

PediRhythm XII

**WORLD CONGRESS OF PEDIATRIC
& CONGENITAL HEART RHYTHM
DISORDERS**

SEPTEMBER 10-12, 2026
EUGENIDES FOUNDATION
ATHENS, GREECE

**ABSTRACT
BOOK**

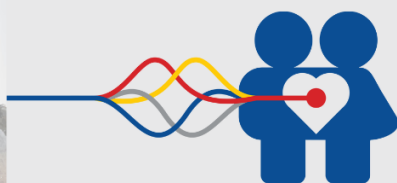
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ORAL PRESENTATIONS



OR01 | CLINICAL OUTCOMES AND THERAPEUTIC STRATEGIES IN PEADIATRIC LONG QT SYNDROME: INSIGHTS FROM THE SPANISH NATIONAL REGISTRY

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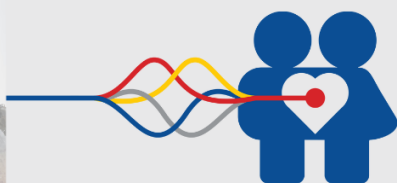
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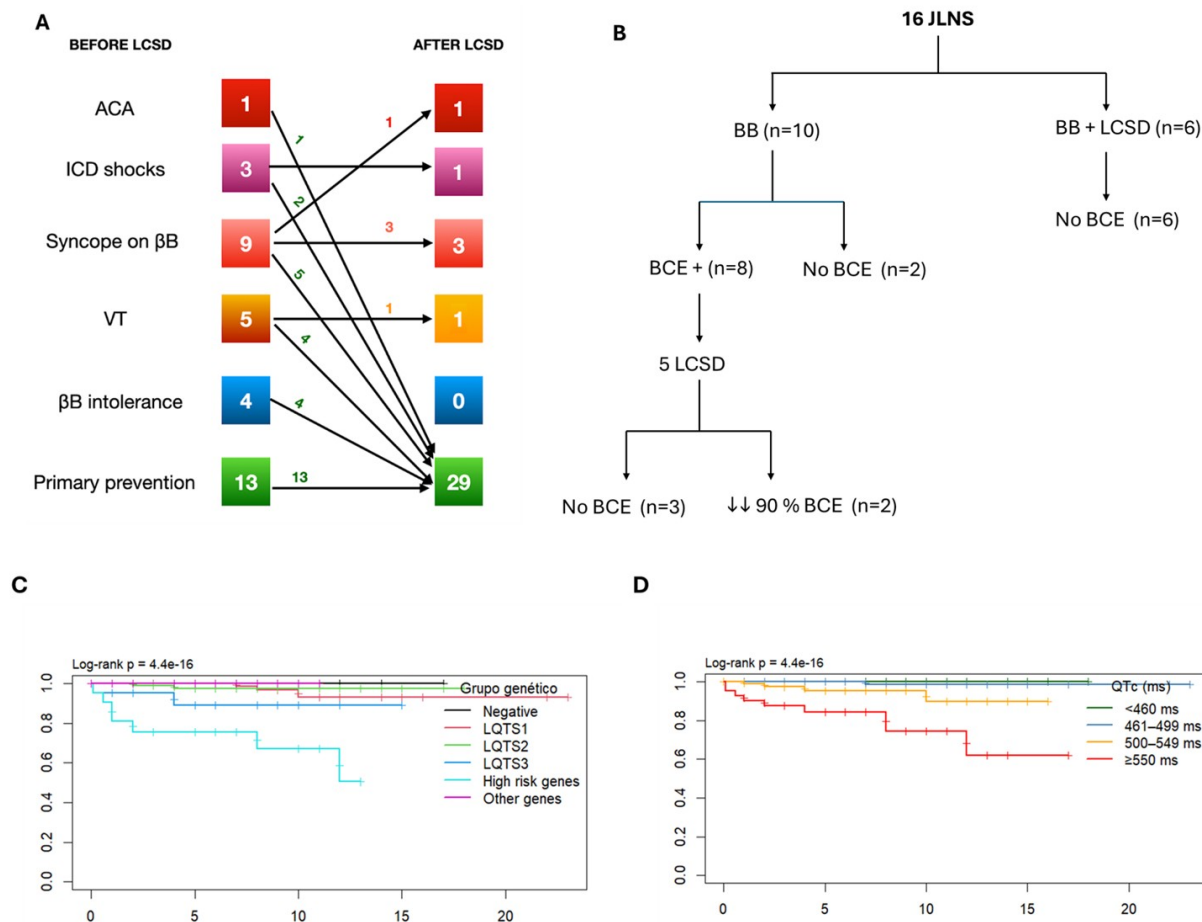
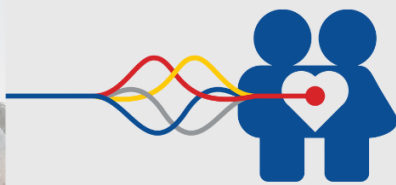
Background and aims: Congenital Long QT syndrome (LQTS) is a heterogeneous disorder in which genotype and QTc duration modulate the risk of major arrhythmic events (MAEs), but contemporary paediatric outcome data remain limited. This study aimed to characterise clinical features, management strategies, and predictors of MAEs in a nationwide paediatric LQTS cohort.

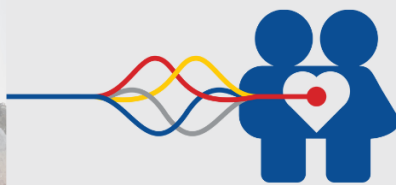
Methods: This retrospective multicentre study analysed children (<18 years) diagnosed with LQTS across 30 tertiary centres (2004–2024) using 2022 ESC criteria. MAEs were defined as sudden cardiac death, aborted cardiac arrest or appropriate implantable cardioverter-defibrillator (ICD) therapy.

Results: The cohort included 371 children (58% male; median diagnosis age 6.0 years; IQR 0.25–11 years). Pathogenic/likely pathogenic variants were identified in 86.5%, mainly in *KCNQ1*, *KCNH2* or *SCN5A*, while 5.7% harboured high-risk genotypes (Jervell–Lange–Nielsen, calmodulinopathies, *CACNA1C*/Timothy). β -blockers were prescribed in 92.4%. Over a median 6-year follow-up, 14 children (3.8%) experienced MAEs, which occurred predominantly in those with high-risk genotypes, $QTc \geq 550$ ms or very early presentation. MAEs were rare in LQT1–3 with $QTc < 500$ ms. Foetal/neonatal bradycardia (5.1%) correlated with high-risk genotypes and adverse outcomes. ICDs were implanted in 33 children (8.9%); ten (30%) received appropriate therapies and eight (24%) experienced complications. Left cardiac sympathetic denervation (LCSD) was performed in 33 (8.9%), mainly high-risk patients, and was associated with >80% arrhythmia-free survival without major complications.

Conclusions: In this nationwide paediatric LQTS cohort, MAEs were uncommon and clustered in children with malignant genotypes, markedly prolonged QTc and very early presentation, particularly foetal or neonatal bradycardia. These data support current genotype and QTc-guided management strategy, with β -blockers as the cornerstone therapy and selective use of LCSD and ICDs in high-risk profiles.

Keywords Long QT Syndrome; Paediatrics; Genotype-Phenotype Correlation; Sudden Cardiac Death; Beta-Blockers; Left Cardiac Sympathetic Denervation





OR02 | OPTIMAL LEAD PLACEMENT FOR MULTI-SITE CARDIAC RESYNCHRONIZATION THERAPY IN PEDIATRIC AND CONGENITAL HEART DISEASE

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Background and Aim:

Cardiac Resynchronization Therapy (CRT) is an effective treatment for select patients with heart failure. For pediatric and congenital heart disease patients (CHD), an epicardial system is often required; however, there is currently no clear guidance for optimal ventricular lead positioning for multisite epicardial CRT.

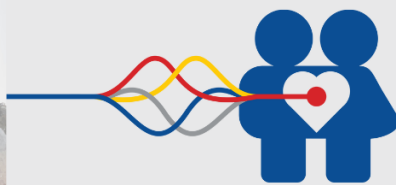
Methods:

Patients were <21 years old at implant or had CHD; had epicardial multisite system placed 2003-2024; had baseline SVEF $\leq 45\%$; and had follow up chest x-ray and echocardiogram. Lead location was determined by review of chest x-ray(s) and categorized as either apex, outflow tract (OT) or sub-systemic (sSV) or sub-pulmonic ventricle (sPV) base/free wall (FW). Accuracy and consistency of lead localization was determined via gold standard of cross-sectional imaging (n=38) and blinded independent review, respectively.

Results:

Of the 222 patients reviewed, 101 met inclusion criteria. The median age (years) was 13.1 [3.9-26.7] and 82 (81.2%) had CHD of which 12 (14.5% of CHD patients) had single ventricle physiology. Identification of lead position based on chest x-ray(s) was accurate ($\kappa = 0.86$) and reproducible (Krippendorff's alpha ($\alpha = 0.87$)). There was no significant difference in lead configuration by era.

Overall, a majority, 60 (59.4%), demonstrated an improvement in SVEF by $>10\%$ with a median absolute SVEF (%) increase of 13.5 [4.3-22.3] compared to baseline. There was no significant difference in percentage of responders ($p=0.904$) or in absolute change in SVEF between the different lead configurations ($p=0.264$). An improved response to CRT was consistently associated with a larger improvement in QRSD z-score and with a younger age on multivariable ordinal logistic regression and linear regression. See *Figure 1*.



Conclusion:

This is the first description of CRT lead placement in pediatric and CHD and identified a broad range of ventricular lead placements. There was no identifiable association between lead positioning and response to CRT. However, younger patients and patients with greater improvement in QRSd z-score were found to have an improved CRT response. This study would suggest that prioritizing shortening of QRSd rather than aiming for specific lead configuration or lead separation is more likely to result in CRT response.

