Pulmonary atresia with intact ventricular septum: long term follow-up after biventricular vs. univentricular correction

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1- To investigate cardiac performance and clinical course after biventricular vs. univentricular repair in PA/IVS. 2- To correlate pre- operative RV and LV function with present RV and LV function. 3- To assess myocardial structural anomalies, which...

Ethical review	Approved WMO
Status	Pending
Health condition type	Congenital cardiac disorders
Study type	Observational invasive

Summary

ID

NL-OMON31965

Source
ToetsingOnline

Brief title
Pulmonary atresia with intact ventricular septum: long term follow-up

Condition

- Congenital cardiac disorders
- Cardiac and vascular disorders congenital

Synonym
cardiac reserve, cardiac response to exercise

Research involving
Human

Sponsors and support

Primary sponsor: Academisch Medisch Centrum
Source(s) of monetary or material Support: Interuniversitair Cardiologisch Instituut

Nederland (ICIN);deel van KNAW

Intervention

Keyword: Biventricular repair, Cardiac reserve, Pulmonary atresia with intact ventricle septum, Univentricular repair

Outcome measures

Primary outcome

The primary endpoints of this study are

1- cardiac reserve, measured as percentual increase in CO or EF

2- Clinical outcome, indicated as a common endpoint including number of

hospital admissions and occurrence of arrhythmia.

Secondary outcome

Secondary endpoints are :

1)VO2 max

2) Quality of Life score

3) Brain natriuretic peptide levels

4) RV function

5) LV function

6) Extent of myocardial scar

Study description

Background summary

Biventricular surgical repair has been employed as a definitive repair in patients with pulmonary atresia with intact ventricular septum (PA/IVS). Theoretically, biventricular physiology is superior to classic univentricular correction (Fontan procedure), because the right ventricle actively sustains the pulmonary circulation. This approach may prevent right sided congestion

with tachyarrhythmias, liver function and coagulation disorders, frequently encountered in the group with univentricular repair. However, biventricular repair may not be a guarantee for superior performance over univentricular repair in PA/IVS as shown by recent small studies. Impaired left and right ventricular performance at the time of definitive repair might be responsible for some disappointing results in the biventricular group. Structural anomalies of the right ventricle such as hypoplasia and coronary perfusion variations may play an important role. In addition, LV function in PA/IVS may be impaired although this has not been adequately investigated in detail. Data on long-term follow-up are limited in this patient group and need further investigation.

Study objective

1- To investigate cardiac performance and clinical course after biventricular vs. univentricular repair in PA/IVS. 2- To correlate pre- operative RV and LV function with present RV and LV function. 3- To assess myocardial structural anomalies, which could contribute to decreased post-operative cardiac performance. 4- To assess retrospectively which patients with PA/IVS could have benefited from biventricular repair.

Study design

Prospective patient based study
Cardiac performance is assessed using dobutamine stress MRI. Delayed contrast hyperenhancement MRI is used for identification of myocardial fibrosis. Clinical outcome will be evaluated by: exercise test, NYHA functional class, and quality of life questionnaires. Serum BNP is measured as parameter for cardiac failure. Echocardiographic studies are performed and compared with preoperative data regarding size of the tricuspid valve and RV. The data of the two PA/IVS groups will be compared.

Study burden and risks

The burden of participation are that the patient has to visit the MAC twice. During the first visit the questionaries will be discussed and the exercise will be performed. During this test the patient will be asked to exercise to his or her maximum. During this test a (pediatric) cardiologist will be present and the patient can resign at any moment. This examination will pose no additional burden or risk than performance of exercise during normal life. During the second visit, blood will be taken during the placement of an IV-canula, neede during the MRI examination, to determine BNP. This procedure is painful but short. During the MRI-examination the IV-canula is used to infuse contrastagent. The patient will notice the infusion because of a cold feeling in his or her arm. This contrastagent poses no risk in this patientgroup and is widely used. Therse is a small risk of allergic reaction for this contrastagent, whcih is rare.

During the exercise part of the MRI-examination, dobutamine will be used to mimic physical exercise. During infusion of dobutamine the heart rate and cardiac output will increase. The patient will notice increase of the heartrate. If all precausions are made this examination will pose minimal risks to the patient. A (pediatric)cardiologist will be present during all examinations and during the MRI-scan the will be monitored using a camera, an alarmbutton, a continuous ECG registration and a continous oxygen-registration

The patient will benefit from participation to this study because his cardiac function and performence will be evaluated in detail and if decreased possible underlying factors can be identified.
Furthermore, the results of this study will help to further rfine the surgical approach to patients born with PA-IVS.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adolescents (12-15 years)
Adolescents (16-17 years)

Adults (18-64 years)
Children (2-11 years)
Elderly (65 years and older)

Inclusion criteria

Treated pulmonary atresia with intact ventricular septum
Age > 8 yrs
No contra-indication for exercise test such as severe aortic-valve stenosis
No contra-indication for MRI examination, such pace maker dependency, cardiac arrhythmia*s or claustrophobia
No contra-indication for Dobutamin, such as prior allergic reaction or cardiac arrhytmias
No contra-indication for contrast agent, such as prior allergic reaction or renal disease

Exclusion criteria

Contra-indications for exercisetest, dobutamine, MRI-scan
Mental retardation

Study design

Design

Study type: Observational invasive

Masking:	Open (masking not used)
Control:	Uncontrolled
Primary purpose:	Diagnostic

Recruitment

NL

Recruitment status:	Pending
Start date (anticipated):	01-05-2008
Enrollment:	30
Type:	Anticipated

Ethics review

Approved WMO
Application type: First submission

Review commission: METC Amsterdam UMC

Study registrations

Followed up by the following (possibly more current) registration

No registrations found.

Other (possibly less up-to-date) registrations in this register

No registrations found.

In other registers

Register	ID
CCMO	NL22564.018.08