

Treatment of PAIVS: what is the best option?

*Carrozza M., Santoro G. (1), Gaio G. (1), Palladino M. T. (1), Del Gaizo F. (1), Capogrosso C. (1), Cirillo A. (1), Molinari D. (1), Giordano M.(1), Farina G. (1), Caianiello G. (1), Russo M. G. (1)
Pediatric Cardiology, Second University of Naples, Monaldi Hospital, Italy*

Background. Innovations in knowledge and techniques have improved survival of pts with PAIVS. Despite this, it is still difficult to define the best treatment for those pts which are “within the spectrum”. Methods: The outcome of 24 newborns with PAIVS (prenatal diagnosis n = 13, postnatal n = 11) was reviewed. Results. The mean age and weight at the observation were 2.5 + 2.4 days and 3.3 + 0.8 kg. The mean Z-Score value of the TV annulus was -3.01 + 1.36. We detected a severe hypoplastic RV (6/24, 25.0%), bipartite RV (8/24, 33.3%) or a hypertrophic tripartite RV in 10/24 (41.6%) pts. Two associated lesions: Ebstein anomaly in one and mitral valve anomaly in another. In one pt we found RV-dependent coronary circulation. Catheter RF was performed in 18/24 pts (72.0%) with favourable anatomy (mean TV Z-score -2.7). Pts with unfavourable anatomy, 1st intervention was MBT shunt in 3 pts (3/24, 12.0%), and PDA stenting in 3 pts (3/24, 12.0%). Perforation was successful in 16 patients (16/18, 88.8%, 1 pt submitted Brock valvotomy and in 1 with pulmonary artery perforation and cardiac tamponade needing neonatal surgery). No deaths. During the first month all pts submitted to RF needed further pulmonary flow. In 13/18 (72.2%) patients ductal stenting and 5/18 pts (27.7%) required a MBTs placement. At a median FU of 6.2 years (range 1.2 to 13.1), among 18 pts underwent RF procedure, 5/18 (27.7%) have a univentricular connection and 13/18 (72.2%) have a biventricular pathway. At FU, in our 24 APSI pts, 9/24 (37.5%) pts have a univentricular connection (3 pts with partial cavo-pulmonary anastomosis and 6 pts Fontan completion), 15 (62.5%) pts got a biventricular correction (2 coil embolization of MBTs with no further intervention, 5 needed RVOT surgery, 3 pts still have PDA flow, and 5 no procedure after MBT or stent spontaneous closure). Conclusion: detection of RV anatomical type and the related-Z Score value of the TV is a predictive index for surgical or percutaneous intervention. Catheter RF is feasible, with low mortality, even if all pts need a pulmonary blood flow after the procedure.