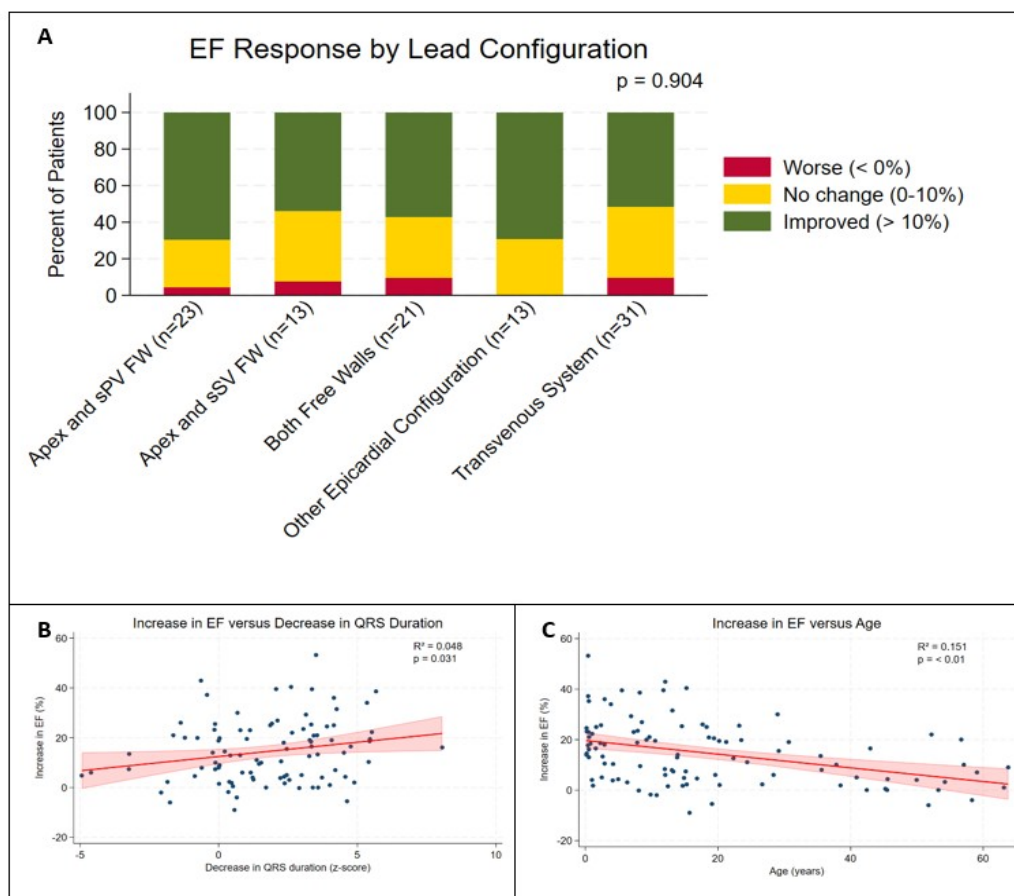
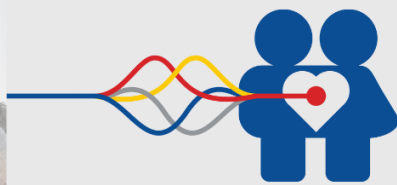


Figure 1. Predictors of response to CRT. (A) SVEF Response by lead configuration. (B) Correlation between SVEF (%) improvement and QRS narrowing. (C) Correlation between diminishing SVEF (%) response with increasing age.



OR03 | CATHETER ABLATION OF MAHAIM PATHWAYS IN CHILDREN: INSIGHTS FROM ONE OF THE LARGEST MULTICENTER COHORTS

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Background:

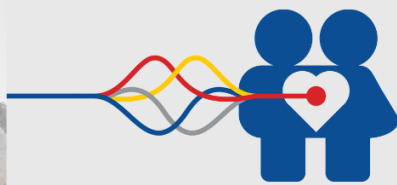
Mahaim accessory pathways are rare decrementally conducting pathways that typically support antidromic atrioventricular reentrant tachycardia and may mimic ventricular tachycardia. Contemporary multicenter pediatric data regarding their electrophysiological characteristics, associated arrhythmias, and long-term ablation outcomes remain limited.

Methods:

We retrospectively analyzed consecutive pediatric patients who underwent electrophysiological study and catheter ablation for Mahaim pathway-mediated tachycardia between January 2015 and December 2025 at three tertiary pediatric electrophysiology centers. Clinical characteristics, electrophysiological findings, pathway localization, procedural features, and long-term outcomes were evaluated.

Results:

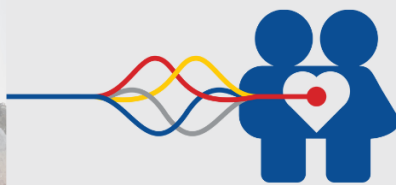
Fifty-six children (29 males, 51.8%) with a mean age of 12.9 ± 3.5 years were included. Structural heart disease was present in 19.6% of patients, most commonly Ebstein anomaly (16.1%). A history of previous failed ablation was documented in 19.6%. Antidromic Mahaim tachycardia was inducible in all patients. A discrete Mahaim potential was identified in 52 patients (92.8%). Concomitant arrhythmia substrates were present in 19 patients (33.9%), including typical atrioventricular nodal reentrant tachycardia (n=8), Wolff–Parkinson–White syndrome (n=7), concealed accessory pathways (n=3), and atrial flutter (n=1). Pathways were predominantly right-sided (98.2%), most frequently located in the right anterolateral (43.0%), right lateral (23.2%), and right posterolateral (23.2%) regions.



Radiofrequency ablation alone was used in 50.0%, cryoablation alone in 16.1%, irrigated radiofrequency ablation alone in 12.5%, and combined-energy strategies in 21.4% of patients. Long sheaths were required in 35.7%, and a transjugular approach was necessary in 7.1%. Mean procedure and fluoroscopy times were 184.7 ± 73.5 minutes and 1.6 ± 3.9 minutes, respectively; 69.6% of procedures were performed without fluoroscopy. Acute procedural success was achieved in 53 patients (94.6%). During a mean follow-up of 29.9 ± 21.8 months, recurrence occurred in five patients (9.4%), all successfully treated with repeat ablation. One major complication (right coronary artery occlusion) was successfully managed with balloon angioplasty.

Conclusions:

Mahaim pathway-mediated tachycardia in children is characterized by a high prevalence of associated arrhythmia substrates and a marked predominance of right free-wall localization. Catheter ablation guided by contemporary electroanatomical mapping provides excellent acute and long-term outcomes with minimal fluoroscopy exposure, even in patients with congenital heart disease and previous failed ablations.

**OR04 | GENETIC VARIANTS AND ARRHYTHMIAS IN CONGENITAL HEART DISEASE**

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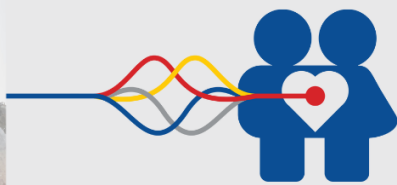
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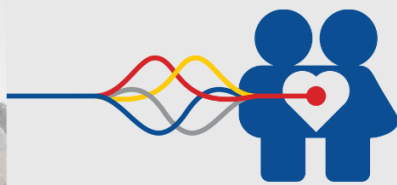
Background and Aims: Arrhythmias in patients with congenital heart disease (CHD) are traditionally attributed to hemodynamic abnormalities, surgical interventions, and myocardial scar formation. Although genetic variants are well recognized contributors to CHD, their role in arrhythmia susceptibility remains poorly understood.

Methods: We analyzed patients with CHD enrolled in the Pediatric Cardiac Genomics Consortium with exome sequencing and longitudinal arrhythmia phenotypic data. We tested associations between clinically significant arrhythmias and rare predicted damaging variants within predefined genes with dominant inheritance: arrhythmia (76 genes, 10.3 Mb), cardiomyopathy (156 genes, 20.8 Mb), CHD (245 genes, 28.7 Mb) and heterotaxy (18 genes, 0.9 Mb). Rare (gnomAD maximum population allele frequency $<1 \times 10^{-4}$) and potentially damaging (loss-of-function, predicted splice-altering (SpliceAI ≥ 0.7), or deleterious missense variants (AlphaMissense ≥ 0.56)) de novo or transmitted variants were identified.



Results: Among 8,828 patients, 1,013 (11.5%) had at least one arrhythmia requiring intervention (8.2% supraventricular tachycardia (SVT), 6.1% ventricular tachycardia (VT), 2.5% atrioventricular block (AVB), 1.7% sinus node dysfunction (SND)). At least one rare, predicted damaging variant within all pre-defined genes was found in 346 (34.2%) of patients with arrhythmia requiring intervention versus 2,319 (29.7%) of those without (OR 1.23 [95% CI 1.07, 1.41], $p=0.004$). Variants in heterotaxy (OR 1.66 [95% CI 1.14, 2.44], FDR 0.019), arrhythmia (OR 1.38 [1.13, 1.67], FDR 0.007) and CHD (OR 1.26 [95% CI 1.07, 1.50], FDR 0.015) genes were associated with arrhythmia requiring intervention, but cardiomyopathy genes were not associated (OR 1.15 [95% CI 0.98, 1.36]). Risk increased with variant burden (OR 1.20 [95% CI 1.09, 1.33] per additional variant). Associations were observed for SVT and VT, but not for AVB or SND requiring PPM.

Conclusion: Rare predicted damaging variants were common in children with CHD, and a greater burden of variants in arrhythmia, CHD, and heterotaxy genes was associated with tachyarrhythmias, supporting a role for genetic susceptibility. In contrast, bradyarrhythmias appeared to be driven predominantly by anatomic and surgical factors.



OR05 | PREDICTION OF ARRHYTHMIAS FOLLOWING CONGENITAL HEART SURGERY USING 12-LEAD ECG AND ARTIFICIAL INTELLIGENCE

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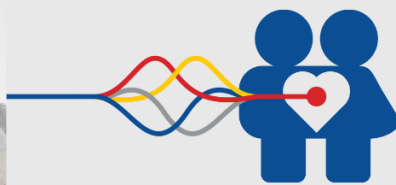
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Background and Aims: Postoperative arrhythmias occur in 7-26% of congenital heart surgery and are associated with prolonged length of stay and mortality. We evaluated whether artificial intelligence (AI) analysis of 12-lead ECGs can predict postoperative arrhythmias.

Methods: Retrospective cohort of patients undergoing CHD surgery (2016–2024). Outcomes included postoperative arrhythmias requiring medication and atrioventricular block (AVB) requiring permanent pacemaker (PPM) before discharge. ResNet convolutional neural networks were trained on preoperative and first postoperative 12-lead ECG, with penultimate layer activations used as embedding (patient-level 70%/15%/15% train/validation/test splits). Clinical variables were then integrated with the embeddings using XGBoost. ECG waveform features were extracted and SHAP analysis performed for interpretability.

Results: Among 5,704 surgeries (age 2.4 [IQR 0.4, 8.8] years, 54% male), arrhythmias requiring medication occurred in 584 (10%) and AVB requiring PPM in 108 (2%). Using ECG alone, the postoperative ECG predicted arrhythmia with an AUROC of 0.72 (95% CI 0.66–0.77), whereas the preoperative ECG had limited value (AUROC 0.63, 95% CI 0.56–0.68). Preoperative ECG predicted AVB requiring PPM with an AUROC of 0.73 (95% CI 0.62–0.82). For arrhythmia, adding clinical variables improved performance (AUROC 0.76, 95% CI 0.72–0.81; DeLong 0.003), with ECG embeddings accounting for 71% of model importance (postoperative 49%, preoperative 22%) and clinical variables 29%. For AVB requiring PPM, adding clinical variables did not improve performance (AUROC 0.74, 95% CI 0.61–0.87; DeLong 0.49). Of 1,647 ECG features, 296 (18%) differed significantly for arrhythmia and 149 (9%) for AVB requiring PPM (FDR $q < 0.05$). Features associated with arrhythmia included prolonged QT interval, reduced R-wave amplitude, and altered waveform skewness in leads V4–V6. Features associated with AVB requiring PPM included increased terminal force, QRS area, and P-wave amplitude in V3–V6.



