Thu. Jul 16, 2015

第2会場

海外招請講演

海外招請講演1

What Is the Data? Biventricular Conversion and Techniques for Left Sided Reconstruction and Growth

座長:佐野 俊二 (岡山大学大学院)
9:50 AM - 10:20 AM 第2会場 (1F ペガサス B)

[IL01-01] What Is the Data? Biventricular Conversion and Techniques for Left Sided Reconstruction and Growth

○Emile A. Bacha （Cardiothoracic Surgery, Columbia University Medical Center, NewYork-Presbyterian Morgan Stanley Children's Hospital , USA）

海外招請講演

海外招請講演2

New Insights in the Marfan Syndrome

座長:丹羽 公一郎 (聖路加国際病院)
11:35 AM - 12:05 PM 第2会場 (1F ペガサス B)

[IL02-01] New Insights in the Marfan Syndrome

○Barbara JM Mulder （Academic Medical Center, The Netherlands）

海外招請講演

海外招請講演3

Long term results after surgery for adults with Congenital heart disease （tentative）

座長:芳村 直樹 (富山大学)
1:10 PM - 1:40 PM 第2会場 (1F ペガサス B)

[IL03-01] TBA

Christian Pizzaro （Cardiothoracic Surgery, Nemours Cardiac Center, A.I. du Pont Hospital for Children, USA）

第3会場

海外招請講演

海外招請講演4

Fetal Cardiology in 2015: What Can We Achieve?

座長:稲村 昇 (大阪府立母子保健総合センター)
4:00 PM - 4:30 PM 第3会場 (1F ペガサス C)

[IL04-01] Fetal Cardiology in 2015: What Can We Achieve?

○Gurleen Sharland （Evelina London Children's Hospital, UK）

Fri. Jul 17, 2015

第2会場

海外招請講演

海外招請講演5

Long-term Outcomes after Surgery

座長:宮地 鑑 (北里大学)

11:30 AM - 12:00 PM 第2会場 (1F ペガサス B)

[IL05-01] Long-term Outcomes After Surgery

○Yves d'Udekem （The Royal Children's Hospital , Australia）

海外招請講演

海外招請講演1

What Is the Data? Biventricular Conversion and Techniques for Left Sided Reconstruction and Growth

座長:佐野 俊二 (岡山大学大学院)

Thu. Jul 16, 2015 9:50 AM - 10:20 AM 第2会場 (1F ペガサス B)

IL01

所属正式名称：佐野俊二(岡山大学大学院医歯薬学総合研究科　心臓血管外科)

[IL01-01] What Is the Data? Biventricular Conversion and Techniques for Left Sided Reconstruction and Growth

○Emile A. Bacha （Cardiothoracic Surgery, Columbia University Medical Center, NewYork-Presbyterian Morgan Stanley Children's Hospital , USA）

(Thu. Jul 16, 2015 9:50 AM - 10:20 AM 第2会場)

[IL01-01] What Is the Data? Biventricular Conversion and Techniques for Left Sided Reconstruction and Growth

○Emile A. Bacha （Cardiothoracic Surgery, Columbia University Medical Center, NewYork-Presbyterian Morgan Stanley Children's Hospital , USA）

Patients born with Borderline left ventricles (LV) continue to pose a vexing problem in terms of management. They are best analyzed sequentially anatomically, starting with the mitral valve morphology, size and function of LV, presence of Endocardial Fibroelastosis (EFE), LVOT obstruction, aortic valve, and aortic arch, as well as physiologically (PGE-dependency, clinical features such as weight, prematurity, presence of other non-cardiac malformations or genetic syndromes). There is currently no best formal scientific way to determine whether a 2-ventricle or a single ventricle pathway is preferable for a given patient. Several echo criteria have been developed, but none have been found to be universally useful. MRI criteria of minimal LV volumes are more specific, but MRIs are not always available to newborns. Thus, this matter remains still as much art as science. In recent years we have been able to "push" more patients with borderline left hearts towards a 2V circulation. However, this has come at a cost of repeated surgeries and the ever-present specter of left atrial hypertension, LV diastolic dysfunction and eventual lung damage (and need for heart-lung transplantation) remains. Surgical techniques used are adapted to the cardiac morphology, and include valve plasty, EFE resection (sometimes repeatedly), creation of a restrictive ASD to force blood flow through the diminutive LV, and valve replacements including mechanical mitral valve replacement, mitral implantation of stented Melody or Sapien valves, or Ross procedures). Meanwhile, outcomes with Fontan circulation continue to improve and "good" Fontan patients do exist.
In summary, 2V repairs in borderline left heart patients should be pursued, but only in selected patients and ideally in centers with extensive experience. Typically, when in doubt, one would initially perform a Stage I Norwood operation (which includes a restrictive ASD and maybe "modified" to allow easier take-down to 2V repair), then re-assess at about 6 months of age with catheterization and MRI before deciding which route to take.

