Early clinical outcomes for fetuses with severe or greater tricuspid regurgitation in the era of cone reconstruction


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Objectives: Since cone tricuspid valve (TV) reconstruction was introduced, the treatment strategy for fetuses with severe or greater tricuspid regurgitation (TR) has been dramatically changed. However, surgical indications and criteria for biventricular repair (BVR) still remain unclear.

Methods: During 2013, 5 fetuses were diagnosed with severe or greater TR and functional (n=2) or membranous (n=3) pulmonary atresia related with Ebstein anomaly (n=3) or congenital dysplastic TV (n=2). The median cardiothoracic area ratio was 0.69 (range, 0.64-0.70). Three babies were prematurely born due to fetal hydrops (n=2) or fetal growth restriction (n=1) and the median birth weight was 2982g (1712-3240). One baby was diagnosed with Down syndrome.

Results: Soon after birth, mechanical ventilator support was initiated in 4 patients and peritoneal dialysis was performed to attenuate whole body edema in 2 patients. The median cardiothoracic ratio (CTR) and TR pressure gradient (TRPG) was 91.0% (84.3-91.5) and 27.0mmHg (17.4-49.0). One patient with fetal hydrops died at 4 days of age without surgical intervention due to low output syndrome. The remaining 4 patients underwent complete BVR consisting of cone reconstruction, the creation of right ventricle to pulmonary artery continuity, and closure of atrial communication at the median age of 10.5 days (8-15). Right atrial cryoablation was concomitantly performed in 3 patients with ectopic atrial tachycardia. TR decreased to a moderate severity level in all patients. There was 1 in-hospital mortality on post-operative day 41 due to persistent pulmonary hypertension related to chromosomal anomaly, and 1 late mortality on day 56 due to septic shock caused by necrotizing enterocolitis The latter patient underwent the Starnes operation conversion as a single ventricular palliation 8 days after BVR because the right ventricle did not compensate. Two patients successfully survived at discharge and the latest follow-up confirmed that the TR grade remained moderate and the CTR was less than 70%.

Conclusions: With cone reconstruction, 2 of 5 critically ill patients successfully obtained complete biventricular circulation. TRPG of more than 40mmHg without primary pulmonary arterial hypertension seems to be essential for BVR.