Conclusion: Children who develop postoperative arrhythmias following CHD surgery exhibit distinct abnormalities in conduction and repolarization on both preoperative and early postoperative ECGs. AI-enabled ECG analysis with clinical variables predicted arrhythmia and AVB requiring PPM with moderate performance, with ECG embeddings contributing most. Prospective external validation, prediction of individual arrhythmia subtypes, and continuous telemetry data may further refine performance.

A. Arrhythmia requiring medication



B. Atrioventricular block requiring pacemaker

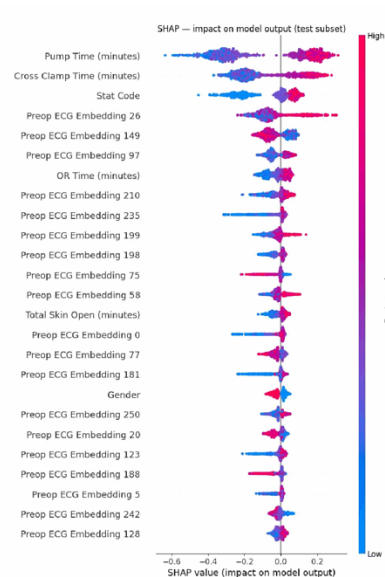
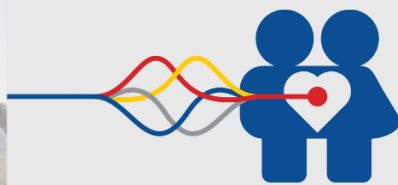


Figure. SHapley Additive exPlanations (SHAP) impact on model output (per-feature) for each integrated model; each point is a patient, colored by feature value (red = high, blue = low). A, arrhythmia requiring medication; B, Atrioventricular block requiring pacemaker

**OR06 | GENOTYPE-SPECIFIC QTc TRAJECTORIES IN NEONATES WITH SUSPECTED LONG QT SYNDROME**

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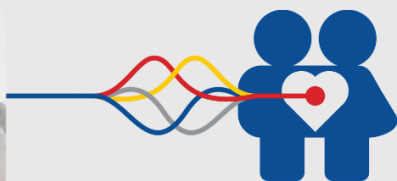
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Background and Aim: The optimal timing of the initial electrocardiogram (ECG) in neonates with suspected Long QT Syndrome (LQTS) remains debated. Early ECGs may capture transient-QTc prolongation, while delayed screening risks missing critically prolonged intervals warranting immediate intervention. We aimed to assess the diagnostic value of QTc<48 hours in predicting genotype positivity by evaluating QTc changes between the first and second neonatal ECG.

Methods: We retrospectively analyzed 98 neonates with suspected LQTS. Genotype distribution: LQTS1 (n=18), LQTS2 (n=19), LQTS3 (n=12), genotype-negative (n=36), and other (n=13). The first ECG was obtained <48 hours of birth. The second ECG was targeted at two weeks of age; when unavailable, the nearest subsequent ECG within the first three months of life was used. QTc intervals were measured at both timepoints. Linear mixed models with random intercepts for patients were fitted to assess QTc change.

Results: Mean QTc decreased from 481±76 to 456±47ms between the first and second ECG. The largest reductions were observed in transient QT prolongation ($\Delta=-115$ ms) and LQTS1 ($\Delta=-68$ ms). LQTS3 showed minimal change ($\Delta=-8$ ms, Time×Genotype $\beta=+51.0$, $p=0.006$). Beta blocker (BB) treated neonates (n=26) demonstrated greater QTc reduction ($\Delta=-68$ ms vs. -24ms; Time×BB $\beta=-46.4$, $p<0.001$), with a more pronounced decrease in LQTS1 (541±30 to 461±27ms, $\Delta=-80$ ms) than LQTS2 (534±38 to 486±52ms, $\Delta=-47$ ms). BB initiation in LQTS2 (10/14) was driven by QTc≥500ms and/or severe fetal phenotype. Six neonates presented with 2:1 atrioventricular block (2 LQTS2, 1 LQTS3, 2 Timothy, 1 genotype elusive), one with Torsade de Pointes. Despite these reductions, the second ECG QTc remained longer in both groups than in genotype-negative (420 ms) neonates. ROC analysis of the first ECG QTc discriminated LQTS1 (AUC 0.985) and LQTS2 (0.987) from genotype-negative neonates with high accuracy, but performed less well for LQTS3 (0.848) (Figures 1 and 2).



Conclusions: First ECG QTc <48 hours of birth discriminates genotype-positive from genotype-negative neonates with high accuracy for LQTS1 and LQTS2, supporting the value of early screening and providing an evidence base for postnatal management protocols. LQTS3 remains a diagnostic challenge due to lower baseline QTc, requiring genotype-guided surveillance. Disentangling BB efficacy from genotype-specific QTc trajectories requires larger within-genotype cohorts.

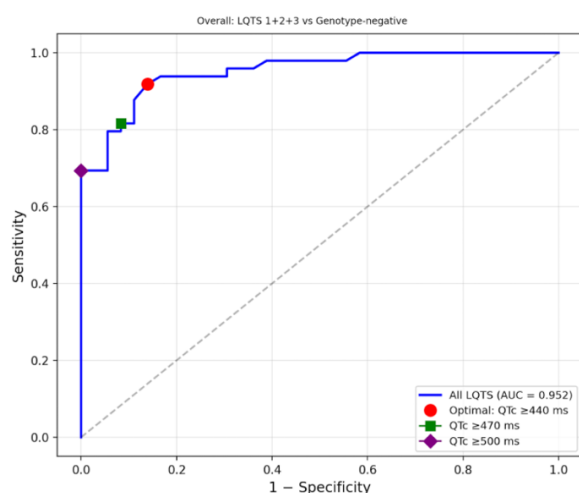


Figure 1: ROC curve for the first ECG QTc (<48 hours) discriminating all LQTS subtypes from genotype-negative neonates (AUC 0.952).

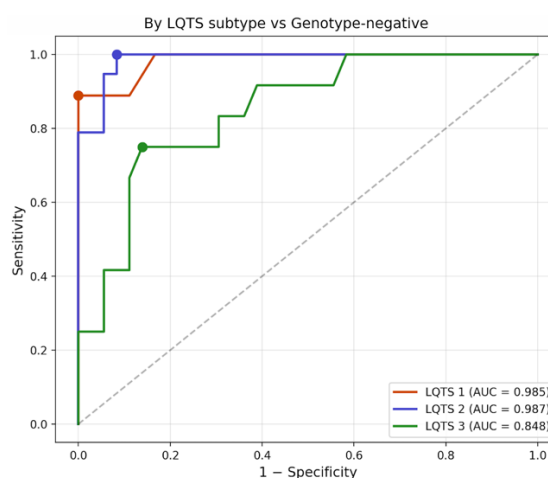
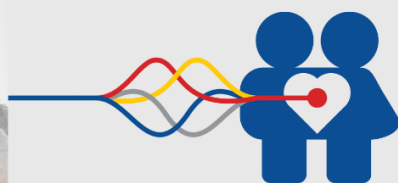


Figure 2: Subtype-specific ROC curves. LQTS type 1 (AUC 0.985; optimal cutoff ≥498 ms) and LQTS type 2 (AUC 0.987; optimal cutoff ≥475 ms) LQTS type 3 (AUC 0.848; optimal cutoff ≥440 ms)

**OR07 | LEFT VENTRICULAR ISOVOLUMETRIC RELAXATION TIME IN FETAL LONG-QT SYNDROME**

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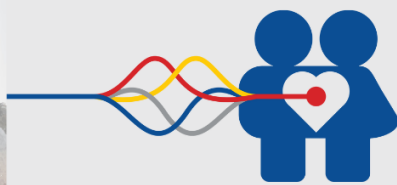
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Background:

Long QT syndrome (LQTS) is an inherited channelopathy characterized by prolonged cardiac repolarization, predisposing to life-threatening arrhythmias. The clinical diagnosis relies on electrocardiography, which is not currently feasible in the fetus, making non-invasive prenatal detection challenging. Previous studies showed prolonged left ventricular isovolumetric relaxation time (LVIRT) and sinus bradycardia in LQTS fetuses. We aimed to evaluate LVIRT and fetal heart rate (FHR) differences between LQTS and controls in a larger cohort and among LQTS subgroups, and their utility for risk stratification.

Methods:

This multicenter study included retrospective (28%) and prospective (72%) data from at risk fetuses. Pulsed Doppler measured LVIRT, cycle length (CL), and FHR, were stratified by gestational age (GA) (≤ 20 , 21–30, ≥ 30 weeks). LVIRT was indexed to cycle length (IRT/CL) to account for FHR effects. ROC curves assessed diagnostic performance of IRT/CL, FHR, and their combination. LQTS-positive fetuses were stratified by arrhythmia (Torsades de Pointes/Ventricular Tachycardia or AV-block) and presentation (index vs. familial).



Results:

104 fetal echocardiograms from 62 LQTS patients were compared with 53 from 29 genotype-negative family members and 703 from 467 controls. IRT/CL was significantly higher in LQTS fetuses across all GA groups (≤ 20 weeks: 13.4 ± 3 vs 10.7 ± 1.7 , $p=0.0006$; 21–30 weeks: 13.1 ± 2.5 vs 11.5 ± 1.6 , $p=0.0007$; ≥ 30 weeks: 13.1 ± 2.7 vs 11.2 ± 1.9 , $p<0.0001$). IRT/CL combined with FHR provided the best diagnostic performance (figure 1).

Within LQTS-positive fetuses, median LVIRT/CL was significantly higher in those with arrhythmia (21–30 weeks: 18.05 [15.13 – 18.73] vs 12.05 [11.12 – 13.17], $p=0.0010$; ≥ 30 weeks: 15.16 [14.17 – 16.09] vs 12.39 [11.25 – 13.66], $p=0.0046$). FHR showed no consistent pattern. Index versus familial comparison suggests FHR may be a stronger differentiator than IRT/CL.

Conclusion:

Fetuses with LQTS have consistently elevated IRT/CL across all GA groups. IRT/CL combined with FHR provides the best diagnostic performance and may help identify fetuses at highest risk for arrhythmia. The effect of maternal betablocker (39%) as a potential confounder still needs to be analyzed. Due to the low arrhythmia rate ($<10\%$) and few index cases (18%), data from recently published cohorts will be included to further validate these findings.

