#### Mon. Nov 23, 2020

#### Track2

JSPCCS-AEPC Joint session

JSPCCS-AEPC Joint session ( II-AEPCJS)

座長:坂本 喜三郎 (静岡県立こども病院) 座長:Katarina Hanseus (Children's Heart Center, Skane University Hospital, Lund, Sweden) 4:00 PM - 5:30 PM Track2

[II-AEPCJS-1] Impacts of transcatheter balloon aortic valvuloplasty in children with aortic valvular stenosis

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[II-AEPCJS-3] Current surgical approaches to congenital aortic valve disease in Japan 
<sup>O</sup>Akio Ikai, Kentaro Watanabe, Tomonori
Ishidou, Hiroki Ito, Masaya Murata, Keiichi
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[II-AEPCJS-4] Optimal indication and surgical

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Surgery technique for AS/AR: the role of aortic valve replacement in the very young patient

ORoberto Di Donato (Al Jalila Children's Hospital / Dubai, United Arab Emirates)

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### [II-AEPCJS-1] Impacts of transcatheter balloon aortic valvuloplasty in children with aortic valvular stenosis

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[II-AEPCJS-2] A national study of the outcome after treatment of critical aortic stenosis in the neonate

Cecilia Kjellberg Olofsson<sup>1, 2</sup>, <sup>O</sup>Katarina Hanseus<sup>3</sup>, Jens Johansson Ramgren<sup>3</sup>, Mats Johansson Synnergren <sup>4</sup>, Jan Sunnegårdh <sup>2,4</sup> (1.Department of Pediatrics, Sundsvall Hospital, Sundsvall, Sweden, 2.Department of Pediatrics, Institute of Clinical Sciences, University of Gothenburg, Gothenburg, Sweden, 3.Children's Heart Center, Skåne University Hospital, Lund, Sweden, 4.Children's Heart Center, The Queen Silvia Children's Hospital, Sahlgrenska University Hospital, Gothenburg, Sweden)

[II-AEPCJS-3] Current surgical approaches to congenital aortic valve disease in Japan

OAkio Ikai, Kentaro Watanabe, Tomonori Ishidou, Hiroki Ito, Masaya Murata, Keiichi Hirose, Kisaburo Sakamoto (Mt.Fuji Shizuoka Children's Hospital, Cardiovascular Surgery)

[II-AEPCJS-4] Optimal indication and surgical technique for AS/AR: the role of aortic valve replacement in the very young patient

<sup>O</sup>Roberto Di Donato (Al Jalila Children's Hospital / Dubai, United Arab Emirates)

(Mon. Nov 23, 2020 4:00 PM - 5:30 PM Track2)

# [II-AEPCJS-1] Impacts of transcatheter balloon aortic valvuloplasty in children with aortic valvular stenosis

OJun Muneuchi <sup>1</sup>, Ayako Kuraoka <sup>2</sup>, Yusaku Nagatomo <sup>3</sup>, Koichi Yatsunami <sup>4</sup>, Koichi Sagawa <sup>2</sup>, Hazumu Nagata <sup>3</sup>, Yuichiro Sugitani <sup>1</sup>, Mamie Watanabe <sup>1</sup>: Kyushu Congenital Heart Research Group. (1.Department of Pediatrics, Kyushu Hospital, Japan Community Healthcare Organization, 2.Department of Cardiology, Fukuoka Children's Hospital, 3. Department of Pediatrics, Graduate School of Medical Science, Kyushu University, 4.Department of Pediatric Cardiology, Kumamoto City Hospital)

Background: It is controversial whether children with isolated aortic valvular stenosis undergo transcatheter balloon aortic valvotomy (BAV) or surgical aortic valvotomy (SAV) as the first intervention. This multicenter retrospective cohort study aimed to explore the rates of survival and reintervention after BAV or SAV among them.

