare congenital defect in the fusion of ventral chest wall resulting in extra thoracic location of the heart which is usually associated with other congenital anomalies and intra cardiac defects with an estimated incidence of 5 to 8 per million live births.[2] Depending upon the position of the heart ectopia cordis can be classified into five types:-

1. Cervical: In which the heart is located in the neck with sternum i.e. usually intact.
2. Thoraco-cervical: In which the heart is partially in the cervical region but the upper portion of the sternum is split.
3. Thoracic: in which the sternum is split or absent and heart lies partially or completely outside the thorax.
4. Thoraco-abdominal: which usually accompanies Cantrell's syndrome.
5. Abdominal: in which the heart passes through a defect in the diaphragm to enter the abdominal cavity. Kim *et al*[1], Dobell *et al*[3].

The pathogenesis of ectopia cordis and associated anomalies has been the subject of research. The prenatal diagnosis of ectopia cordis is carried out using ultrasound, which allows visualization of the heart outside the thoracic cavity. Earliest diagnosis has been reported by Bick *et al*[4]. and Tongsong *et al*[5]. at 11 and 9 weeks of gestation, respectively. Three-dimensional with Doppler allows for a more accurate early diagnosis. Magnetic resonance imaging (MRI) is also helpful in prenatal evaluation to plan for management of associated congenital anomalies.

While ectopia cordis is generally considered to be an isolated, there have been a number of reports linking it to chromosomal abnormalities. Reported karyotype abnormalities include trisomy 18, Turner syndrome and 46, XX, 17q+.7. Immediate surgical correction of ectopia cordis is often difficult, owing to the inability to enclose the ectopic heart within a hypoplastic thoracic cage. Although it has high mortality, corrective and palliative cardiovascular operations have been performed in different centres around the globe.

It is recommended that aggressive surgical procedures should be carried out without delay. Overall, the prognosis is poor. Antenatal management should include a careful search for associated anomalies, especially cardiac ones, and assessment of fetal karyotype. Early diagnosis by routine prenatal Ultrasonography as early as 10-12 weeks of pregnancy & termination of pregnancy prior to viability should be considered after discussion with parents.

Prognosis of ectopia cordis is generally poor and most cases results in still birth or die shortly after birth due to infection, cardiac failure, or hypoxemia because it is associated with intracardiac and other congenital anomalies. Surgical correction of Ectopia cordis is complex and generally requires a staged closure.

## Conclusion

This case emphasizes the need to improve the ante natal screening, existing referral system and raise the awareness of the health personnel on the proper diagnosis and prompt referral of rare and lethal cases like this.

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