

Case Report of Pentalogy of Cantrell

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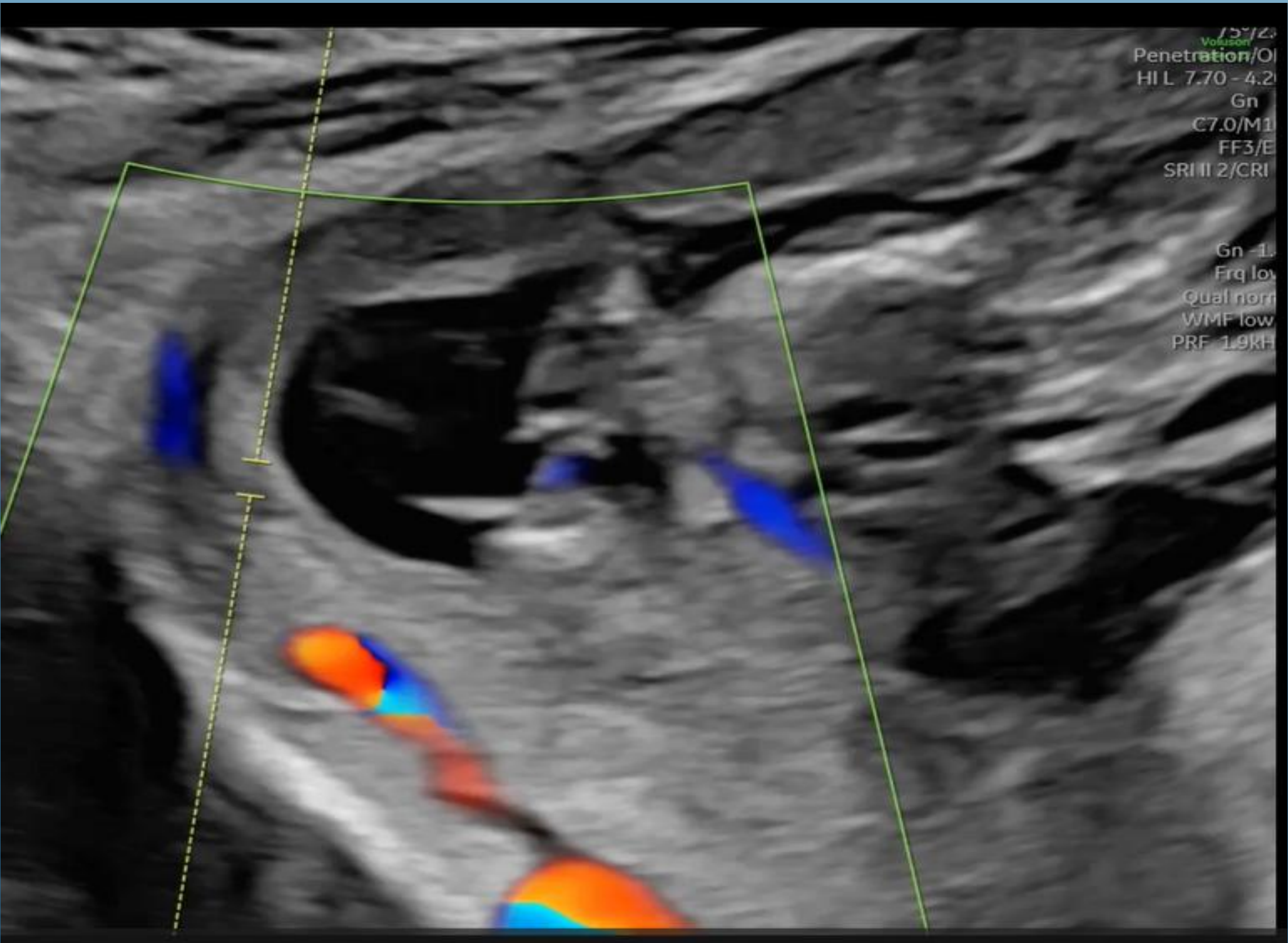
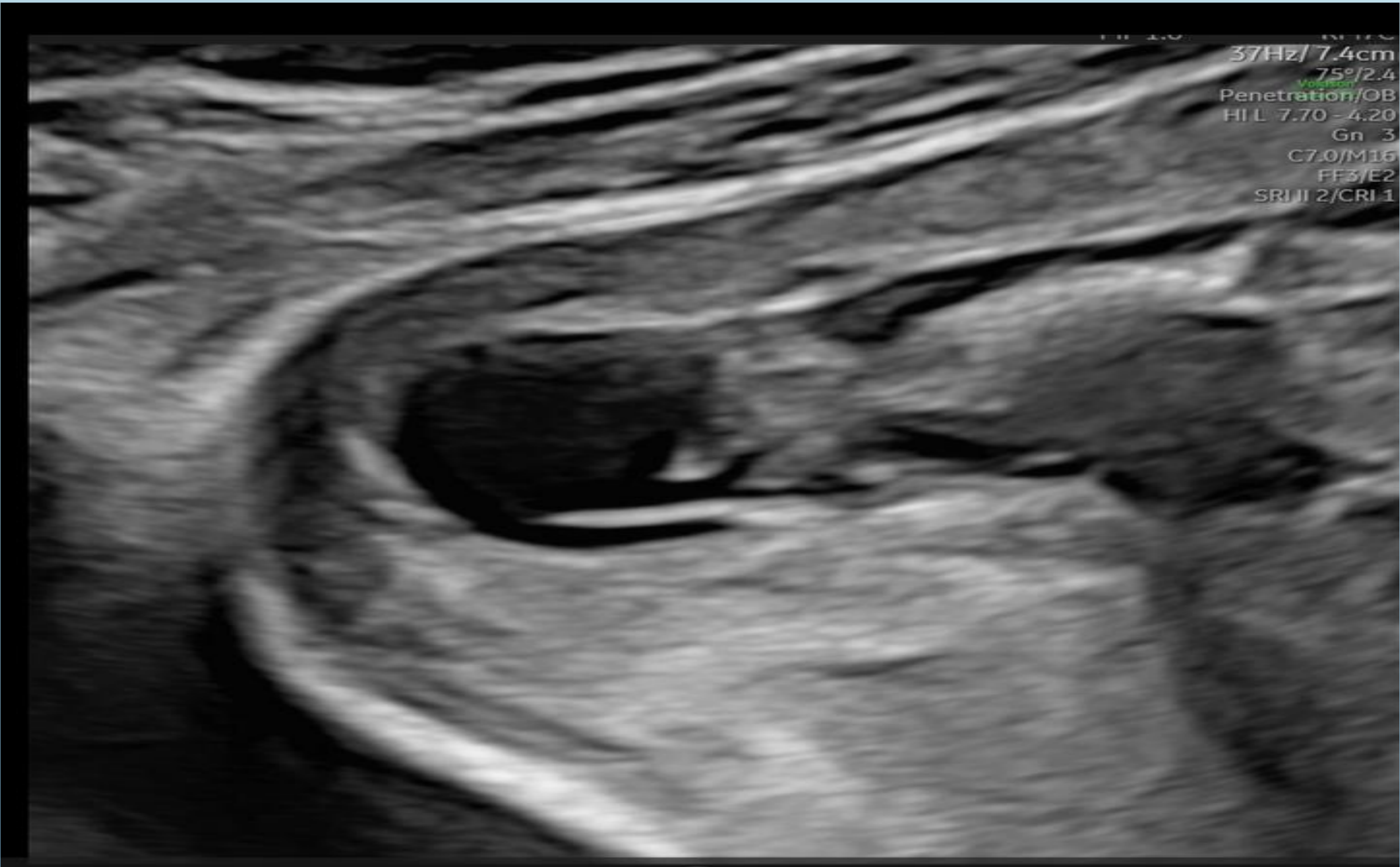
Introduction