Methods: We studied subjects who underwent BAV or SAV at 4 tertiary congenital heart centers in Kyushu, Japan. We compared survival, and freedom from reintervention and aortic valve replacement (prosthetic or autograft) between the groups

Results: A total of 78 subjects were enrolled. Age and aortic valve annulus z-score at the first intervention were 70 (23—415) months and -0.72 (-1.84—0.60), respectively. There were 62 BAV and 16 SAV. During the follow-up period of 13.3 (5.9—16.7) years, there was no significant difference in 10-year survival between the groups (BAV: 88% vs SAV:100%, P= 0.162). Reinterventions included BAV in 3, surgical valve plasty in 9, Ross operation in 17, and prosthetic valve replacement in 6. Freedom from reintervention at 10 years were 46% and 65% in BAV and SAV subjects, respectively (P=0.592), while freedom from valve replacement at 10 years were 77% and 85% in BAV and SAV subjects, respectively (P=0.988).

Conclusions: Long-term survivals after BAV and SAV were acceptable. Freedom from reintervention and from prosthetic/autograft valve replacement were similar between the groups.

(Mon. Nov 23, 2020 4:00 PM - 5:30 PM Track2)

# [II-AEPCJS-2] A national study of the outcome after treatment of critical aortic stenosis in the neonate

Cecilia Kjellberg Olofsson<sup>1, 2</sup>, <sup>O</sup>Katarina Hanseus<sup>3</sup>, Jens Johansson Ramgren<sup>3</sup>, Mats Johansson Synnergren <sup>4</sup>, Jan Sunnegårdh <sup>2,4</sup> (1.Department of Pediatrics, Sundsvall Hospital, Sundsvall, Sweden, 2.Department of Pediatrics, Institute of Clinical Sciences, University of Gothenburg, Gothenburg, Sweden, 3.Children's Heart Center, Skåne University Hospital, Lund, Sweden, 4.Children's Heart Center, The Queen Silvia Children's Hospital, Sahlgrenska University Hospital, Gothenburg, Sweden)

Aortic stenosis presenting with symptoms in the neonate is a serious condition that has been associated with significant risk for pre-, intra- and postoperative mortality and the need for repeated reinterventions. This study describes short-term and long-term outcome after treatment of critical valvular aortic stenosis in neonates in Sweden, with surgical valvotomy as first choice intervention. Methods: All neonates in Sweden treated for critical aortic stenosis between 1994 and 2016 were included. Patient files were analyzed and cross-checked against the Swedish National Population Registry as of December 2017, giving complete survival data. Diagnosis was confirmed by

reviewing echo studies. Critical aortic stenosis was defined as valvular stenosis with duct-dependent systemic circulation or depressed left ventricular function with an echocardiographic measurement of fractional shortening of 28% or below. Primary outcome was all-cause mortality and secondary outcomes were reintervention and aortic valve replacement. Results: Sixty-one patients were identified (50 boys, 11 girls). Primary treatment was surgical valvotomy in 52 neonates and balloon valvotomy in 6. Median age at initial treatment was 5 days (0-26), and median follow-up time was 10.8 years (0.14-22.6). There was no 30-day mortality but four late deaths. Freedom from reintervention was 66%, 61%, 54%, 49%, and 46% at 1, 5, 10, 15, and 20 years, respectively. Median time to reintervention was 3.4 months (4 days to 17.3 years). Valve replacement was performed in 23 patients (38%). Conclusions: Aortic stenosis in the neonate is the start of lifelong need for surgical and catheter-based interventions and follow-up. Primary treatment with high short and long-term survival is possible. Surgical valvotomy is a safe and reliable treatment in these critically ill neonates, with no 30-day mortality and long-term survival of 93% in this national study. At 10 years of age, reintervention was performed in 54% and at end of follow-up 38% had had an aortic valve replacement.