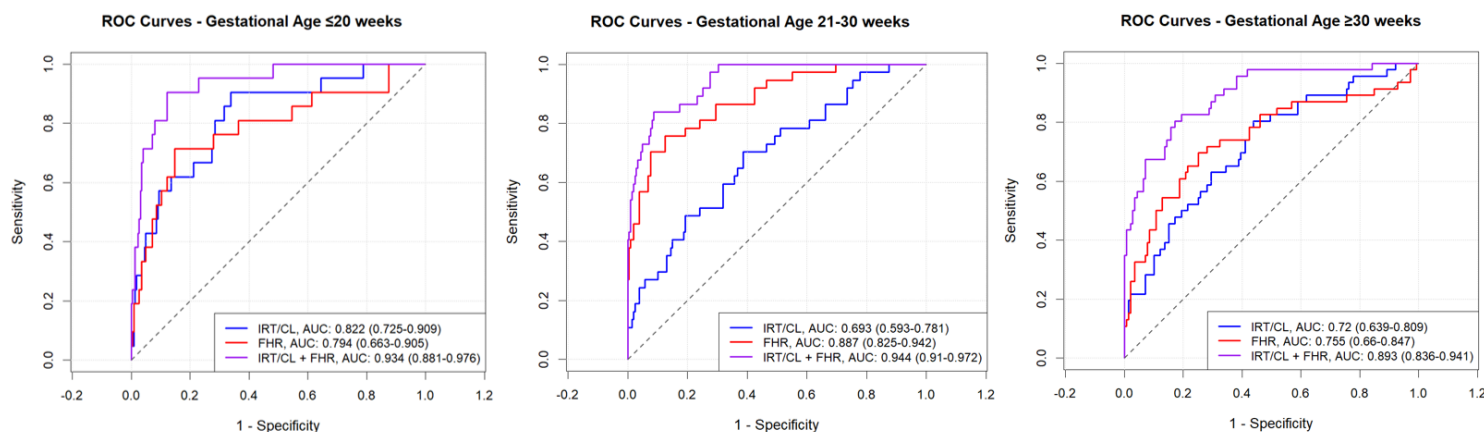
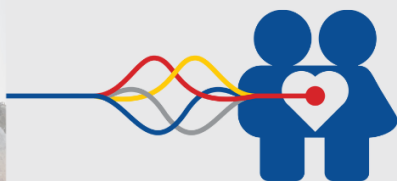


Figure 1. Receiver operating characteristic (ROC) curves stratified by gestational age.



OR08 | INTERNATIONAL PEDIATRIC BRUGADA REGISTRY: A PHENOTYPICAL REVIEW OF THE COHORT AND EVALUATION OF CURRENT METHODS FOR INDIVIDUAL RISK STRATIFICATION

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Background and aim:

The paediatric Brugada syndrome (BrS) is a rare genetic condition that predisposes to dangerous cardiac arrhythmia and sudden death. Its underlying genetic substrate remains incompletely understood. *SCN5A* loss-of-function variants are a recognized driver of disease pathogenesis. However, the clinical spectrum of cardiac expression is highly variable, and individual risk assessment remains challenging.

We aim to describe the spectrum of disease expression in young patients worldwide and to assess the performance of the existing paediatric risk score.

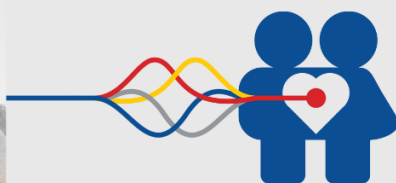
Methods:

We describe a cohort of young patients with sodium channelopathy. For this, we use the data of the International Pediatric Brugada Registry, which includes patients diagnosed via either type 1 Brugada ECG or pathogenic familial *SCN5A* variant by the age of 20 years.

Life-threatening arrhythmic events (LAEs) were defined as (aborted) sudden cardiac death ((a)SCD), documented VF and/ or appropriate ICD shock. Discriminatory performance of the score was assessed using sensitivity and specificity.

Results:

A total of 340 young patients were analysed. First degree AV-block was present in 10% (n=34), sinus node dysfunction (SND) in 5% (n=18) and atrial arrhythmia in 2% (n=7). About one third (n=120, 35%) had symptoms of possible cardiac origin, and 8.8% (n=30) experienced LAEs; of these, 13 were (a)SCD, 3 were documented VF and 14 were appropriate ICD shocks. Spontaneous type 1 ECG (OR 1.6), syncope (OR 2.4) and sinus node dysfunction (OR 4.2) showed association with LAE.



The proposed paediatric risk score showed a sensitivity of 60% and a specificity of 89% for predicting LAE at initial evaluation. Exclusion of patients with overt high-risk features resulted in significant reduction in the model's predictive performance.

Conclusion:

We describe a very heterogeneous paediatric population with a high rate of conduction abnormalities and SND. Most patients are asymptomatic, and our findings suggests that a combination of ECG markers may help define clinical risk. However, LAEs can occur without preceding symptoms or ECG abnormalities, which makes individual risk stratification extremely difficult.

As previously documented for proposed adult risk scores, the paediatric scoring model equally falls short in reliably identifying individuals at risk of LAE without overt high-risk features.



OR09 | POLYGENIC QTc BURDEN MODIFIES EARLY-LIFE PENETRANCE OF CONGENITAL LONG QT SYNDROME IN FINNISH FOUNDER VARIANT CARRIERS

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Background and Aim

The clinical expressivity of LQTS is highly variable ranging from concealed and asymptomatic disease to a malignant form with torsades de pointes leading to syncope or sudden cardiac death. The highly variable penetrance of pathogenic LQTS variants is poorly understood. Polygenic variation modifies the corrected QT-interval (QTc), and higher QTc increases the risk of severe arrhythmic events. However, whether polygenic QTc burden modifies LQTS event penetrance remains unclear. Our aim was to assess whether QTc polygenic risk score (PRS) modifies lifetime LQTS event penetrance in Finnish founder variant carriers.

Methods

We used FinnGen participants genotyped for Finnish founder variants KCNQ1 G589D, KCNH2 R176W, and KCNH2 L552S (N=418,310). We ascertained LQTS events (1970—) from electronic health registers. Early-life analysis from birth to age 30 included those with complete LQTS event follow-up (N=140,298). We used cox proportional hazards (CPH) to model the joint effect of founder variant carrier status and QTc PRS on early-life LQTS event risk. Follow-up ended at a severe LQTS event, death, or end of register follow-up (latest August 2024). We included CPH interactions between QTc PRS and carrier status. QTc PRS was based on 1,110,494 variant weights from UK Biobank (PGS catalog PGS002276).

Results

The study population is presented in Table 1. One standard deviation increase in QTc PRS was associated with 50% higher LQTS event risk. The associations were carrier-specific (statistical interaction) with no PRS effect in non-carriers (HR 1.01; 95% CI 0.99-1.03)). Carriers with QTc PRS >90% had 207% higher LQTS event risk (HR 3.07; 95% CI 1.81-5.21) while carriers with PRS 10-90% had 52% higher LQTS event risk (1.52; 1.11–2.08) relative to non-carriers (Table 2 and Figure 1).



Conclusion

We observed that QTc PRS stratifies early-life LQTS penetrance in carriers, but not non-carriers of Finnish LQTS founder variants. QTc PRS may help stratify lifetime congenital LQTS penetrance in pathogenic variant carriers.

Group	N	Female (%)	Median follow-up (% complete)	Person years	Number of LQTS events / total	Event rate per 1000 person years
Carrier & PRS > 90%	46	29 (63.0)	30 (78.3)	1,267	6 (46)	4.7
Carrier & PRS 10-90%	430	288 (67.0)	30 (80.9)	12,074	36 (430)	3.0
Population	111,808	76,620 (68.5)	30 (80.3)	3,197,386	5,453 (111,808)	1.7
Carrier & PRS < 10%	74	48 (64.9)	30 (89.2)	2,143	0 (74)	0

Table 1. Study population characteristics

Group	HR (95% CI)	P value	Risk by age 30 % (95% CI)
Carrier & PRS > 90%	3.07 (1.81–5.21)	3.38×10^{-5}	12.5 (6.06–18.5)
Carrier & PRS 10-90%	1.52 (1.11–2.08)	9.52×10^{-3}	6.40 (4.40–8.36)
Population	1.00	-	4.26 (4.01–4.51)
Carrier & PRS < 10%	0.76 (0.39–1.50)	0.43	3.27 (1.07–5.41)

Table 2. Cox proportional hazards analysis of study cohort

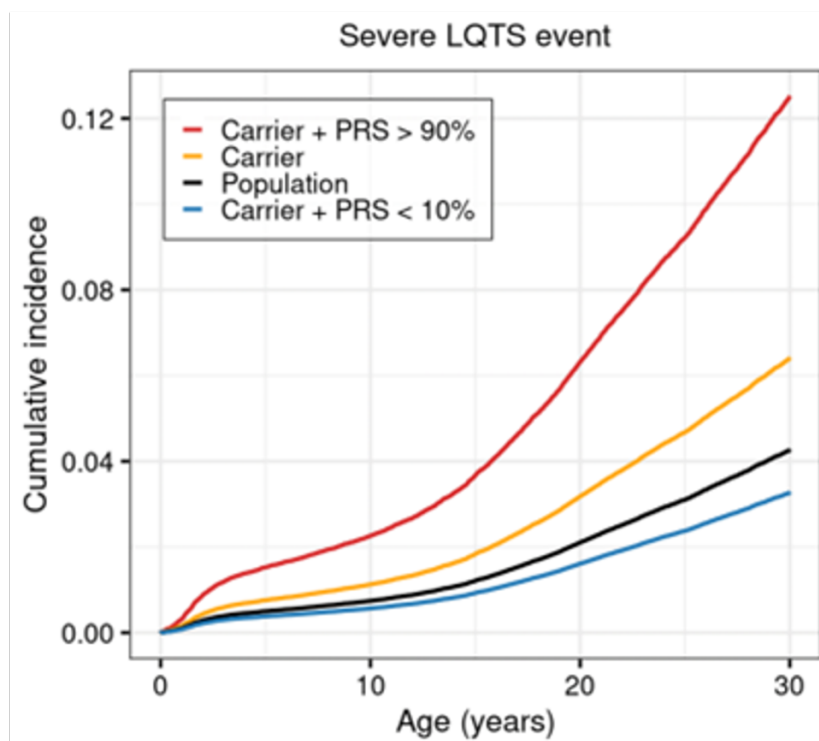


Figure 1. Polygenic risk modifies the early-life penetrance of LQTS founder mutations. Adjusted survival curves showing long QT syndrome (LQTS) event risk stratified by pathogenic founder variant carrier status and QTc polygenic risk score (PRS) categories. Adjusted survival curves were derived from a CPH model including an interaction term between continuous QTc PRS and founder variant carrier status.



OR10 | LONG-TERM OUTCOMES AND APPROPRIATE ICD THERAPIES IN PEDIATRIC HCM: ARE WE MISSING HIGH-RISK PATIENTS?

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Background: Implantable cardioverter defibrillators (ICDs) are recommended for sudden cardiac death prevention in selected children with hypertrophic cardiomyopathy (HCM). However, pediatric data on long-term efficacy and device-related complications remain limited.

