Fri. Jul 6, 2018

第2会場

AEPC-JSPCCS Joint Symposium

AEPC-JSPCCS Joint Symposium（ II-AEPCJS）

Congenitally corrected TGA

座長:坂本 喜三郎（静岡県立こども病院心臓血管外科）
座長:Gurleen Sharland（Evelina Children's Hospital）
8:40 AM - 10:10 AM 第2会場 (301)

[II-AEPCJS-01] How to assess the Tricuspid valve and RV function in CCTGA by CMR

○Inga Voges （University Hospital Schleswig-Holstein, Campus Kiel, Department of Congenital Heart Disease and Paediatric Cardiology, Kiel, Germany）

[II-AEPCJS-02] TBA

○Nicolaas Andreas Blom （Department of Pediatric Cardiology Leiden University Medical Center）

[II-AEPCJS-03] Right ventricular function in patients with congenitally corrected transposition of the great arteries

○Ken Takahashi （Department of Pediatrics, Juntendo University Faculty of Medicine ）

[II-AEPCJS-04] Double switch operation for atrioventricular discordance with balanced ventricle

○Hajime Ichikawa, Takaya Hoashi, Tomohiro Nakata, Masatoshi Shimada, Motoki Komori （Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center）

AEPC-JSPCCS Joint Symposium

AEPC-JSPCCS Joint Symposium（ II-AEPCJS）

Congenitally corrected TGA

座長:坂本 喜三郎（静岡県立こども病院心臓血管外科）
座長:Gurleen Sharland（Evelina Children's Hospital）
Fri. Jul 6, 2018 8:40 AM - 10:10 AM 第2会場 (301)

[II-AEPCJS-01] How to assess the Tricuspid valve and RV function in CCTGA by CMR

○Inga Voges （University Hospital Schleswig-Holstein, Campus Kiel, Department of Congenital Heart Disease and Paediatric Cardiology, Kiel, Germany）

[II-AEPCJS-02] TBA

○Nicolaas Andreas Blom （Department of Pediatric Cardiology Leiden University Medical Center）

[II-AEPCJS-03] Right ventricular function in patients with congenitally corrected transposition of the great arteries

○Ken Takahashi （Department of Pediatrics, Juntendo University Faculty of Medicine ）

[II-AEPCJS-04] Double switch operation for atrioventricular discordance with balanced ventricle

○Hajime Ichikawa, Takaya Hoashi, Tomohiro Nakata, Masatoshi Shimada, Motoki Komori （Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center）

(Fri. Jul 6, 2018 8:40 AM - 10:10 AM 第2会場)

[II-AEPCJS-01] How to assess the Tricuspid valve and RV function in CCTGA by CMR

○Inga Voges （University Hospital Schleswig-Holstein, Campus Kiel, Department of Congenital Heart Disease and Paediatric Cardiology, Kiel, Germany）

Congenitally corrected Transposition of the great arteries (ccTGA) is a rare cardiac malformation characterized by the combination of discordant atrioventricular and ventriculo-arterial connections and is usually accompanied by other malformations such as ventricular septal defects, obstruction of the morphologically LV outflow tract and lesions of the tricuspid valve.
In patients with ccTGA the systemic right ventricle (RV) and its tricuspid valve are exposed to systemic arterial afterload and great concern exists about the ability of the anatomic RV to sustain the systemic circulation over the long term. Cardiovascular magnetic resonance imaging plays an important role in the evaluation of ccTGA patients during follow-up as it offers an accurate quantification of RV volumes and function as well as evaluation of the tricuspid valve.
In this presentation the various CMR methods for the detailed assessment of the systemic RV and the tricuspid valve are presented in the light of the current literature in ccTGA.

(Fri. Jul 6, 2018 8:40 AM - 10:10 AM 第2会場)

[II-AEPCJS-02] TBA

○Nicolaas Andreas Blom （Department of Pediatric Cardiology Leiden University Medical Center）

TBA

(Fri. Jul 6, 2018 8:40 AM - 10:10 AM 第2会場)

[II-AEPCJS-03] Right ventricular function in patients with congenitally corrected transposition of the great arteries

○Ken Takahashi （Department of Pediatrics, Juntendo University Faculty of Medicine ）
Keywords: Congenitally corrected transposition of the great arteries, Right ventricular function, Systemic right ventricle

Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart disease. Right ventricular (RV) function has a great impact on clinical outcome in patients with ccTGA. Patients without significant associated intracardiac lesions usually do not have symptoms of heart failure in childhood and do well until adult life. However, the gradual dysfunction of the RV, which supports systemic circulation, will occur after decades in life. RV dysfunction usually remains subclinical for decades. By 45 years of age, more than 30% of patients with isolated ccTGA and more than two thirds of patients with associated lesions develop clinical congestive heart failure.
As mechanisms of RV dysfunction, high pressure inside of the RV causes increased ventricular wall stress

and dysfunction of the trabecular component and valvular apparatus of the RV. Another mechanism of RV dysfunction is thought to be inadequate perfusion of the coronary artery, as only one coronary artery supplies the hypertrophied RV mass. Tricuspid regurgitation (TR) has also been found to be a major contributor to the development of progressive RV dysfunction. In ccTGA, the tricuspid annulus dilates, the papillary muscles move away from the inflow tract, and the tethered cords do not allow the leaflets to occlude the orifice. These mechanisms predispose TR to a steady increase over time.
In this presentation, the mechanisms of RV dysfunction are summarized on the basis of reviewed articles. This will be helpful to the audience in understanding these mechanisms and in their clinical practice.

(Fri. Jul 6, 2018 8:40 AM - 10:10 AM 第2会場)

[II-AEPCJS-04] Double switch operation for atrioventricular discordance with balanced ventricle

○Hajime Ichikawa, Takaya Hoashi, Tomohiro Nakata, Masatoshi Shimada, Motoki Komori （Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center）
Keywords: congenitally corrected transposition of great arteries, double switch operation, atrioventricular discordance

The long-term outcome of the patients with ccTGA has been thought to be affected by the anatomical right ventricular volume overload and tricuspid regurgitation. Although it is almost a common concept that the anatomical repair is ideal, the database of Japanese Society for Thoracic Surgery reports that since 2011, the number of double switch operations (DSO) for ccTGA has been between 10 to 20 per year. On the other hand, physiological or any other procedure for ccTGA is around 100 per year in Japan. This is partly because of the fact that only a small fraction of the ccTGA cohort is suitable for DSO or the physician still hesitate to put the patients on DSO strategy which is a high risk procedure. In the presentation, we revisit the detailed indication for DSO.
There were 81 patients with conventional repair including simple VSD closure or complex LV-PA conduit with interventricular rerouting. DSO was performed in 73 patients with either Senning/Mustard plus Jatene/Rastelli type operation. Ten patients underwent only pacemaker implantation or palliative surgery. Results: Survival rates in the conventional group were poor with 10, 20 and 30 year freedom from death after surgery of 75, 71 and 65%, respectively. The age at initial surgical intervention inversely correlated with the survival (expired 9±16 vs survived 19±20). The survival rate of simple tricuspid valve replacement in 22 patients (average 29 years old) were 91, 91 and 91% in 10, 20 and 30 years after the operation, respectively. Since 1997, the survival rates after double switch operation were 97 and 91.3% at 10 and 20 years, respectively. The median age at the initial surgical intervention was 2.1 year old. Conclusion: When earlier surgical intervention is needed, conventional repair only provide poor outcome. Earlier decision of treatment strategy may improve the clinical outcome of patients with ccTGA.