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PRENATAL DIAGNOSIS OF TWO CASES OF PENTALOGY OF CANTRELL THAT SHOW THE BROAD ESPECTRUM OF THE SYNDROME.

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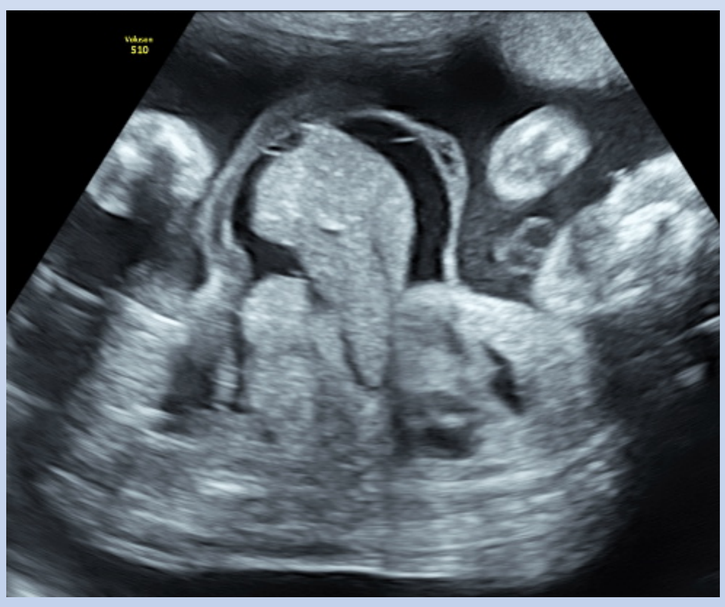
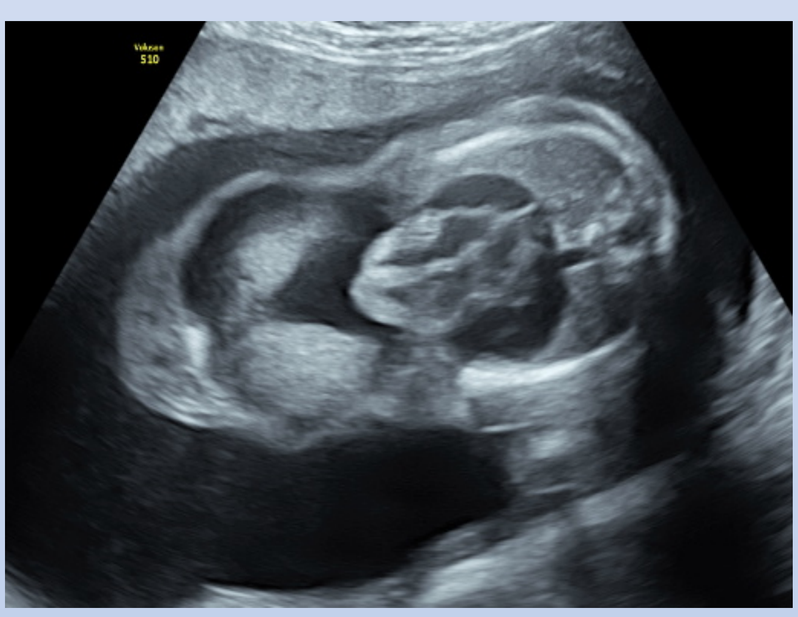
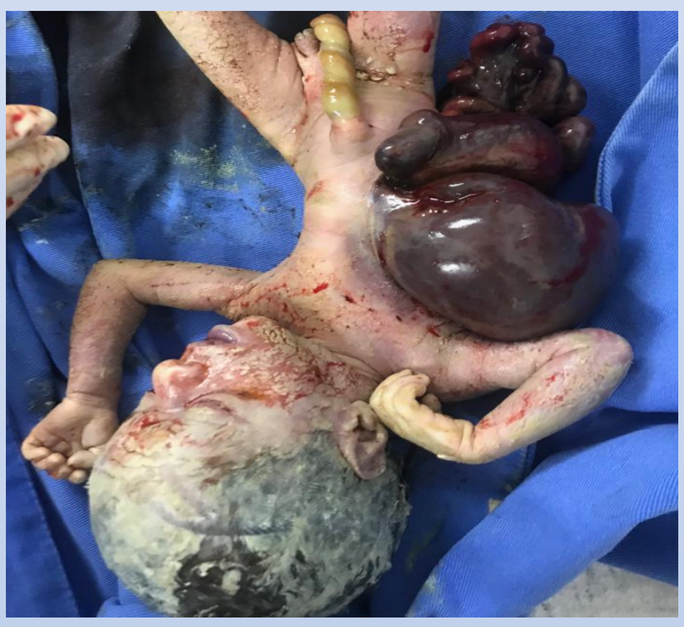
OBJETIVE:

Pentalogy of Cantrell is a non common syndrome with a reported prevalence from 1: 65 000 to 1: 100 000 cases (Desselle 2007). In 1972 Tayoma, proposed a classification: Type I: with the defects at five levels; (1) lower sternum, (2) anterior diaphragm, (3) diaphragmatic pericardium, (4) supraumbilical abdominal wall region, and (5) heart. Type II with only 4 defects but with cardiac and abdominal defects definitively present, and Type III with incomplete presentation, but with the a defect in the sternum. Our aim is to report two cases of live new-borns, diagnosed prenatally, which show the broad spectrum of the syndrome. One of the new-borns died minutes after birth, and the other survived.

