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**Pulmonary atresia and intact ventricular septum (PA-IVS) – a population based follow-up in Sweden between 1980 and 2016**

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**Introduction.** Pulmonary atresia and intact ventricular septum is a rare and heterogeneous congenital heart defect that still represent a challenge to pediatric cardiac surgeons and cardiologists.

**Objective.** To study the outcome of all children born with pulmonary atresia and intact ventricular septum in Sweden between January 1980 and August 2016.

**Methods.** A retrospective, descriptive study based on medical reports and the initial echocardiographic examination.

**Results.** The cohort consists of 171 subjects, giving an incidence of 4.6 per 100 000 live births. 74/171 (43%) had ventriculo coronary arterial communications (VCAC). 164 subject were offered treatment, for 147 subjects the initial treatment was based on surgery and for 17 it was based on percutaneous treatment. At the end of the study period 130 subjects were alive, 71 had biventricular repair, five one- and an half ventricular repair and 55 univentricular palliation with completed total cavopulmonary connection in 42 cases. Follow-up time ranged from 2.5 months to 35 years (median 14 years). The overall mortality was 37 % (34/91) between 1980 and 1999, and 8 % (7/80) between 2000 and 2016. Of the 41 that died, 34 (83%) died before the age of one year, no death due to the heart defect after the age of nine years. In 26 subjects the heart defect was diagnosed prenatally, of whom 10 ended up with a biventricular circulation. Statistical analysis of incremental risk factors for death showed statistical significance for low birth weight, having systemic-to-pulmonary shunt as the only surgical procedure. The survival after surgery is statistically ameliorated during the time period.

**Conclusions.** The incidence of 4.6 per 100 000 live births remain unchanged in spite of developed prenatal imaging. The majority 71/130 (55%) of the survivors has biventricular repair. The mortality rate has declined over time, and there was no cardiac death after the age of nine years.