(Mon. Nov 23, 2020 4:00 PM - 5:30 PM Track2)

# [II-AEPCJS-3] Current surgical approaches to congenital aortic valve disease in Japan

OAkio Ikai, Kentaro Watanabe, Tomonori Ishidou, Hiroki Ito, Masaya Murata, Keiichi Hirose, Kisaburo Sakamoto (Mt.Fuji Shizuoka Children's Hospital, Cardiovascular Surgery)

Surgical treatment of aortic valve disease in children remains challenging due to the child's underlying factors, such as growth potential, the valve morphology itself, and unavailability of right size of mechanical valve. In addition, treatment strategies for critical aortic stenosis in newborns are controversial because catheter intervention, which is less invasive than surgical treatment, is an alternative approach. However, catheter interventions are still palliative due to higher re-intervention rate than surgical interventions. Ross procedure is an additional alternative approach. However, the Ross procedure for newborns and infants is technically demanded to achieve good results. Furthermore, the long-term results of the Ross procedure are still skeptical.

Last two decades, we have performed aortic valvuloplasty whenever possible, if the size of the patient's aortic annulus is acceptable. Since 1999, we have been aortic valve plasty in 69 cases consisting of 47 simple cases without patch and 22 cases with patch. Aortic valvuloplasty techniques using autologous pericardial patch consist of cusp extension, cusp reconstruction, and commissural reconstruction. Moreover, we have developed a new surgical approach, "open sleeve technology". It makes an incision in the hypoplastic commissure below the aortic annulus to obtain a good surgical field.

Aortic valve repair less than 3 years old shows acceptable survival rate and suboptimal reintervention rate. However, the group was able to grow and develop without anticoagulant therapy while maintaining acceptable left ventricular size and function. They have a possibility to reach mechanical valve replacement or Ross procedure with lower risk. In patients more than 4 years old, the results of aortic valve repair with and without patch showed reasonable mortality and reintervention rate.

In this session, we hope to discuss lifelong treatment strategies for congenital aortic valve disease.

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ORoberto Di Donato (Al Jalila Children's Hospital / Dubai, United Arab Emirates)

Aortic valve replacement may be required at a very early age (neonates, infants and very young children) for severe aortic stenosis and/or regurgitation, either as a primary or a secondary choice, the latter in the cases of failed balloon or surgical aortic valvuloplasty. Surgical options for this patient population are particularly limited because of patient size and lack of suitable prosthetic valves (mechanical or biological). Therefore, in the past decade, the use of pulmonary autograft has been increasingly recognized as the best solution. In fact, it almost entirely satisfies all the criteria of the ideal prosthesis in aortic position, being: 1) Available, i.e. collected from the same patient, provided the absence of pulmonary regurgitation. 2) Efficient, in a similar way to a normal aortic valve, with early normalization of left ventricular function. 3) Durable, based on a dual mechanism of growth and dilatation, though variable from patient to patient. 4) Thromboresistant and fully biocompatible, without need for anticoagulation. 5) Implantable, although usually at the expense of aortic annulus enlargement by means of a challenging surgical procedure, the Ross/Konno operation. Neonates and infants undergoing this operation are a high-risk group, with an average hospital mortality in the neonatal group of up to 20-30% in multi-institutional studies, i.e. comparable with some of the highest risk procedures. Several risk factors have been identified in this age cohort, e.g.: critical aortic stenosis with preoperative ventricular dysfunction, need for postoperative mechanical circulatory support, aortic arch hypoplasia and mitral disease. Procedurerelated long-term survival is generally reported as very satisfactory with excellent left ventricular functional parameters. Conversely, freedom from reoperation on either the left or the right ventricular outflow tract (or both, "two-valve disease") declines progressively with time, although variably from institution to institution. Interestingly, it has been noted that the reintervention rate on the aortic outflow tends to be lowest in infants compared with older patients, whereas age is inversely related to need for reintervention on the pulmonary outflow tract. In fact, the planned need for right ventricle-to-pulmonary artery conduit is generally considered the true Achilles' heel of the Ross (± Konno) operation.

Among alternative options for aortic valve replacement at a very young age (including infants), it is worth mentioning the initial experience with the Ozaki procedure, recently reported by the Boston group. Encouraging short-term results were achieved also in young patients with small aortic annulus, adding a procedure for surgical annular enlargement.