Panel Discussion

Fri. Jul 10, 2026 2:00 PM - 3:30 PM JST | Fri. Jul 10, 2026 5:00 AM - 6:30 AM UTC | Room1（Conference Hall A）

[PD6] パネルディスカッション 06（II-PD6）

小児期のブルガダ症候群およびSCN5A遺伝子バリアント関連疾患

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[II-PD6-1] Brugada syndrome and cardiac conduction in children

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Over the past decades, increasing recognition of pediatric loss-of-function SCN5A channelopathies has demonstrated that these disorders represent a broad and evolving spectrum of electrical heart disease rather than isolated Brugada syndrome. In genetically confirmed pediatric SCN5A disease, affected children may present with highly variable phenotypes including Brugada syndrome, progressive cardiac conduction disease, sinus node dysfunction, atrial arrhythmias, ventricular arrhythmias, aborted cardiac arrest, and sudden cardiac death. In many patients, conduction abnormalities such as PR prolongation and QRS widening are the earliest and most consistent electrocardiographic manifestations, often preceding development of a spontaneous type 1 Brugada ECG pattern.

A major pediatric-specific feature is the strong association between fever and arrhythmic manifestations, with febrile illness frequently unmasking latent disease or triggering malignant arrhythmias, particularly in younger children. Phenotypic expression is dynamic and age dependent, with considerable overlap between conduction disease and Brugada phenotypes within the same patient or family. Our observations further highlight the importance of family screening and serial ECG evaluation, as clinical manifestations may evolve over time despite initially limited findings.

Management of pediatric loss-of-function SCN5A channelopathies remains challenging because risk stratification in children differs substantially from adults. Current strategies emphasize aggressive antipyretic treatment during fever, hospital admission during fever episodes, avoidance of sodium channel–blocking drugs, and individualized therapy. Quinidine may play an important role in selected patients with high risk Brugada phenotype, while beta-blocker therapy can be beneficial in specific patients with cardiac conduction disease and monomorphic ventricular tachycardia. Implantable cardioverter-defibrillator therapy is usually reserved for patients with aborted cardiac arrests. Recognition of the diverse pediatric phenotypes associated with loss-of-function SCN5A variants is essential for timely diagnosis, prevention of arrhythmic events, and development of age-specific management strategies