Methods: We retrospectively analyzed children with HCM undergoing ICD implantation at a tertiary center. Appropriate therapies were defined as ICD shocks delivered for ventricular tachycardia (VT) or ventricular fibrillation (VF). The predictive performance of the HCM Risk-SCD score was evaluated using receiver operating characteristic analysis. Progression of hypertrophy was assessed as the difference between baseline and follow-up left ventricular maximal wall thickness z-scores.

Results Twenty-eight children underwent ICD implantation at a median age of 13 years. Twenty-six patients (93%) received an ICD for primary prevention and 2 (7%) for secondary prevention. Ten patients received transvenous ICDs and eighteen subcutaneous ICDs. Median follow-up was 3.8 years (IQR 1.4–5.7).

Seven patients (25%) experienced appropriate ICD therapies. Five episodes were triggered by VF and two by sustained VT. Patients receiving appropriate therapies were younger at implantation (11.0 vs 15.0 years, $p=0.042$), despite longer follow-up duration (7.3 vs 2.3 years, $p=0.021$). Kaplan-Meier analysis demonstrated a significantly higher incidence of appropriate ICD therapies among secondary prevention patients (log-rank $p=0.0005$). Of note, both patients implanted for secondary prevention exhibited exercise-induced ECG abnormalities and advanced diastolic dysfunction, whereas their estimated HCM Risk-Kids score was $<6\%$.

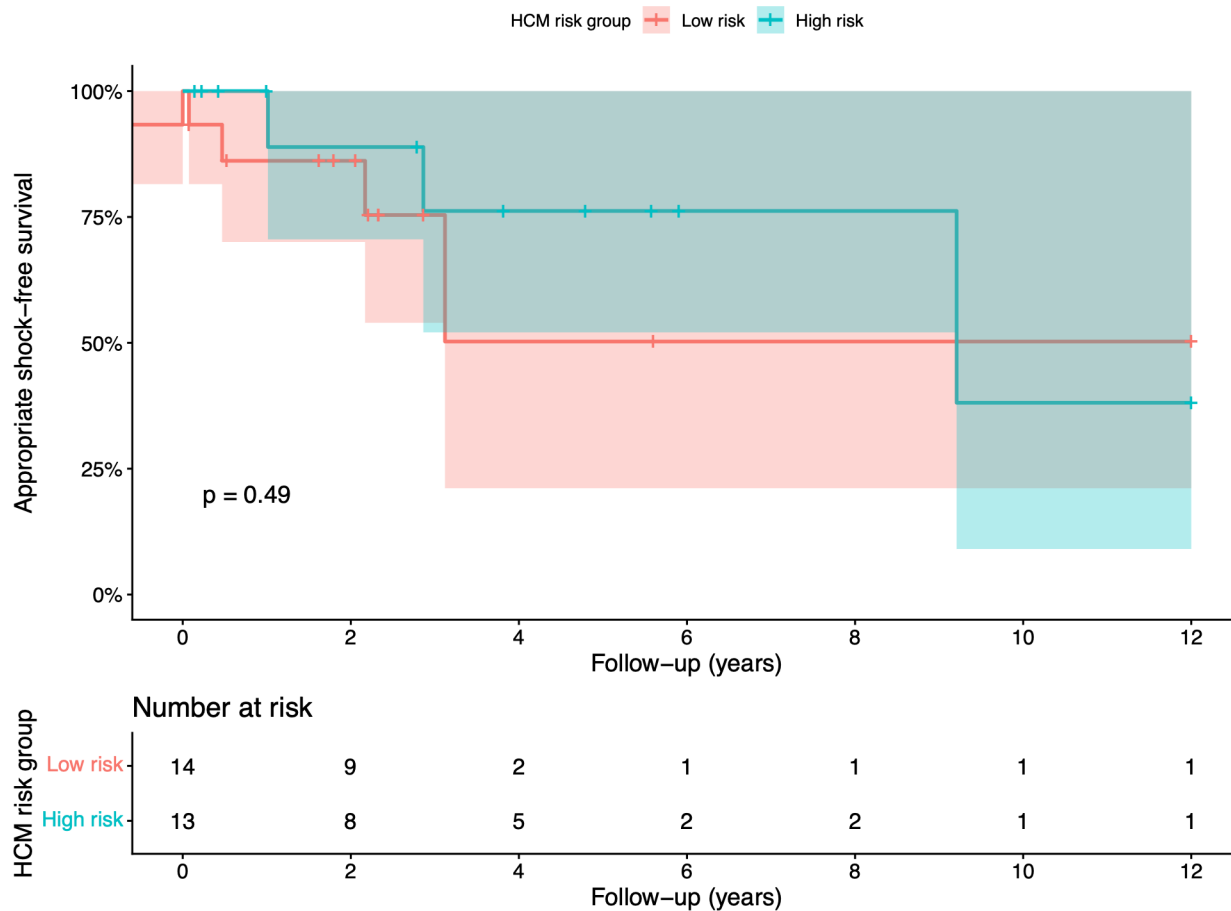


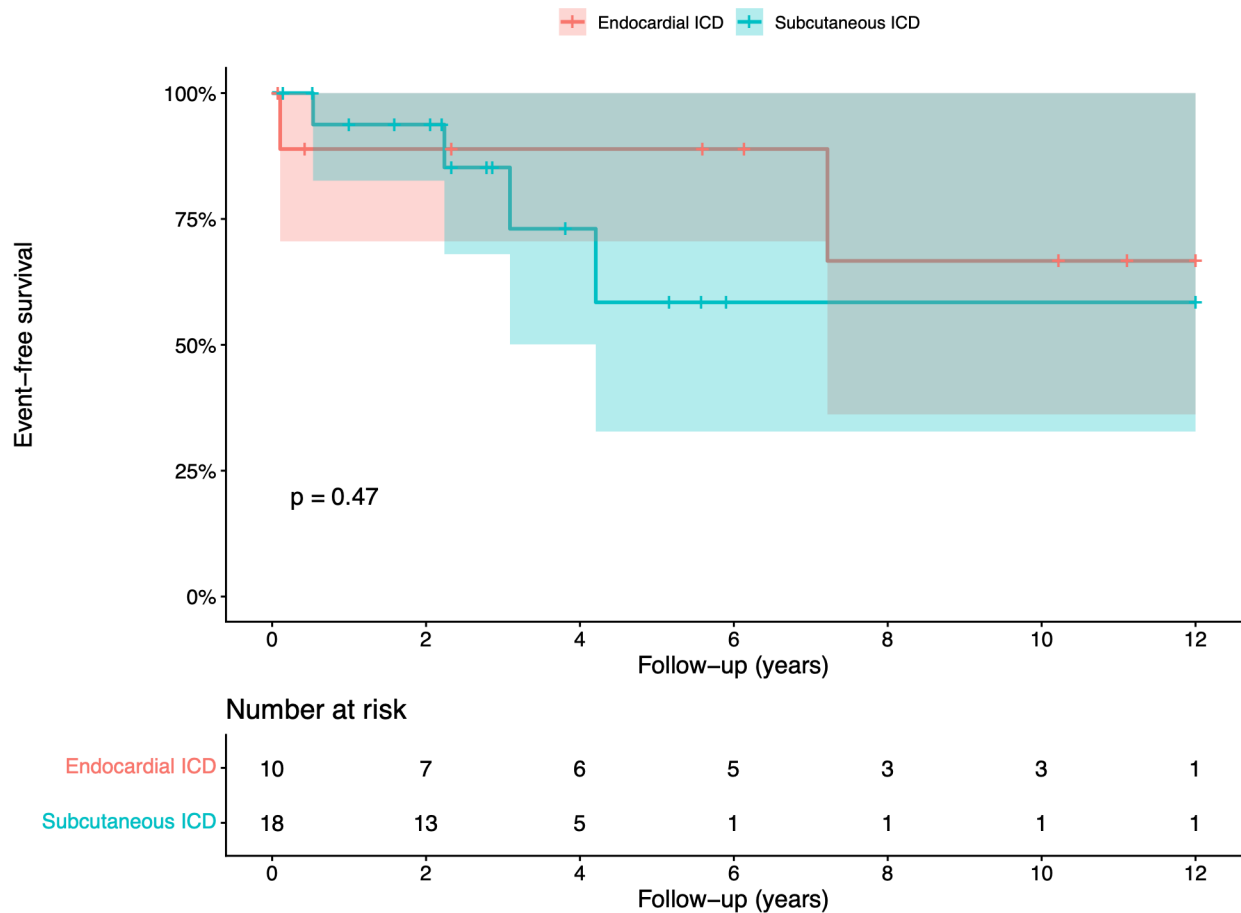
The HCM Risk-Kids score showed limited ability to identify patients experiencing appropriate therapies (AUC 0.50, 95% CI 0.22–0.78) and freedom from appropriate shocks did not differ between patients with HCM Risk-Kids $\geq 6\%$ and $< 6\%$ (log-rank $p=0.49$). Device-related complications occurred in six patients (21%), including five inappropriate shocks and one infection with erosion requiring reintervention. Complication-free survival was not significantly different between transvenous and subcutaneous ICD recipients (log-rank $p=0.47$). Progression of hypertrophy, assessed by changes in left ventricular wall thickness z-score over time, was not associated with device-related complications ($\beta = -0.04$, $p=0.54$)

Conclusion ICDs provided potentially life-saving therapies in 25% of our cohort. Guideline-recommended risk stratification tools seem to show limited ability to predict appropriate ICD interventions. The presence of ischemic ECG abnormalities and advanced diastolic dysfunction may provide incremental information beyond conventional risk models.



