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When Coronary Arteries Need Right Ventricular Systolic Pressure

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Introduction Patients with pulmonary atresia and intact ventricular septum (PA IVS) can have coronary sinusoids connected to a hypertensive RV. Coronary perfusion can be dependent on the RV systolic pressure wave (RVDCC), even when dual supply exists. Decompression of RV then can be deleterious. We evaluated the outcome of different treatment strategies.

Patients and methods: national, multicentre (4), restrospective analysis. Of 207 patients (pts) born 1985-2010 with PA IVS, 41 had coronary sinusoids. All angiograms, cardiac ultrasounds, surgical reports, pathology reports and outcome data were reviewed.

Results 13 patients had normal coronary flow, 17 had dual flow of which 7 had significant systolic RV flow. 11 patients had segments with only sinusoidal perfusion. Thirty five pts (85%) received a BT-shunt at 11 days (2d–16m); 22pts (53%) had a Glenn shunt at 8 months (2m–5y). Twelve pts (29%) went to full Fontan at 3y (2.5–19y).
Eleven pts (27%) had fulguration and balloon dilation of pulmonary valve, 3 with RVDCC; 2pts (5%) RVOT surgery. Only one patient with RV decompression (dual flow) died at the age of 7 months short after bidirectional Glenn due to collaps of the right coronary artery; in retrospect RVDCC. In 3pts (7%) with dual flow, decompression resulted in disappearance of sinusoids.
Eighteen patients (43%) had RVDCC. Twelve patients died (29%), 8 (19%) with RVDCC at age 93.5 days (9d – 2.5y): 7 at BT-shunt, 2 at bidirectional Glenn; all suffered massive ischemia. 3 patients died >1y suddenly (no pathology). Pathology in 4 showed marked coronary hypertensive vasculopathy, intimal fibrosis and medial hypertrophy.

Conclusion Decompression of the RV appears to be safe if dual flow without RVDCC; frequently regression of sinusoidal flow was observed. If RVDCC, reduction of coronary perfusion to “normal aortic diastolic flow” may result in acute ischemia and infarction. Cardioplegia (administration and washout) is different in RVDCC and should be avoided. Natural history of RVDCC is dreadfull with progressive endothelial proliferation resulting in late ventricular ischemia. Heart transplantation is probably the best long term option in RVDCC.