JSPCCS-TSPC Joint Session

Fri. Jul 11, 2025 8:30 AM - 9:30 AM JST | Thu. Jul 10, 2025 11:30 PM - 12:30 AM UTC Room2（Fine Arts Center 1F Middle Auditorium）

JSPCCS-TSPC Joint Session(II-TSPCJS)
Functional Limitations and Management of the Systemic Right Ventricle

Chair:Shintaro Nemoto（Department of Thoracic and Cardiovascular Surgery, Osaka Medical and Pharmaceutical University）

[II-TSPCJS-1]

Hemodynamic Assessment by Catheterization in the Management of Patients with TGA after Atrial Switch

○Ayako Ishikita[1], Tomoyasu Suenaga[1], Akiko Nishizaki[1], Takamori Kakino[1], Ichiro Sakamoto[1], Eiko Terashi[2], Kenichiro Yamamura[2], Kohtaro Abe[1] (1.Department of Cardiovascular Medicine, Kyushu University Hospital, 2.Department of Pediatrics, Kyushu University Hospital)

[II-TSPCJS-2]

Sudden Cardiac Death in Adults with a Systemic Right Ventricle

○Chun-Wei Lu (Adult Congenital Heart Center, Department of Pediatric Cardiology, National Taiwan University Children's Hospital, Taipei, Taiwan)

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Keywords：Systemic RV、GDMT、pulmonary hypertension

Systemic RV dysfunction and concomitant pulmonary hypertension (PH) are important prognostic factors in patients with transposition of the great arteries (TGA) after atrial switch, but their management has not been standardized. Here, we present our practice based on hemodynamic assessment by catheterization. Patients with RV dysfunction are treated with medical therapy (GDMT) according to current heart failure (HF) guidelines; response to GDMT varied as shown below:**Case1.** A 37-year-old man presented with HF and severe TR. Initially, TVR for severe TR was not indicated due to combined pre- and post-capillary PH (mean PAP 64mmHg, PCWP 17mmHg, and PVR 13w.u.). After GDMT, PH was improved (mean PAP 22mmHg, PCWP 11mmHg, and PVR 2.0w.u.). TVR was subsequently performed, and the patient is now NYHA Class I.**Case2.** A 35-year-old man presented with HF, but no PH. HF symptoms were controlled with maximal GDMT. However, routine catheterization 3 years after starting GDMT revealed postcapillary PH (PCWP 22mmHg, mean PAP 28mmHg, and PVR 2.8w.u.). He was registered as a recipient for heart transplantation. Now he is waiting for heart transplant with support by aRVAD.**Case3.** A 48-year-old woman presented with severe TR, combined pre- and post-capillary PH (mean PAP 80mmHg, PCWP 30mmHg, PVR 14.0w.u). Despite maximal GDMT, clinical improvement was limited (mean PAP 60mmHg, PCWP 21mmHg, PVR 4.7w.u). In severe PH, additional surgical intervention or heart transplantation was contraindicated.Repeated evaluation by catheterization is a valuable in the management of TGA after atrial switch.

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Keywords：Systemic Right Ventricle、Sudden Cardiac Death、Adult Congenital Heart Disease

A systemic right ventricle (SRV) is most commonly seen in patients with congenitally corrected transposition of the great arteries (ccTGA) or in those who have undergone atrial switch procedures for complete transposition of the great arteries (d-TGA). Advances in medical and surgical care have significantly improved long-term survival in this population. However, adults with an SRV remain at a markedly increased risk of sudden cardiac death (SCD), arising from a range of causes and triggers. This presentation explores the current understanding of SCD in this unique group, with a focus on pathophysiological mechanisms, challenges in risk stratification, exercise recommendations, and clinical management. Key issues include progressive SRV dysfunction, arrhythmogenic substrates, and the limitations of conventional SCD risk models. We will examine the emerging roles of advanced imaging, electrophysiological studies, and novel biomarkers in identifying high-risk individuals. In addition, current guideline recommendations and ongoing controversies regarding the use of implantable cardioverter-defibrillators (ICDs) will be discussed. Ultimately, the aim is to highlight strategies for improved surveillance and individualized interventions to reduce SCD risk and enhance long-term outcomes in adults with an SRV.