Variable	No appropriate shock (n=21)	Appropriate shock (n=7)	P value
Male sex, n (%)	15 (71.4)	3 (42.9)	0.362
Symptoms, n (%)			0.017
• None	20 (95.2)	4 (57.1)	
• Syncope	0 (0.0)	2 (14.3)	
• Neurally mediated syncope	1 (4.8)	0 (0.0)	
Height, cm	158.0 [150.0–166.0]	150.5 [146.8–155.0]	0.220
Weight, kg	52.0 [45.0–70.0]	41.0 [36.0–52.0]	0.089
Age at implant, years	15.0 [12.0–16.0]	11.0 [10.5–12.5]	0.042
Subcutaneous ICD, n (%)	15 (71.4)	3 (42.9)	0.362
Fluoroscopy time, min	1.05 [0.0–8.55]	9.0 [5.8–12.5]	0.198
Procedure time, min	91.5 [80.0–109.3]	105.0 [90.0–120.0]	0.684
ICD complication, n (%)	4 (19.0)	2 (28.6)	1.000
Pocket infection, n (%)	1 (4.8)	0 (0.0)	1.000
Lead erosion, n (%)	1 (4.8)	0 (0.0)	1.000
Inappropriate shock, n (%)	3 (14.3)	2 (28.6)	0.776
HCM Risk-Kids score at implant	8.7 [7.6–10.7]	9.0 [6.4–10.8]	1.000
HCM Risk-Kids score at follow-up	6.3 [4.6–10.2]	9.0 [7.7–9.9]	0.309
Maximal wall thickness at implant, mm	24.0 [21.0–27.0]	23.0 [16.5–27.0]	0.440
LV wall thickness z-score at implant (Boston)	11.0 [7.7–12.7]	15.0 [10.1–19.5]	0.301
Maximal wall thickness at follow-up, mm	27.0 [24.8–30.0]	26.5 [24.3–31.0]	0.927
LV wall thickness z-score at follow-up (Boston)	11.0 [9.7–13.5]	13.5 [7.9–18.2]	0.567
Left atrial diameter at implant, mm	32.0 [28.0–37.0]	29.0 [28.0–33.5]	0.506
Left atrial diameter z-score at implant (Detroit)	1.33 [0.95–2.21]	1.00 [0.88–2.15]	0.657
NSVT at implant, n (%)	6 (28.6)	3 (42.9)	0.815
Family history of SCD, n (%)	12 (60.0)	4 (57.1)	1.000
Syncope at implant, n (%)	1 (4.8)	1 (14.3)	1.000
LGE at implant, n (%)			0.662
• Absent	2 (10.0)	0 (0.0)	
• Present	14 (70.0)	2 (100.0)	
• Extensive	4 (20.0)	0 (0.0)	
End-stage disease at implant, n (%)	1 (4.8)	1 (14.3)	1.000
Apical aneurysm at follow-up, n (%)	0 (0.0)	1 (14.3)	0.151
High HCM risk score , n (%)	10 (47.6)	3 (42.9)	1.000
Massive HCM, n (%)	7 (33.3)	4 (57.1)	0.503





POSTER PRESENTATIONS

**PO.01 | ELECTROMECHANICAL INTERACTION AND EFFECT OF RIGHT VENTRICULAR CARDIAC RESYNCHRONIZATION THERAPY IN PATIENTS WITH EBSTEIN'S ANOMALY UNDERGOING CONE REPAIR**

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Introduction: Ebstein patients after Cone repair frequently have right bundle branch block and decreased right ventricular (RV) function. We sought to study ventricular electromechanical interaction and potential benefit from RV cardiac resynchronization therapy (CRT).

Methods: Cross-sectional single center study in 23 pts after Cone repair aged median (IQR) 6.4 (4.1-10.4) yrs at surgery and 11.2 (6.2-17.2) yrs at evaluation. Permanent RV CRT was applied in 13/23 to improve RV function. Ultra-high frequency ECG (UHF ECG) and speckle tracking echocardiography (STE) were used to detect local ventricular electrical activation and peak segmental contraction timing from each RV and left ventricular (LV) segment and to quantify the electromechanical dyssynchrony at baseline and with RV CRT temporarily off and on.

Results: At baseline all pts. showed right bundle branch block with a mean (SD) QRS duration of 140 (21) ms. UHF ECG visualized local electrical activation timing in all RV and LV segments. There was a significant septal/LV to RV free wall (RVFW) electrical and mechanical delay of mean (SD) 73 (26) and 134 (97) ms, respectively, accompanied by cumulative pre-stretch (median 7.9, IQR 5.0-11.6 %) and post-systolic strain (median 3.4, IQR 1.2-6.6 %) in late activated RVFW segments with a linear correlation of time to peak segmental electrical and mechanical RV activation ($R=0.482$, $p<0.001$). RV CRT (N=13), as compared to baseline, carried an immediate improvement in RV metrics and electromechanical synchrony (Figure) without deteriorating the LV function (Tables and Images).

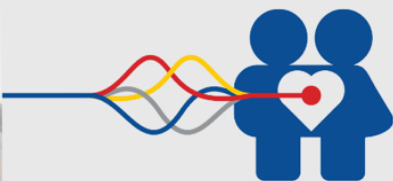


Conclusion: Ebstein patients after Cone repair exhibit significant RV electromechanical dyssynchrony detectable by UHF ECG and speckle tracking echocardiography. UHF ECG allows for accurate detection of local RV electrical activation and presents a simple method of myocardial viability and electrical dyssynchrony assessment. RV CRT improves RV electromechanical coordination, global systolic function and contraction efficiency and may be used as an adjunctive therapy of RV dysfunction.

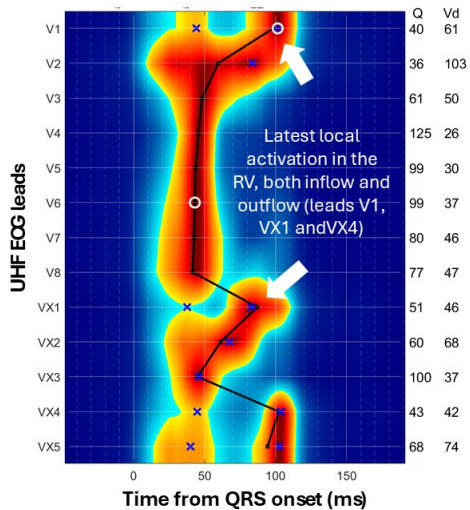
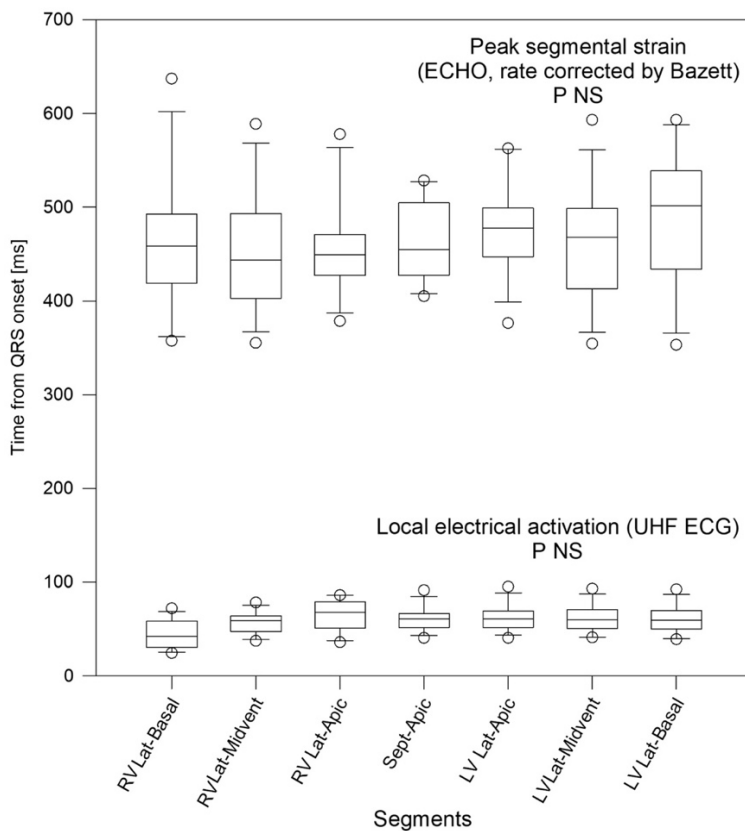
Table: N=13 with RV-CRT

	QRS [ms]	RVA FS [%]	TI dP/dt [mmHg/s]	RV MPI	LV-RV ED [ms]	LV-RV MD [ms]	RV SSF	RVFW PS [%]	RVFW PSS [%]	LV EF [%]	LV MPI
RV CRT off	146 (17)	26.3 (9.0)	214 (33)	0.30 (0.15)	78 (29)	167 (115)	0.36 (0.17)	8.0 (6.0-14.7)	3.4 (1.8-6.0)	61.0 (6)	0.42 (0.18)
RV CRT on	107 (16)	29.9 (9.9)	419 (165)	0.19 (0.09)	-8 (12)	-81 (60)	0.16 (0.14)	2.9 (0.7-5.4)	0.0 (0.0-0.7)	63.8 (6.2)	0.40 (0.13)
P	<0.001	<0.001	=0.007	=0.002	<0.001	<0.001	<0.001	= 0.001	<0.001	=0.010	NS

Mean (SD) or median (IQR). ED, electrical delay; EF, ejection fraction; MD, mechanical delay; MPI, myocardial performance index; PS, pre-stretch; PSS, post-systolic strain; RVA FS, RV area fractional shortening; RVFW, right ventricular free wall; TI, tricuspid insufficiency; SSF, systolic stretch fraction.

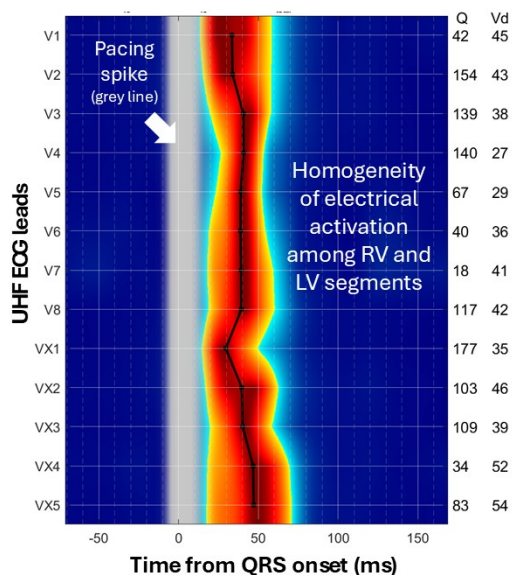
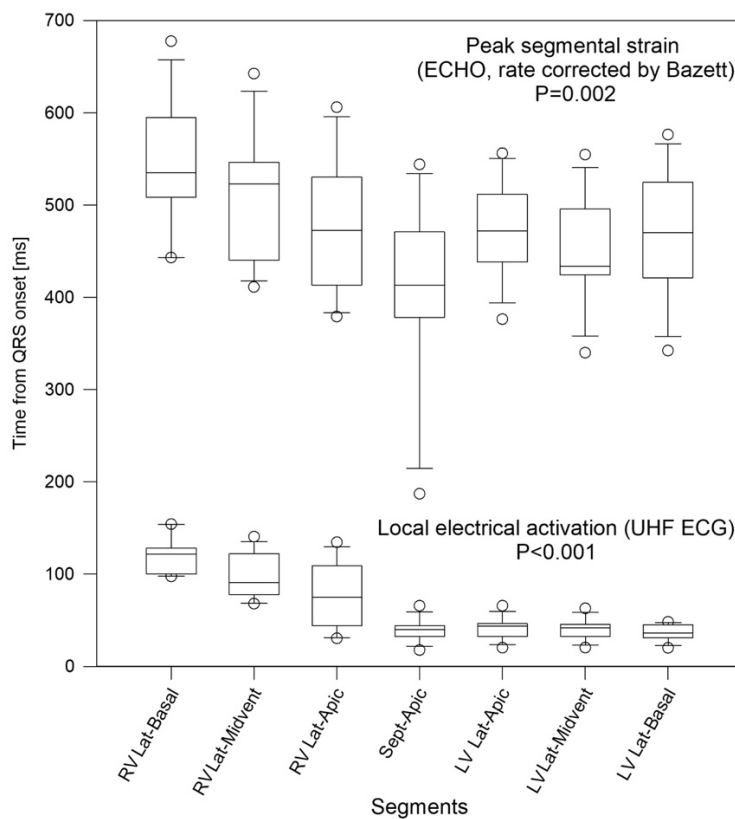


Electromechanical interaction RV CRT on





Electromechanical interaction RV CRT off





PO.02 | OPEN WINDOW MAPPING FOR ACCESSORY PATHWAY LOCALIZATION IN PEDIATRIC WOLFF-PARKINSON-WHITE SYNDROME: FIRST CASE SERIES FROM KAZAKHSTAN

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Background:

Wolff-Parkinson-White (WPW) syndrome is characterized by the presence of an accessory pathway (AP) capable of antegrade or retrograde conduction, predisposing patients to supraventricular tachyarrhythmias. Accurate localization of the AP is crucial for successful catheter ablation. The Open Window Mapping (OWM) technique is a high-resolution mapping approach that allows comprehensive visualization of activation propagation across the atrioventricular (AV) annulus.

