Pulmonary atresia with intact ventricular septum
epidemiology and outcome in children born in Sweden 1980-1999

Akademisk avhandling

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Fakultetsopponent:
Docent Gudrun Björkhem
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Avhandlingen baseras på följande delarbeten:

I. Ekman-Joelsson B-M, Sunnegårdh J, Hanséus K, Berggren H, Jonzon A, Jögi P, Lundell B. The outcome of children born with pulmonary atresia and intact ventricular septum in Sweden from 1980 to 1999. Scand Cardiovasc J 2001; 35: 192-198.

II. Ekman-Joelsson B-M, Gustafsson PM, Sunnegårdh J. Exercise performance after surgery for pulmonary atresia and intact ventricular septum. In manuscript.

III. Ekman-Joelsson B-M, Berggren H, Boll A-B, Sixt R, Sunnegårdh J. Abnormalities in myocardial perfusion after surgical correction of pulmonary atresia with intact ventricular septum. Cardiol Young 2008; 18: 89-95.

IV. Ekman-Joelsson B-M, Berntsson L, Sunnegårdh J. Quality of life in children with pulmonary atresia and intact ventricular septum. Cardiol Young 2004; 14: 615-621.

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Abstract

Aims: To describe children born with pulmonary atresia with intact ventricular septum (PA-IVS) in Sweden between 1980 and 1999, the incidence and outcome of PA-IVS, to examine cardio-pulmonary outcomes in survivors and to evaluate their quality of life.

Material and methods: Eighty-four subjects were identified. All available medical data were evaluated. Among 52 survivors, 29 underwent cardiopulmonary exercise testing and lung function tests at rest and 12 subjects underwent myocardial scintigraphy during exercise test and echocardiography at rest. A questionnaire concerning quality of life was completed by 42 subjects.

Results: The incidence was 4.2/100, 000 live births. Eight subjects had an Ebstein-like tricuspid ostium, 31 had a muscular pulmonary atresia and 40 had a membranous pulmonary atresia. Ventriculo coronary arterial communications (VCAC) were found in 36 subjects (43%). Follow-up time was 14 days to 20 years (median 6 years). Among 52 survivors 32 had biventricular repair and 20 univentricular palliation. The survival rate was 68% ten years after initial surgery. Exercise capacity was reduced, but subjects without VCAC and operated with biventricular repair had better exercise capacity than the others. Lung function was an independent predictor of exercise capacity. Nine of 12 subjects examined had myocardial perfusion defects during exercise, and these were associated with VCACs**.** Right ventricular function, as judged from echocardiography at rest, was impaired, while left ventricular function was normal or slightly impaired. Overall quality of life was similar to that of a healthy control group, but subjects with PA-IVS reported more psychosomatic symptoms.

Summary: PA-IVS is an unusual and heterogeneous congenital heart defect associated with high mortality during the first years of life. Membranous pulmonary atresia was associated with a better outcome than muscular pulmonary atresia with respect to survival, myocardial perfusion defects and exercise capacity. The majority of the survivors had biventricular repair. Overall quality of life was good.

Key words: pulmonary atresia with intact ventricular septum, ventriculo coronary arterial communications, biventricular repair, univentricular palliation, myocardial perfusion, myocardial function, cardiopulmonary exercise, lung function, quality of life, mortality, outcome

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