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Outcome after surgery for pulmonary atresia and VSD; a long-term follow study in a single institution in Sweden

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Objective. To examine the outcome after treatment of pulmonary atresia and VSD.

Methods. All children in our referral area (50% coverage of the Sweden population) operated upon due to pulmonary atresia and VSD from Jan 1st 1994 to Dec 31st 2017 were included. Patient files were analyzed and cross-checked against the Swedish National Population Registry as of Jan 1st 2018 allowing for reliable and complete data on survival. Primary outcome was all cause mortality, secondary outcomes were incidence of extracardiac diseases, reoperations and catheter interventions.
Results. Seventy patients were identified (31 girls, 39 boys) with a median age of one day (0-480) at presentation. All medical files were retrieved with no patient lost to follow-up. The pulmonary arteries were confluent in 61 (87.1%) patients, in whom a ductal supply was diagnosed in 44 (62.8%) and no ductal supply was found in 17 (24.2%). The pulmonary arteries were non-confluent in five patients (7.1%), and in four (5.7%) no central arteries were found. A right aortic arch was seen in 14 patients (20%), 22q11 deletion syndrome in 16 (22.8%) and other significant syndromes in a further nine patients (12.8%). Major aorto-pulmonary collateral arteries (MAPCAs) were found in 29 patients (41.4%), in whom 12 a unifocalization procedure was performed. Corrective surgery was accomplished in 58 patients (82.8%) at a median age of 1.3 years (0.01-19). Eight patients received a Melody® valve. Death occurred in 18 patients (25.7%), at a median age of 1.3 years (0.02-19), of whom three died within and 15 later than 30 days after the last major surgery. Mortality was similar in patients with and without MAPCAs (27.6% vs 24.4%). All deaths were cardiac except one, where the child died in an accident. The median follow-up time in survivors was 13 years (0.8-39.4) after birth.

Conclusion. Corrective surgery was accomplished in 82.8% of all patients, and long-term survival was 74.3%, with no difference found in patients with or without MAPCAs. The incidence of associated syndromes was high, with 22q11 syndrome in 22.8% and other significant syndromes in another 12.8%.