Case Report

# Cardia Ectopia with Omphalocele Detected at 11+5 Weeks

**Prabha Sinha[1], Shabnum Sibtain[2]**

[1]Consultant Tawam Hospital, Abu Dhabi, UAE.
[2]Assistant Professor, Azra Naheed Medical College, Lahore

*Correspondence: Dr. Shabnum Sibtain*
*Asst Prof Azra Naheed Medical College, Lahore*
*Email: s_sibtain@hotmail.com*

## Abstract

Ectopia cordis is a rare, severe congenital anomaly.[1] This may be of various types depending on the extent of soft tissue involved. It is characterized by complete or partial displacement of the heart outside the thoracic cavity. The prenatal sonographic diagnosis of ectopia cordis is usually easy and can be diagnosed in the first trimester scan. Due to associated intracardiac and extracardiac anomalies outcome is very poor. This case of ectopia cordis with omphalocele, was diagnosed at 11 weeks +5 days and ended in termination of pregnancy. This defect might have started at 21 days of gestation, due to thoracic cavity compression by ruptured chorion/yolk sac.

Key Words: Ectopia cordis, omphalocele, pentalogy of Cantrell, thoraco-abdominal wall defect

Cite this article as: Sinha P, Sibtain S. Cardia Ectopia with Omphalocele Detected at 11+5 Weeks. J. Soc. Obstet. Gynaecol. Pak. 2018; Vol 8(2):135-136.

## Case Report

Forty three years old, G9 P7+1. All spontaneous pregnancies and spontaneous vaginal deliveries. Last delivery was 9 years ago. She is a second wife of a 67-year-old husband. He had three wives. Her LMP was 02/08/2017. She had a dating ultrasound with sonographer. The ultrasound suggested single intrauterine pregnancy, heart and liver seen outside the fetal abdomen, Nuchal translucency was 0.56 cm with positive fetal heart beats. she was referred to Fetal Medicine Unit on the same day.

Ultrasound in Fetal Medicine Unit suggested, crown rump length (CRL) 4.78cm equivalent to 11 weeks and 5 days with exomphalos measuring 1.0 cm x 0.82 cm with liver and heart seen outside the abdominal cavity. The diagnosis was made of Cardiac ectopia with omphalocele. The bad prognosis was explained to the couple. Both medical and conservative managements were offered. The parents decided to continue with the pregnancy. A week later intrauterine death was confirmed, and termination of pregnancy was carried out. She declined Chromosomal analysis.

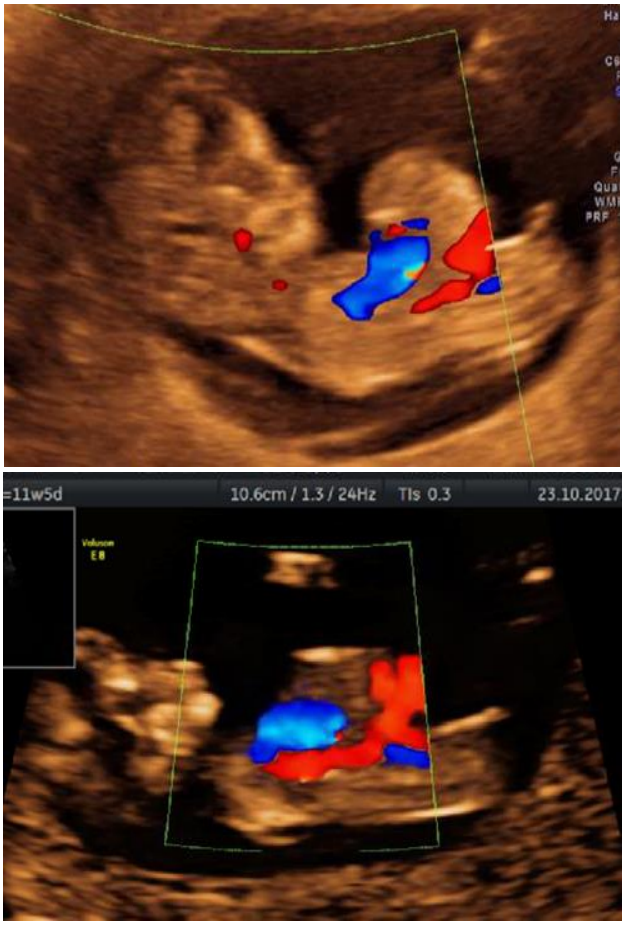


**Figure 1. Cardia Ectopia with Omphalocele**

## Discussion

Ectopia cordis is a rare congenital malformation, characterized by the heart completely displaced outside the chest. It is mostly on the thoracoabdominal side. Its