Objective:

To present the first case series from Kazakhstan demonstrating the feasibility and efficacy of OWM for accessory pathway localization and ablation in pediatric WPW patients.

Methods:

Three pediatric patients (mean age 15 years; 66.7% female) with manifest WPW syndrome underwent catheter ablation guided by the OWM technique using EnSiteX and Carto3 systems. Mapping was performed during sinus rhythm or retrograde pacing under local anesthesia with sedation. The timing and location of the earliest AV conduction were identified to guide targeted radiofrequency ablation.

Results:

OWM enabled precise localization of the accessory pathway in all three patients (100% success). The identified AP sites included one left posteroseptal, one right posteroseptal, and one left posterolateral pathway. The mean fluoroscopy time was 3 ± 2 minutes. There were no procedural complications. During a 12-month follow-up, no arrhythmia recurrence or delta-wave reappearance was observed.

Conclusion:

This is the first reported experience with OWM for WPW ablation in Kazakhstan. The technique proved safe, effective, and capable of providing superior spatial resolution compared with conventional mapping. Integration of OWM into clinical practice may improve ablation outcomes in complex or pediatric WPW cases, especially in developing electrophysiology centers.



Keywords: open window mapping; accessory pathway; catheter ablation; electrophysiology



Accessory Pathway Localization and Ablation Outcomes

Age	Sex	AP localization	Fluoroscopy Time (min)	Recurrence (1 year)
16	F	Left posteroseptal	2	No
14	M	Right posteroseptal	5	No
15	F	Left posterolateral	2	No



Procedural Outcomes

Parameter	Result
Acute success rate	100% (3/3)
Mean fluoroscopy time	3 ± 2 minutes
Complications	None

**PO.03 | ELECTROCARDIOGRAPHIC MARKERS OF SUDDEN UNEXPECTED DEATH RISK IN PEDIATRIC EPILEPSY: A COMPARATIVE STUDY OF GENERALIZED AND FOCAL SEIZURES**

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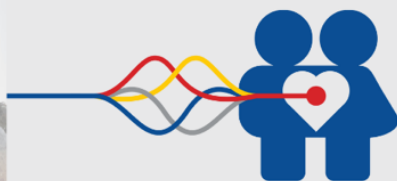
Background/Objectives: Sudden unexpected death in epilepsy (SUDEP) is a major cause of mortality in pediatric epilepsy. Cardiac arrhythmias, possibly reflected by electrocardiographic (ECG) abnormalities, are thought to contribute significantly to SUDEP risk. This study aimed to evaluate ECG indices associated with an increased risk of both atrial and ventricular arrhythmias and sudden cardiac death in pediatric patients with generalized and focal seizures, excluding those with underlying channelopathies. Materials and

Methods: Pediatric patients aged 0–18 years with generalized or focal epilepsy followed at our center between October 2024 and April 2025 were enrolled. Comprehensive cardiac evaluations, including echocardiography and 12-lead ECG, were conducted. Patients with channelopathies, structural heart defects, or significant congenital heart disease were excluded. ECG parameters—QT dispersion (QT Disp), corrected QT interval (QTc), QTc dispersion (QTc Disp), P-wave dispersion (P Disp), and T peak-T end interval (Tp-e)—were analyzed across epilepsy subgroups and compared to healthy controls. Effects of antiepileptic drug (AED) use and gender were also assessed.



Results: A total of 151 participants were included (generalized: $n = 51$; focal: $n = 50$; controls: $n = 50$). QTc and Tp-e intervals were prolonged in both epilepsy groups compared to controls ($p = 0.001$ and $p = 0.036$, respectively), however, they fell within the conventional parameters. AED use was associated with further prolongation of QTc ($p = 0.035$) and Tp-e ($p = 0.037$), these metrics were similarly found to be within the established normative boundaries. Phenobarbital and lamotrigine users showed the longest QTc, albeit not statistically significant. Males with generalized seizures had longer maximum P-wave duration (P Max) than females ($p = 0.009$). A moderate correlation was found between Tp-e and QTc ($r = 0.557$, $p = 0.001$).

Conclusions: Although there are findings in our study that may suggest a relationship between SUDEP and arrhythmia according to electrocardiographic markers associated with arrhythmia risk, larger and prospective studies with long-term follow-up are needed in the future.



PO.04 | CARDIONEUROABLATION AS A TREATMENT OF CHOICE IN CHILDREN WITH VASOVAGAL SYNCOPE AND SYMPTOMATIC BRADYARRHYTHMIAS: EFFICACY AND SAFETY

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1) Morozovskaya Children's City Clinical Hospital

2) I.V. Davydovsky City Clinical Hospital of the Moscow Department of Health

3) Patrice Lumumba Peoples' Friendship University of Russia

The authors of the study declare no conflict of interest.

Background and Aim: cardioneuroablation (CNA) is nowadays considered a promising treatment for patients, including children, with vasovagal syncope and/or symptomatic bradycardia caused by vagal hyperactivity. The aim of this study was to evaluate the safety and efficacy of CNA in children with vasovagal syncope and/or symptomatic bradycardia caused by vagal hyperactivity.

Methods: patients had recurrent syncope with documented asystole and/or symptomatic bradycardia, refractory to conservative therapy. Preprocedural atropine tests revealed abnormally increased vagal tone in all patients. The Carto 3 navigation system was used for anatomical and electrophysiological identification of the ganglionated plexi. (GPs). Intraoperative criteria for the effectiveness of ganglionated plexi RFA were increased heart rate at least for 20 BPM, reduced antegrade Wenckebach point and improvement in atrioventricular conduction, as well as the absence of a bradycardia to intravenous phenylephrine insertion. Follow-up with HMECG was performed at 3, 6, 9, and 12 months postoperatively.

Results: between February 2021 and March 2026, 14 children (7 girls) underwent RFA at the Morozov Children's City Clinical Hospital in Moscow. The average age was 13.8 ± 3.7 years. During a follow-up period of 24.2 ± 21.7 months after RFA, 12 of 14 patients with previously documented syncope did not experience any further syncope. All children showed an improvement in mean and minimum heart rate after CNA. None of the patients experienced vascular injury, thromboembolism, pericardial effusion or other surgical complications.

Conclusion: cardioneuroablation is an effective and safe treatment for vasovagal syncope and/or symptomatic bradycardia caused by vagal hypersensitivity. However, careful patient selection and long-term regular follow-up are necessary for this procedure. Larger studies are needed to confirm these results.



PO.05 | EVOLUTION OF IMPLANTABLE CARDIOVERTER-DEFIBRILLATOR SHOCK THERAPY IN CHILDREN IN THE ERA OF OPTIMIZED ICD PROGRAMMING AND REMOTE MONITORING

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Aims

Implantable cardioverter defibrillator (ICD) therapy is effective in preventing sudden cardiac death in children. Unnecessary shocks should be avoided. ICD programming strategies and remote monitoring have been proven to be effective in preventing ICD shock therapy in adults; the present study evaluates the effectiveness in children.

Methods

Retrospective multi-center study, including children with transvenous or epicardial ICD implantation. During follow-up data on ICD shocks, programming variables and remote monitoring were collected.



Results

One hundred sixteen children were included, median age 13.4 years (min–max 0.3–18) and median follow-up 5.2 years (IQR 3.7–6.6), 53 with an ICD implanted before 2010 and 63 after 2010. The total appropriate and inappropriate annual shock rate decreased from 10.5 to 8% [difference in mean cumulative function (MCF) $P = 0.008$], 7.8 to 5.8% (MCF $P = 0.036$), and 4.3 to 2.6% (MCF $P = 0.28$), respectively, without increase in cardiac-related death. Remote monitoring was associated with a decrease of total shocks (MCF $P = 0.013$) and appropriate shocks (MCF $P = 0.052$). The ventricular fibrillation (VF) zone was programmed higher (≥ 210 bpm) at the time of appropriate vs. inappropriate shocks (86 vs. 79%; $P = 0.0016$), with no significant change in VF zone programming after 2010 compared to before 2010 (90 vs. 76%; $P = 0.142$) and in patients with vs. without shocks (79 vs. 89%; $P = 0.243$).

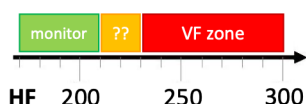
Conclusion

The incidence of ICD shocks has significantly decreased in children with implantation after 2010, which appears to be multifactorial. A changed programming behaviour and remote monitoring are associated with this decrease and support its use in strategies to prevent unnecessary shocks in children.

Evolution of implantable cardioverter-defibrillator shock therapy in children in the era of optimized ICD programming and remote monitoring.



Annual shock rate in 116 children decreased from 10.5% before 2010 to 8% after 2010 ($P=0.008$)

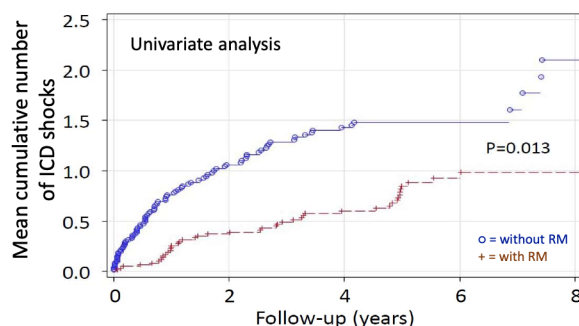


VF zone programmed ≥ 210 in:
86% of appropriate vs 79% of inappropriate shocks ($N=194$, $P=0.0016$)
76% before 2010 vs 90% after 2010 ($N=106$, $P=0.142$)
79% with vs 89% without ICD shocks ($N=106$, $P=0.243$)



Remote Monitoring

RM increased from 34% to 83% before 2010 versus after 2010 ($P<0.001$)





PO.06 | LEFT BUNDLE BRANCH AREA PACING IN PEDIATRIC PATIENTS: SINGLE-CENTER EXPERIENCE

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Keywords. Left bundle branch area pacing, atrioventricular block.