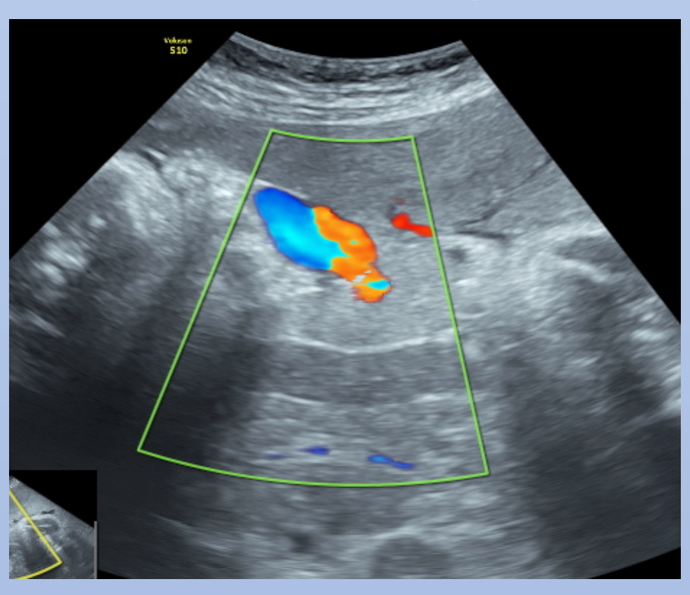


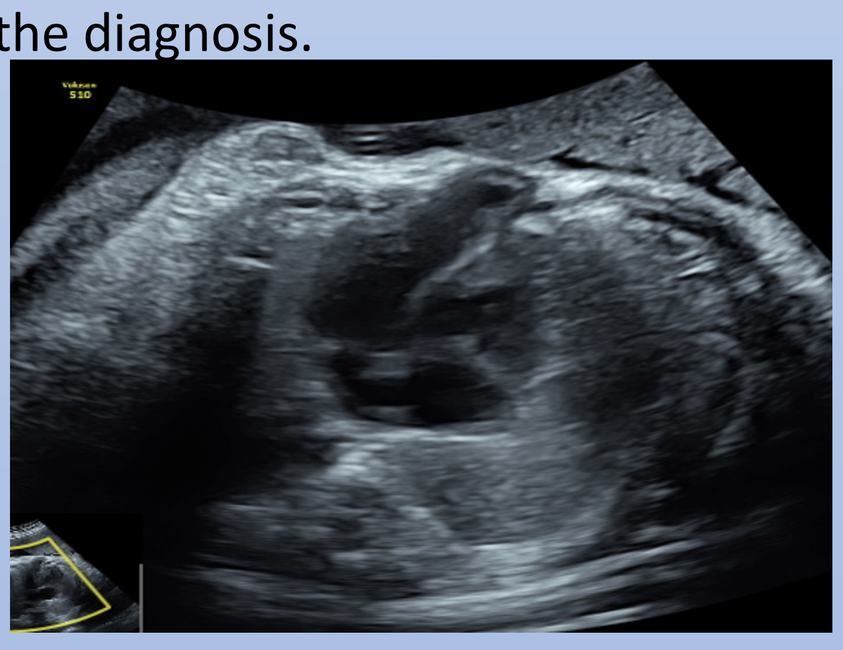


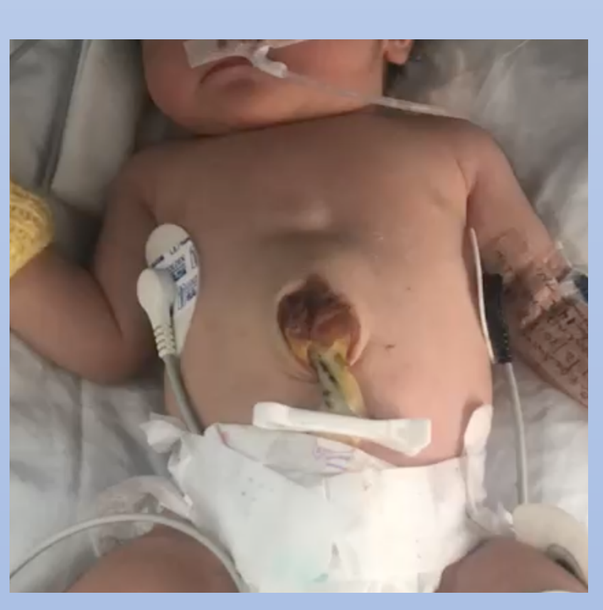


CASE 1

22 years old woman, G2A1, with no remarkable personal history, and no infection or genetic history. At 25 weeks of gestational age, was referred to our health centre, where the diagnosis of Pentalogy of Cantrell was made. She decided to continue the pregnancy. Penatal cariotype: 46XY normal. At 32 weeks of gestation, preterm labour began, a boy of 1690 gr was born, apgar 2-1-1, and died 10 min after birth. Pathology confirmed the diagnosis.







CASE 2

32 yeasr old woman, G2P1CO, with no remarkable personal history, and no infection or genetic history. No prenatal care. At 37 weeks of gestational age, omphalocele, partial agenesis of sternum, and aneurysm of the apex of the left ventricle, was documented. At 38 weeks, labour started, and a vaginal delivery of a boy of 2790 g occurred. The baby had partial agenesis of the sternum, small omphalocele in the heart, a ventricular aneurysm, and small defect of the diaphragm. At 12 days of life, a surgical correction of the pericadium, diaphragm and abdominal Wall defects was made. At 24 days of life, the new-born was released from the hospital with no complications. Postnatal karyotype: 46xy normal .

CONCLUSION

Pentalogy of Cantrell has a broad spectrum of occurrence; on one hand with severe defects with very poor prognosis, and on the other hand, an incomplete occurrence with better prognosis. This is why it is so important to do a very accurate prenatal diagnosis, so proper council to the parents can be given.