海外招請講演

海外招請講演2

New Insights in the Marfan Syndrome

座長:丹羽 公一郎 (聖路加国際病院)

Thu. Jul 16, 2015 11:35 AM - 12:05 PM 第2会場 (1F ペガサス B)

IL02

所属正式名称：丹羽公一郎(聖路加国際病院　心血管センター)

[IL02-01] New Insights in the Marfan Syndrome

○Barbara JM Mulder （Academic Medical Center, The Netherlands）

(Thu. Jul 16, 2015 11:35 AM - 12:05 PM 第2会場)

[IL02-01] New Insights in the Marfan Syndrome

○Barbara JM Mulder （Academic Medical Center, The Netherlands）

Marfan syndrome is a connective tissue disorder, caused by mutations in the FBN1 gene encoding fibrillin-1 protein, a structural component of elastic fibers in the tunica media in large arteries. Mortality and morbidity is mainly determined by the development of an aortic aneurysm and subsequent dissection. Prophylactic aortic surgery has increased survival tremendously. In addition, apart from β-blockers the angiotension blocker losartan has recently been shown to slow down aortic dilatation. However, the beneficial effect of losartan appears to be heterogeneous and is more pronounced in patients with a mutation causing haploinsufficiency (mutations resulting in deficient fibrillin-1 protein) compared to patients with a dominant negative mutation (mutations resulting in abnormal fibrillin-1). Around one third of Marfan patients have a haploinsufficiency mutation. They are at increased risk for cardiovascular death, aortic dissection or prophylactic surgery compared to dominant negative patients. So, especially in these haploinsufficient Marfan patients, losartan therapy should be advised both in unoperated patients and after elective aortic root surgery. After aortic root replacement the distal part of the aorta is at increased risk of type B dissection. Type B dissection has become a major problem in these patients since it may occur when the proximal descending aorta is only slightly dilated. Losartan appears to reduce the incidence of type B dissections. In conclusion, for optimal assessment of prognosis and treatment of Marfan patients, more extensive genetic screening, and evaluation of the FBN1 mutation effect on fibrillin-1 protein is warranted. Treatment with losartan seems beneficial in many Marfan patients, but for assessment of the exact role of losartan in Marfan syndrome, the results of running trials should be awaited.

海外招請講演

海外招請講演3

Long term results after surgery for adults with Congenital heart disease （tentative）

座長:芳村 直樹 (富山大学)

Thu. Jul 16, 2015 1:10 PM - 1:40 PM 第2会場 (1F ペガサス B)

IL03

所属正式名称：芳村直樹(富山大学)

[IL03-01] TBA

Christian Pizzaro （Cardiothoracic Surgery, Nemours Cardiac Center, A.I. du Pont Hospital for Children, USA）

(Thu. Jul 16, 2015 1:10 PM - 1:40 PM 第2会場)

[IL03-01] TBA

Christian Pizzaro （Cardiothoracic Surgery, Nemours Cardiac Center, A.I. du Pont Hospital for Children, USA）

海外招請講演

海外招請講演4

Fetal Cardiology in 2015: What Can We Achieve?

