Sat. Jul 8, 2017

ROOM 3

AP Target Symposium

AP Target Symposium 3 (II-APT3)

Dealing with congenitally corrected transposition of the great arteries - Efforts to minimize late development of systemic ventricular dysfunction

Chair:Yih-Sharng Chen(National Taiwan University Hospital, Taiwan)
Chair:Jun Yoshimoto(Pediatric Cardiology, Shizuoka Children's Hospital, Japan)
10:15 AM - 11:45 AM ROOM 3 (Exhibition and Event Hall Room 3)

[II-APT3-01] Morphological aspects

○Hideki Uemura （Congenital Heart Disease Center, Nara Medical University, Japan）

10:15 AM - 11:45 AM

[II-APT3-02] The late outcome of systemic right ventricle in congenitally corrected transposition of great arteries: Functional repair or anatomical repair

○Hajime Ichikawa[1], Takaya Hoashi[1], Tomohiro Nakata[1], Masatoshi Shimada[1], Akihiko Higashida[1], Hideo Ohuchi[2], Kenichi Kurosaki[2], Isao Shiraishi[2] （1.Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Japan, 2.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Japan）

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[II-APT3-03] Electrical issue of corrected TGA

○Jun Yoshimoto （Department of Cardiology, Shizuoka Children's Hospital, Shizuoka, Japan）

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[II-APT3-04] Congenitally Corrected Transposition: Efforts to minimize late development of systemic RV Failure

○Sertaç M. Çiçek （Department of CV Surgery, Mayo Clinic, USA）

10:15 AM - 11:45 AM

[II-APT3-05] The Senning operation in anatomical repair of congenitally corrected transposition

○James S. Tweddell （Cardiothoracic Surgery, Cincinnati Children’s Hospital Medical Center, USA）

10:15 AM - 11:45 AM

[II-APT3-06] Role of Fontan operation in cc-TGA

○Tae-Jin Yun （Asan Medical Center, University of Ulsan, Korea）

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[II-APT3-01] Morphological aspects

○Hideki Uemura (Congenital Heart Disease Center, Nara Medical University, Japan)

It is essential to clarify the concept behind the terms ' congenitally corrected transposition of the great arteries', ' discordant atrioventricular connections', ' l-transposition', and so on. Overall understanding then will require recognition of morphological diversity and architectural spectra within this entity; not only for atrioventricular/ventriculo-arterial connections, but also atrial arrangement, nature of a ventricular septal defect (if any), obstruction across the ventricular outflow tracts (particularly to the pulmonary arteries), and anatomy of the coronary circulation. The feature of the conduction system has been well documented in this setting.
Such background knowledge should contribute to better comprehension of functional results of the systemic right ventricle or those subsequent to the so-called anatomic biventricular repair in the longer term. The investigators may not necessarily discuss the outcome through a uniform language. Thus, confusion or misleading analysis could go further in this difficult clinical area.

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[II-APT3-02] The late outcome of systemic right ventricle in congenitally corrected transposition of great arteries: Functional repair or anatomical repair

○Hajime Ichikawa[1], Takaya Hoashi[1], Tomohiro Nakata[1], Masatoshi Shimada[1], Akihiko Higashida[1], Hideo Ohuchi[2], Kenichi Kurosaki[2], Isao Shiraishi[2] (1.Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center, Japan, 2.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Japan)

The poor outcome of systemic right ventricle (RV) in congenitally corrected transposition of great arteries (ccTGA) is a well-known fact. Our strategy for the treatment of ccTGA is to place the left ventricle in a systemic position since 1990. The outcome is compared with the functional repair.
Patients and method: One hundred sixty one ccTGA patients with balanced ventricle were included in this retrospective non-randomized analysis. There were 81 patients with conventional repair including simple VSD closure or complex LV-PA conduit with interventricular rerouting. Seventy patients underwent double switch operation of 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 1.8 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.

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[II-APT3-03] Electrical issue of corrected TGA

○Jun Yoshimoto (Department of Cardiology, Shizuoka Children's Hospital, Shizuoka, Japan)

In congenitally corrected TGA (ccTGA), there are several electro-anatomical issues. No.1 is an anatomical issue . SA node lies in normal position, but AV node (AVN) is in abnormal position. The actual position and course of AVN and His bundle differs case by case, especially because of straddling and DORV. Also connective tissue lies superficially and fragile. This seems to be the reason of occurrence of congenital and postsurgical complete AV block. No.2 is electrical and mechanical dyssynchrony. Bundle branch block of systemic ventricle causes electrical and mechanical dyssynchrony and they can be the cause of heart failure. In the case of univentricular physiology, however, interventricular dyssynchrony can cause severe heart failure. No.3 is tachy arrythmia post double switch operation. Both Senning and Mustard operation can cause intra atrial reentry tachycardia(IART). Ablation of IART of ccTGA has difficult problem because of anatomical abnormality of AVN, therefore brockenbrough puncture of intraatrial septum or baffle. Ablation of AVnodal Reentry tachycardia is quite difficult because of anatomical abnormality.

