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Pentalogy of Cantrell. An eighteen case series

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Objective

The purpose of this study its to describe the prenatal diagnosis and perinatal outcome in 18 cases diagnosed with Pentalogy of Cantrell (POC), in our centers.

Methods

Retrospective study in which all cases diagnosed prenataly with Pentalogy of Cantrell were collected in three tertiary centers. We reviewed the files of all POC cases between the years 2003 and 2018. The criteria were thorax - abdominal wall defect, presence of heart malformation, aneuploidy association and perinatal outcome. Continuous variables with abnormal distribution data were described as a mean and range. Categorical variables are shown as absolute value and proportion.

Results

Eighteen cases of POC were diagnosed during the study period. Maternal age was 27 years (15 to 45 years), Gestational age of diagnosis was 20 weeks (11 to 31 weeks). Ultrasound findings were: omphalocele in 18/18 patients, diaphragmatic defect 18/18, pericardial defect 17/18, sternal defect 17/18, cardiac malformation 13/18. Chromosomal analysis was conducted in 10/18 cases, 9 were normal and 1 was abnormal, which came back as trisomy 18. 7/18 patients were stillborn at 23 weeks (12 to 31 weeks) and 8/18 died in the first 6 hours of life. 2/17 died at 21 days of life, and only one was alive at one year of birth.

Conclusion

According our knowledge this is the largest published series of prenatal diagnosis of Pentalogy of Cantrell. Pentalogy of Cantrell is a rare multiple malformation syndrome, as a results of abnormal embryo development, consisting in a supra umbilical abdominal wall defect, pericardial defect, anterior diaphragmatic defect, pericardial defect and or heart malformation. It is more likely to be associated with with normal kartotype. Perinatal outcome is poor with less than 10% of perinatal survival.