Fetal ventricular aneurysms and diverticula: Nine new cases and a review of the current literature

Jones C.B., Simpson J.M., Miller O.I., Sharland G.K.
Department of Fetal and Paediatric Cardiology, Evelina Children's Hospital, London, UK.

Introduction: Congenital ventricular aneurysms (CVA) and diverticulum (CVD) are rare conditions with a variable prognosis when diagnosed in fetal life. There have been isolated case reports but little evidence to predict survival or outcome. Here we present a large fetal case series and review a total of eighty-two cases to ascertain prognostic factors that may aid with prenatal counselling.

Methods: Retrospective search of our fetal cardiology database between 1991 and 2011 for cases of CVA or CVD. Patient notes, fetal echocardiograms and relevant postnatal investigations were reviewed. A literature search identified 73 previously published cases.

Results: Two fetuses had CVD, one arising from the left ventricle (LV) and one from the right ventricle (RV). There were seven cases of CVA with 5 arising from the LV and 2 from the RV. Two cases of CVA had poor ventricular function and parents opted for termination of pregnancy (TOP). One patient with a rapidly expanding LV aneurysm suffered an intrauterine death (IUD). All six live-born infants remain well on follow-up (range 3 months – 18 years). Reviewing all published cases, there were 57 survivors, 10 TOP, 11 IUD and 4 neonatal deaths. Fetal hydrops, ventricular dysfunction, increasing size and cardiomegaly were all independently associated with non-survival (p<0.05). These poor prognostic signs were more frequent in those with a LV aneurysm. Eleven fetuses had associated structural heart defects (predominantly septal defects), overall this did not affect outcome. Presentation in later gestation and those with only fetal dysrhythmia tended to have a favourable outcome (p<0.05). Pericardial effusion was present in 33 cases and associated with a poor outcome in the LV group (p<0.05). Prenatal intervention with pericardiocentesis was performed most frequently in those with RV diverticulum who had a favourable outcome with or without intervention.

Conclusion: CVA and CVD are a heterogeneous group of conditions where prognosis depends on location and type of lesion. Our review demonstrates that detailed fetal cardiology assessment to look for associated sequelae can further help to predict prognosis and guide prenatal counselling. Intervention should be discussed on a case by case basis. The aetiology of these lesions remains uncertain.