Pulmonary atresia with intact ventricular septum

Akademisk avhandling

som för avläggande av medicine doktorsexamen vid Sahlgrenska akademin,
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av
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Fakultetsopponent:
Docent Gudrun Björkhem
Universitetssjukhuset
Lund

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

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.
Pulmonary atresia with intact ventricular septum

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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