ASSOCIATED COMPLEX CONGENITAL HEART DISEASE BEST PREDICTS REPAIR AND OUTCOME IN PRENATALLY DIAGNOSED ATRIOVENTRICULAR SEPTAL DEFECTS

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Authors: Andrea Z. Beaton, Jodi Pike, Mary Donofrio, Children’s National Medical Center, Washington, DC, USA

Background: Atrioventricular septal defects (AVSD) are a common prenatal diagnosis that encompasses a wide spectrum of abnormalities. When associated with complex disease, little information exists to predict postnatal outcome.

Methods: This is a retrospective review of fetuses diagnosed with AVSD. The mid-gestation echocardiogram and postnatal medical record were reviewed.

Results: From 2001 to 2011, 106 fetuses with AVSD were identified; 66 (61%) were balanced with mean LV:RV ratio 0.85 to 1.15, and 40 (39%) were unbalanced. Of the balanced defects, 15 were terminated and 9 were lost to follow-up. Of the remaining 42, 37 had successful biventricular (2V) repair; 25 with no associated cardiac disease, 9 with transitional AVSD, and 3 with tetralogy of Fallot (TOF). Five, all with additional complex disease, had univentricular (1V) repair, with one death. Concurrent complex disease, heterotaxy (3/37, 8%, p<0.01) and/or double outlet right ventricle (3/37, 8%, p<0.01), was associated with 1V repair. Of the unbalanced defects, 10 (25%) were left-dominant with mean LV:RV ratio of 1.9 +/- 0.61, and 30 (75%) were right-dominant with mean LV:RV ratio of 0.64 +/- 0.16. In the left-dominant group there were 3 terminations, 1 intrauterine death, and 2 lost to follow-up. One child died preoperatively and the remaining 3 underwent 1V repair, with one death. In the right-dominant group there were 12 terminations, 1 intrauterine death, 4 lost to follow-up, and 2 preoperative deaths. Of the surviving 11, there were 3 1V repairs, with 2 deaths (66%), and 8 2V repairs, with 2 deaths (25%). A restrictive ventricular septum (5/8, 63%), LV:RV ratio> 0.55 (8/8, 100%), and the absence of concurrent complex disease (6/8, 75%) was associated with a 2V repair.

Conclusions: When AVSD is diagnosed prenatally, a thorough search for additional cardiac anomalies should be performed. The presence of concomitant complex heart disease is associated with 1V repair whereas a restrictive ventricular septal defect is associated with a 2V repair. Need for 1V repair is associated with a poor prognosis. In fetuses with AVSD, detailed and accurate prenatal imaging is imperative for effective parental counseling and postnatal planning.