What to do for a fetus of 24 weeks of GP with severe absent PV syndrome

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Absent pulmonary valve syndrome (APVS) is a rare congenital heart defect. It is usually associated with tetralogy of Fallot (TOF, TOF/APVS), but APVS without intracardiac anomaly is also found. Although it is morphologically characterized by severely dilated branch pulmonary arteries, the right ventricle and tricuspid valve are typically of normal size. Because dilated peripheral pulmonary arteries compress the small airways, the potential for respiratory failure at birth is high. With the frequent use of fetal echocardiography, most of the patients with APVS are diagnosed prenatally, which leads to a high rate of termination in this subset. In the previous reports, termination rates range from 30% to 40%, and the 1-year mortality rates range from 67% to 75%. The perinatal risk factors for mortality include (1) respiratory distress at birth, (2) presence of a genetic syndrome or abnormal karyotype, and (3) presence of hydrops fetalis. More recent series of TOF/APV have reported better rates of survival, ranging from 72% to 86%. This may be due to better anticipatory planning at birth after prenatal diagnosis and to improved surgical strategies for repair. In patients with APVS without TOF (i.e. isolated APVS or APVS in a functionally single ventricle), there is a normal tricuspid annulus and a normal or dilated right ventricle. Alternatively, it can occur with single-ventricle physiology with underlying tricuspid atresia or stenosis and right ventricular hypoplasia. In most fetal case series of APV, subjects without underlying TOF constitute only 10% to 25% of the study cohort, with relatively high termination rates. We sought to

investigate the current outcomes in fetuses diagnosed as having APVS in Asan Medical Center, in association with the review of the surgical cases.