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[II-APT3-04] Congenitally Corrected Transposition: Efforts to minimize late development of systemic RV Failure

○Sertaç M. Çiçek (Department of CV Surgery, Mayo Clinic, USA)

Congenitally corrected transposition of the great arteries is a rare defect representing approximately 0.5% of all congenital heart disease. ccTGA is characterised by the combination of atrioventricular discordance and ventriculo-arterial discordance. This combination, atrioventricular discordance and ventriculoarterial discordance, produces a unique set of management challenges.
Patients with congenitally corrected transposition of the great arteries have a morphological right ventricle that sustains the systemic circulation. In these patients, regurgitation of the systemic atrioventricular valve, the tricuspid valve, is a common finding. When TV regurgitation becomes severe, it is associated with RV failure and decreased survival. Associated anomalies such as ventricular septal defect, morphologic left ventricular outflow tract obstruction and tricuspid valve abnormalities occur in the majority of these patients. Although the presence of these associated conditions influences the clinical presentation of this condition and the type and timing of surgical intervention, ***the ability of the morphologic right ventricle and the tricuspid valve to withstand a lifetime of exposure to systemic pressure largely determines the ultimate outcome of these patients.***
Progressive dysfunction of the tricuspid valve and right ventricle occurs after prolonged exposure to systemic pressure in a substantial percentage of patients who have undergone an atrial switch operation for transposition of the great arteries with increased risk for patients who have accompanying lesions. If sensitive measures of ventricular function are used, an even larger percentage of cc-TGA patients can be shown to have subclinical evidence of limited cardiovascular reserve. A significant percentage of cc-TGA patients had perfusion defects during exercise stress testing with radionuclide imaging.
Increasing morphologic LV pressure either naturally by PS or banding may maintain RV geometry &prevent progression of TR. Shifting the ventricular septum leftward, reduces left ventricular (LV) end-diastolic volume and causes LV to fall within the Frank-Starling curve and reduces LV filling dynamics

and end-diastolic pressure.
Earlier intervention with tricuspid regurgitation even with minimal severity could reverse the future RV failure. Recent data supports the use of tricuspid valve replacement as an alternative to repair, especially in adult patients. The use of the double switch operation is predicated on the assumption that long-term outcomes will be better with the morphologic left ventricle supporting the systemic circulation.
Surgery for congenitally corrected transposition of the great arteries has evolved overtime. Anatomical repair of cc-TGA is now performed with quite satisfactory outcomes in children. The double switch operation has become the preferred surgical procedure in selected cases. However, Systemic A-V valve regurgitation strongly associated with RV dysfunction and CHF whether this is causative or secondary remains speculative.

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[II-APT3-05] The Senning operation in anatomical repair of congenitally corrected transposition

○James S. Tweddell （Cardiothoracic Surgery, Cincinnati Children's Hospital Medical Center, USA）

Current controversies in the management of the patient with congenitally corrected transposition of the great arteries (ccTGA) revolve around the best choice of procedure among the choices of physiologic repair, anatomic repair and single ventricle palliation.(1-3) While anatomic correction of ccTGA aims to improve long term outcome by placing the morphologic left ventricle (LV) as the systemic ventricle and avoiding systemic right ventricular failure the long-term outlook is complicated by a high rate of re-intervention, conduction abnormalities combined with systemic left ventricular dysfunction. Options for the atrial level switch component of the anatomic correction include; the Senning operation, the Mustard procedure or the so called hemi-Mustard combined with a bidirectional Glenn shunt.(4) To maximize the benefit of anatomic repair and limit reoperations, an atrial level switch that minimizes sinus node dysfunction, baffle obstruction and supraventricular arrhythmias is essential.(5-7) While, potentially more complicated to perform the Senning procedure uses native tissue and results in a low rate of late baffle obstruction. Technical strategies to avoid sinus node dysfunction can be successfully applied.(8) The Senning, as opposed the hemi-Mustard, maintains the superior caval vein connection to the atrium which may be important for access for pacemaker and arrhythmia management. The steps of the Senning procedure will be demonstrated with a video and the late outcomes reviewed.

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[II-APT3-06] Role of Fontan operation in cc-TGA

○Tae-Jin Yun （Asan Medical Center, University of Ulsan, Korea）

Patients with congenitally corrected transposition of the great arteries (cc-TGA) can be categorized into discrete three subgroups with respect to the surgical strategies: 1) cc-TGA with ventricular septal defect (VSD) and no pulmonary stenosis (PS) 2) cc-TGA with intact ventricular septum (IVS), and 3) cc-TGA, VSD and PS or pulmonary atresia (PA). The first subset is thought to be the best candidates for double switch operation (DSO), because patients rapidly develop severe congestive heart failure which necessitates urgent surgical interventions (i.e. pulmonary artery banding for staged DSO or early primary DSO). In the second subgroup, pulmonary artery banding for the training of the morphologic left ventricle is commonly indicated when regurgitation of the atrioventricular valve of the systemic right ventricle (i.e. tricuspid regurgitation) develops. As long as the tricuspid valve is competent, however, it is very difficult to determine the exact timing of any surgical intervention. The best surgical option for the third group is still under debate. Anatomical repair (i.e. Intraventricular rerouting from the morphologic LV to the aortic valve in association with atrial switch operation and right ventricle to pulmonary artery extracardiac conduit interposition) has been attempted for this subset, but it has been pointed out that the potential risks of morbidities associated with atrial switch operation, RV-PA conduit stenosis, and left ventricular outflow tract obstruction, particularly in case of restrictive VSD,

may outweigh the potential benefits of recruiting the morphological left ventricle to the systemic circulation. Furthermore, superiority of anatomical repair over the Fontan procedure does not seem to be fully substantiated by the clinical reports pertaining to the long-term outcome of anatomical repair for cc-TGA with VSD and PS.