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Prenatal Diagnosis of Total Anomalous Pulmonary Vein Connection: Our Institutional Experience.

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Introduction: Prenatal diagnosis of total (TAPVC) anomalous pulmonary venous connection in isolation or associated with other cardiac disease is rare and may reflect the difficulty in detecting them in utero.

Objective: To describe and evaluate the spectrum of fetal cardiac anomalous pulmonary venous connections and associated cardiac defects, at our tertiary referral center.

Methods: Retrospective review of 1934 fetal echocardiograms, performed between 2010-2011. All pregnancies with a fetal diagnosis of TAPVC, prenatal and postnatal echocardiograms and medical records were reviewed.

Results: We identified 8 fetuses with prenatal diagnosis of TAPVC. Mean gestational age at diagnosis was 29 weeks. 3 supracardiac, 3 intracardiac(coronary sinus) and 2 infracardiac type. Seven had an additional major cardiac defect, including 3 atrio-ventricular channel defects with associated right heterotaxy syndrome, one congenital corrected transposition of large vessels, one hypoplastic left ventricle, one double outlet right ventricle and one interventricular septal defect. Visualization of a dilated superior cava vein(2/3) and a vertical vein(1/3) in the supracardiac type, a dilated coronary sinus(3/3) in the intracardiac type and a vertical vein draining into the portal sinus(2/2), were the most consistent echocardiographic clues. In five cases, prenatal diagnosis was only made at follow-up assessment. 2 had altered karyotype, one prenatal diagnosis of Cat-Eye-Syndrome and one postnatal diagnosis of Edwards-Syndrome (parents refused to undergo amniocentesis). Diagnosis was confirmed postnatally in 7/8, one suspected supracardiac TAPVC had left pulmonary drainage obstruction confirmed by MRI. In five fetuses with TAPVC and obstruction, confirmed postnatally, continuous turbulent flow in the drainage site were demonstrated by Color-Doppler. Complete TAPVC surgical correction was performed in 7/8 with an average life of ten days. Premature death(<2months) occurred in 3 cases(2 Heterotaxy syndromes and Edwards-Syndrome).

Conclusion: Detailed assessment of the PV connection, both at initial examination and in serial fetal studies, is important for the prenatal diagnosis of TAPVC. The high morbidity and mortality were largely due to the presence of severe additional pathologies. In these patients fetal diagnosis did not significantly improve the prognosis, although it was critical for parental counseling and perinatal management.