Pentalogy of Cantrell is vey very rare congenital anomaly, its prevalence range from 1 in 5.5-1 million live births, it is associated with the multiple congenital anomalies specially involving Thorax and abdominal structures.

here we are presenting a case report on Pentalogy of Cantrell diagnosed in our hospital.

Case Report

28years old , Caucasian women, G1P0, BMI 25.8, blood group O positive, smoker, fit and well , had first trimester ultrasound at 12+5/40 weeks of gestation showed Suspicion of omphalocele containing Bowels and Stomach with possible Ectopia Cordis, Increased NT ,Mega cystis 7.9mm . Hence referred to Fetal medicine unit urgently. Scan in FMU showed Hydrops fetalis. Large Exomphalos containing Bowels, Liver and Stomach, Ectopia cordis: Large portion of the Heart is outside the baby’s chest, Bladder extrophy,the bladder is connected with the large exomphalos, The left leg was deformed. Scan findings explained to the couple and offered expectant management, invasive testing or Termination of pregnancy due to severe nature of congenital anomalies incompatible with life. Couple opted for termination of pregnancy and underwent medical Termination of Pregnancy and miscarried pregnancy at 13+3/40 weeks of gestation.



Discussion

Pentalogy of Cantrell is very rare congenital anomaly, It usually involves Thoracic and abdominal structures, its combination of five congenital abnormalities including Sternum, diaphragm, pericardium, abdominal wall and Heart which involves Ectopia Cordis almost all of the reported cases, omphalocele, Sternal Aplasia, sternal and diaphragmatic defects. The exact etiology of this congenital anomaly is unknown its is thought to be associated with sporadic or genetic mutation on x chromosomes or abnormality in the development of mesoderm during embryonic period. . Increased NT can be detected in POC and can be related to cardiac or chromosomal anomalies. usually its diagnosed in first trimester and foetus with confirmed POC is incompatible with life so once the diagnosis is established, the termination of the pregnancy is an appropriate decision.

Conclusion

We are reporting this case to increase the awareness of early detection and to assess foetus carefully in the first trimester of pregnancy for any major structural abnormalities that is not compatible with life, diagnosing the condition before viability allows the parents to consider the option of terminating the pregnancy and comparatively less traumatic psychologically.

References

- 1-Cantrell syndrome in the first trimester of pregnancy: imagistic findings and literature review
- 2-PENTALOGY OF CANTRELL: REPORT OF A UNIQUE PHENOTYPE .