incidence is 1:100 000 live births in developed countries.[2] The thoraco-abdominal ectopia cordis can present anywhere in the world. It is often associated with Cantrell's pentalogy of anomalies.[3] Cantrell's pentalogy is an embryological anomaly. It was first described by Cantrell in 1958.[4] It has five classic midline deficiencies and it is often associated with ectopia cordis. The five classic anomalies are deficiency of the anterior diaphragm, a midline supraumbilical abdominal wall defect, a defect in the diaphragmatic pericardium, various congenital intracardiac abnormalities, and a defect of the lower sternum.[4] The pentalogy of Cantrell is an extremely rare congenital anomaly and is usually incompatible with life, with a male preponderance has been described in literature in developed countries.[5]

It is assumed that Ectopia cordis starts early in the embryonic period around the 8th and 9th week of pregnancy.[6] The prognosis of complete ectopia cordis depends on the degree of intracardiac and associated malformations and the degree to which the heart is exposed. If not treated it is fatal.[7] Congenital heart defects associated with Ectopia cordis may be due to the mechanical distortion of the developing heart following early rupture of the chorion and/or yolk sac.[8] The majority of neonates soon die after birth. Surgical correction is widely performed, with immediate covering of the heart and exposed abdominal contents using silastic prosthesis.[9]

## Conclusion

In conclusion, ectopia cordis with omphalocele is a rare congenital malformation and require to be adequately evaluated for appropriate prenatal and postnatal management. The prenatal diagnosis is made easily with ultrasound, as the heart is visualized outside the thoracic cavity. The diagnosis of ectopia cordis can be confidently made during the first-trimester. Once detected, the parents should be counselled about the poor prognosis. In view of the poor prognosis, termination of pregnancy can be considered if diagnosis is made before viability. As there is increasing availability of ultrasound, such cases can be diagnosed early in pregnancy. The ultrasonologists should be vigilant for such rare congenital anomalies. The increasing awareness of this rare condition among the obstetricians and ultrasonologists would result in making a diagnosis before viability and an option of termination of the pregnancy being offered.

## References


1. Baral K, Bandyopadhyay M, Das P, Dutta A, Mukherjee S. Ectopia cordis with omphalocele--a case report. J Indian Med Assoc. 2010;108(10):697-8.
2. Pannová J, Kováčová H, Matura D, Gruszka T, Pavlíček J, Delongová P. Ectopia cordis - case report. Ceska Gynekol. 2015; 80(3):214-7.
3. Chishugi JB, Franke TJ. Thoraco-abdominal ectopia cordis in southwest Cameroon. Pan Afr Med J. 2014; 18:124.
4. Morales JM, Patel SG, Duff JA, Villareal RL, Simpson JW. Ectopia cordis and other midline defects. Ann Thorac Surg. 2000; 70(1):111-4
5. Puvabanditsin S, Di Stefano V, Garrow E, Wong R, Eng J, Balbin J. Ectopia cordis. Hong Kong Med J. 2013; 19(5):447-50.
6. Taksande AM, Vilhekar KY A case report of ectopia cordis and omphalocele. Indian J Hum Genet. 2013; 19(4):491-3.
7. Alphonso N, Venugopal PS, Deshpande R, Anderson D. Complete thoracic ectopia cordis. Eur J Cardiothorac Surg. 2003; 23(3):426-8.
8. Ucima N, Tuka DD, Kimbongila MM, Lumbala PK, Biselele T, Tady BM, et al. Rare Occurrence of Ectopia Cordis in a Congolese Neonate. 2015; 56(2):132–133
9. Basant Kumar B, Sharma C, Sinha DD, Sumanlata Ectopia cordis associated with Cantrell's pentalogy. Ann Thorac Med. 2008;3(4):152-3. doi: 10.4103/1817-1737.43084