座長:稲村 昇 (大阪府立母子保健総合センター)

Thu. Jul 16, 2015 4:00 PM - 4:30 PM 第3会場 (1F ペガサス C)

IL04

所属正式名称：稲村昇(大阪府立母子保健総合センター　小児循環器科)

[IL04-01] Fetal Cardiology in 2015: What Can We Achieve?

○Gurleen Sharland （Evelina London Children's Hospital, UK）

(Thu. Jul 16, 2015 4:00 PM - 4:30 PM 第3会場)

[IL04-01] Fetal Cardiology in 2015: What Can We Achieve?

○Gurleen Sharland （Evelina London Children's Hospital, UK）

Cardiac malformations are one of the commonest types of congenital abnormality and remain a major cause of morbidity and mortality in infancy. It is possible to detect most forms of major congenital heart disease (CHD), as well as some of the minor forms, during fetal life. Detection in early pregnancy allows parental choice and allows time for parents to be prepared for the likely course of events after delivery. Also, confirming normality of the fetal heart can also be of great benefit in providing reassurance to parents at high risk of having a child with CHD.

Antenatal screening for CHD was introduced 30 years ago and since then there have been many changes, though prenatal diagnosis of CHD remains a challenge.**Improvement in prenatal screening/detection**

Obstetric screening for CHD, using initially the four chamber view and then views of the outflow tracts and more lately the 3 vessel view, plays a vital role in prenatal detection. There is still significant variation in the effectiveness of screening but overall this has been improving.

Changes in spectrum of abnormality detected

The severe end of the spectrum of CHD is usually detected before birth but more types of lesion are increasingly detected.

Advancement in precision of fetal cardiac diagnosis

Improvement in ultrasound imaging has allowed more detailed and precise diagnosis and more accurate prediction of postnatal management and outcome.

Newer techniques to help refine diagnosis

Use of techniques such as 3D/4D echocardiography, speckle tracking and MRI can help to refine fetal diagnosis.Management options following prenatal diagnosis vary in different centres and countries. This depends on local laws and customs as well as paediatric cardiology and surgical facilities available for the care of the affected baby. The outcome will also be affected by these factors.

海外招請講演

海外招請講演5

Long-term Outcomes after Surgery

座長:宮地 鑑 (北里大学)

Fri. Jul 17, 2015 11:30 AM - 12:00 PM 第2会場 (1F ペガサス B)

IL05

所属正式名称：宮地鑑(北里大学医学部　心臓血管外科)

[IL05-01] Long-term Outcomes After Surgery

○Yves d'Udekem （The Royal Children's Hospital , Australia）

(Fri. Jul 17, 2015 11:30 AM - 12:00 PM 第2会場)

[IL05-01] Long-term Outcomes After Surgery

○Yves d'Udekem （The Royal Children's Hospital , Australia）

The Australian and New Zealand Fontan Registry collects all data of patients who have undergone Fontan surgery living in the region. With 1300 Fontan patients alive it is the largest database of Fontan patients. It is has allowed to specify outcomes for the Fontan population. The survival is better than expected, with 76% of the patients with an atrial pulmonary connection Fontan alive at 25 years and 97% of those with a lateral tunnel and an extra cardiac conduit alive at 15 years. These results clearly show us that the majority of Fontan patients will survive three decades after their Fontan operations. We have already demonstrated that their physical capacity was better than expected, and that their quality of life was absolutely normal.

We are now facing the challenge to support this population as they age into adulthood. There is doubt about the best medications to support them. We have demonstrated that Warfarin is of no benefit compared to Aspirin. No anti-hypertensive agents bring any benefits. The only therapy that has been proven effective in increasing cardiac output and exercise capacity is regular exercise training. The possibility of a large proportion of these patients facing liver and renal failure is looming. The majority of these patients present mild abnormalities but their real impact is still unclear.

The therapeutic options for patients facing a failure of their circulation are being explored. It has become clear that Fontan conversion of atrio-pulmonary connection should occur early if we want the patients to benefit from this intervention. Supporting the circulation with assist devices met its first success'. Centralized system for heart transplantation for these complex patients is mandatory to achieve favourable outcomes.