Hybrid Approach for Infants with Pulmonary Atresia, Ventricular Septal Defect, extreme Hypoplasia of Central Pulmonary Arteries and Major Aortopulmonary Collaterals.

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Objectives: to report our early experience with treatment of pulmonary atresia with ventricular septal defect (PAVSD), severe hypoplastic central pulmonary arteries (PAs) and major aortopulmonary collaterals (MAPCAs), by a hybrid approach.

Background: PAVSD and MAPCAs is a complex congenital heart defect that can vary according both to PAs anatomy and source of pulmonary blood flow. Management of this lesion is still controversial. Early establishment of a forward flow through the native pulmonary arteries could promote their growth; however results from both surgical and percutaneous approaches are often disappointing when required in small infants.

Methods: the department registry was retrospectively searched for all patients affected by PAVSD, MAPCAs and severe hypoplastic PAs (< 2 mm) and treated with a hybrid approach between February 2007 and March 2012.

Results: a perventricular implantation of a right ventricle (RV) to PA stent was attempted in five hypoxemic infants (mean age, weight and saturation were 65 days, 3.5 Kg and 60%, respectively). The procedure was performed under general anesthesia, throughout a midline sternotomy. Under direct vision a 22 Gauge needle was advanced towards the free wall of the RV into the pulmonary trunk. After angiography, a 0.014 inch floppy guide wire was inserted into a pulmonary branch and a 3Fr sheath placed over that wire. A Taxus Liberté 3.5 mm x 19 mm stent was implanted. The procedure was successful and final angiography showed a well-placed stent in all cases. Mean total procedural time was 175 minutes with a mean x-ray time of 8 minutes. The post-operative course was uneventful in all infants but one in whom we had an early major complication. At a mean follow-up of 43 months all patients are alive. Four of them had a new opened RV to pulmonary artery conduit (VSD left opened), with increased PAs size and an oxygen saturation above 82%. Only one patient was considered unsuitable for other surgical procedure, having showed complete stent occlusion at CT scan control, without any evidence of central PAs.

Conclusions: hybrid approach allows to obtain growth of native PAs even in small infants with the most unfavorable PAs anatomy.