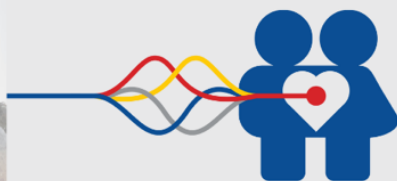
Background and Aim. Left bundle branch area pacing (LBBAP) has emerged as a physiological pacing modality and is an effective and safe alternative to conventional cardiac pacing. We present a single-center experience of LBBAP in 12 children.

Methods. 12 pediatric patients (mean age - $12,67 \pm 3,27$ years old, mean weight - $48,64 \pm 15,83$ kg) were diagnosed with complete atrioventricular block and underwent pacemaker implantation with implantation of a ventricular lead in the left bundle branch area. The mean follow-up period was $10,58 \pm 4,89$ months. QRS duration and echocardiographic parameters were measured before and after surgery. Standard pacing parameters were also assessed during the implantation and after 3 months. The presence of complications was assessed.

Results. Successful LBBAP was achieved in all 12 patients. The commonly used success criteria for LBBAP involve the paced QRS morphology displaying the pattern of right bundle branch block (RBBB), V6 R-wave peak time (V6 RWPT) less than 80 ms, interpeak time V1V6 (IPT V1V6) more than 33 ms. Due to the variability in pediatric body size and continuous growth, individualized LBBAP capture criteria are essential for precise clinical verification. The paced QRS displayed the pattern of RBBB in V1, mean V6 RWPT= $55,17 \pm 8,87$ ms, mean IPT V1V6= $43,50 \pm 6,5$ ms. During the implantation and at the 3-month follow-up optimal and stable ventricular lead pacing parameters were observed (mean pacing threshold after 3 months - $0,54 \pm 0,1$ V, mean sensing - $13,33 \pm 3,45$ mV, mean impedance - $493,25 \pm 92,12$ Ohm). Mean duration of the paced QRS complex was $100,67 \pm 11,14$ ms. Left ventricular dimensions and ejection fraction remained without significant changes (mean left ventricular end-diastolic diameter index - $3,15 \pm 0,57$ cm/m², mean left ventricular ejection fraction - $65,5 \pm 4,74\%$). No implantation-related complications occurred during the perioperative period.



Conclusion. LBBAP can be considered an effective and safe method of surgical treatment of atrioventricular block in pediatric patients, providing optimal pacing parameters, physiological ventricular contraction and potential prevention of pacemaker-induced cardiomyopathy, which necessitates further long-term observation. Finding of individualized LBBAP capture criteria is crucial for pediatric patients.



PO.07 | ARE OVERWEIGHT PEDIATRIC PATIENTS WITH LEFT VENTRICULAR HYPERTROPHY AT RISK OF VENTRICULAR ARRHYTHMIAS?

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Background

Childhood obesity is associated with metabolic alterations and early cardiac remodeling, including left ventricular hypertrophy (LVH). While hypertrophic cardiomyopathy is a recognized arrhythmogenic substrate, the prevalence and characteristics of arrhythmic findings in obesity-related LVH remain poorly defined.

Aim

To assess the prevalence of arrhythmic findings in overweight/obese pediatric patients with LVH and explore their relationship with structural and metabolic parameters.

Methods

Consecutive overweight/obese pediatric patients were included. LVH was defined as left ventricular mass index (LVMI) $\geq 38.6 \text{ g/m}^2$. Patients were stratified according to the presence or absence of structural heart diseases (SHD). SHD included all cardiomyopathies (hypertrophic, dilated, arrhythmogenic, and non-compaction). All patients underwent echocardiography, 12-lead ECG, 24-hour Holter monitoring and metabolic assessment: Homeostasis Model Assessment of Insulin Resistance (HOMA-IR), ApoB, NT-proBNP. Arrhythmic events included non-sustained or sustained ventricular tachycardia (NSVT/VT). Continuous variables are reported as mean $\pm$ standard deviation or median (interquartile range), as appropriate. A p-value < 0.05 was considered statistically significant.



Results

Twenty-four patients (13.0 ± 3.4 years, BMI 30 [27–32] kg/m²) were included. Eleven (46%) had SHD, thirteen (54%) had no SHD. LVMi was 105 (85–165) g/m², significantly higher in SHD than in non-SHD patients (119 [89–196] vs 93 [58–110] g/m², $p=0.02$). Metabolic parameters showed marked inter-individual variability in both groups. HOMA-IR was 3.5 (2.5–5.5), ApoB 0.72 (0.58–0.89) g/L, and NT-proBNP 55 (20–200) pg/mL. NT-proBNP levels were significantly higher in SHD than in non-SHD patients ($p=0.037$), whereas no significant differences were observed for HOMA-IR ($p=0.907$) and ApoB ($p=0.706$). Arrhythmias were observed in 9/24 (38%) patients, NSVT ($n=8$) and SVT ($n=1$), and were significantly more frequent in patients with (8/11, 73%) than without SHD (1/13, 8%) ($p=0.008$). Metabolic parameters did not significantly differ between patients with and without arrhythmias (HOMA-IR, ApoB, and NT-proBNP; all $p>0.05$).

Conclusions

In overweight/obese pediatric patients, arrhythmic events occur mainly in presence of SHD. Left ventricular hypertrophy and metabolic alterations do not seem to be independently associated with arrhythmic risk. Therefore, SHD seem to be the main determinant of arrhythmic events in this population.



PO.08 | LONG QT SYNDROME IN A PEDIATRIC PATIENT WITH EBSTEIN ANOMALY: WHEN THE RARE MEETS THE UNLIKELY

Gabriel Cruz Diaz¹, Leonardo Rivera Rodriguez¹, Alberto Anibal Galvez Valverde², Brenda Salamanca Gil², Javier Andres De Leon Tenaz¹

Introduction

Long QT syndrome is a rare genetic disorder with an estimated prevalence of 1 in 2,000 live births. Patients have an increased propensity for life-threatening arrhythmias, particularly under physical or emotional stress. (1–6) Ebstein's anomaly involves a malformation of the tricuspid valve associated with right ventricular myopathy. The reported incidence is approximately 1 in 20,000 live births. (7,8) Electrocardiographic findings classically include PR interval prolongation, tall P waves, and varying degrees of right bundle branch block. (9)

Case presentation

A 17-year-old patient presented for evaluation due to three episodes of syncope with emotional stress. The patient has a known diagnosis of Ebstein anomaly (Figure 1). There is no history of prior surgical correction or pharmacological management.

A baseline electrocardiogram was obtained (Figure 2), showing sinus rhythm with a heart rate of 99 bpm, QTm 400 ms, and QTcB in lead V5 of 478 ms.

An exercise stress test was then performed, achieving a maximum heart rate of 197 bpm, with a QTcB of 507 ms at peak exercise. Genetic testing was reported negative.

Based on these findings, treatment with Propranolol 3 mg/kg/day was initiated.

Discussion

There are only two reports in the literature describing patients with Ebstein's anomaly and long QT syndrome: one associated with amiodarone use, and a second case involving a 15-year-old patient with an exercise stress test showing a baseline QTcB of 438 ms, increasing to 489 ms at maximal exertion. (10,11)

According to the electrocardiographic criteria proposed by Schwartz, our patient scores 4 points yielding a high probability of congenital long QT syndrome. (6)



Conclusion

Although the genetic testing was negative, the patient's symptoms, along with the prolonged QTc interval that cannot be fully explained by her underlying condition, are noteworthy. Patients presenting with prolonged QTc in the context of concomitant underlying diseases continue to represent a gray area regarding management and prognosis. It is therefore of utmost importance to highlight and report these cases in order to work toward the standardization of management strategies and clinical approaches that allow patients to maintain a good quality of life.

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(INCARD IGNACIO CHAVEZ)
PRECONSULTA C-1

FC 99 . Ritmo sinusal.....eje P normal, frec-V 50- 99
Desviación del eje a la derecha.....eje QRS (111,269)
PR 149 . Bajo voltaje en derivaciones precordiales.....derivaciones precordiales <1.0mV
QRSD 87 . Anomalías de la onda T en el límite.....ángulo del eje QRS-T (91,180)
QT 373 . Intervalo QT prolongado en el límite.....QTc >475ms
QTc 479

--EJES--

P 40

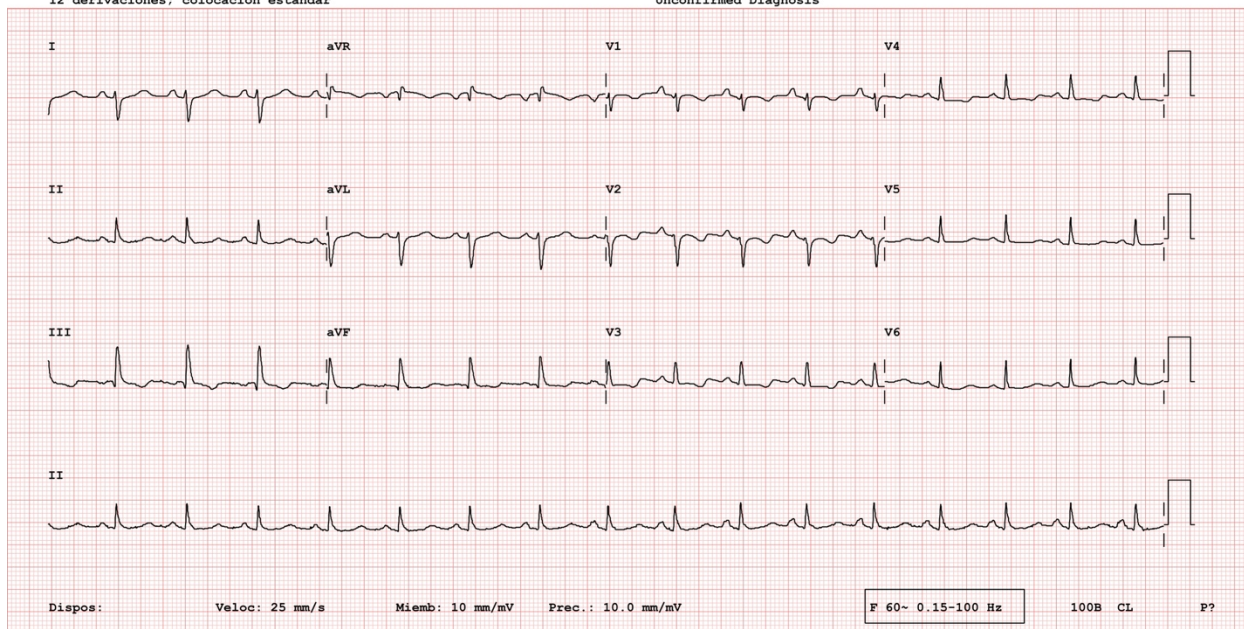
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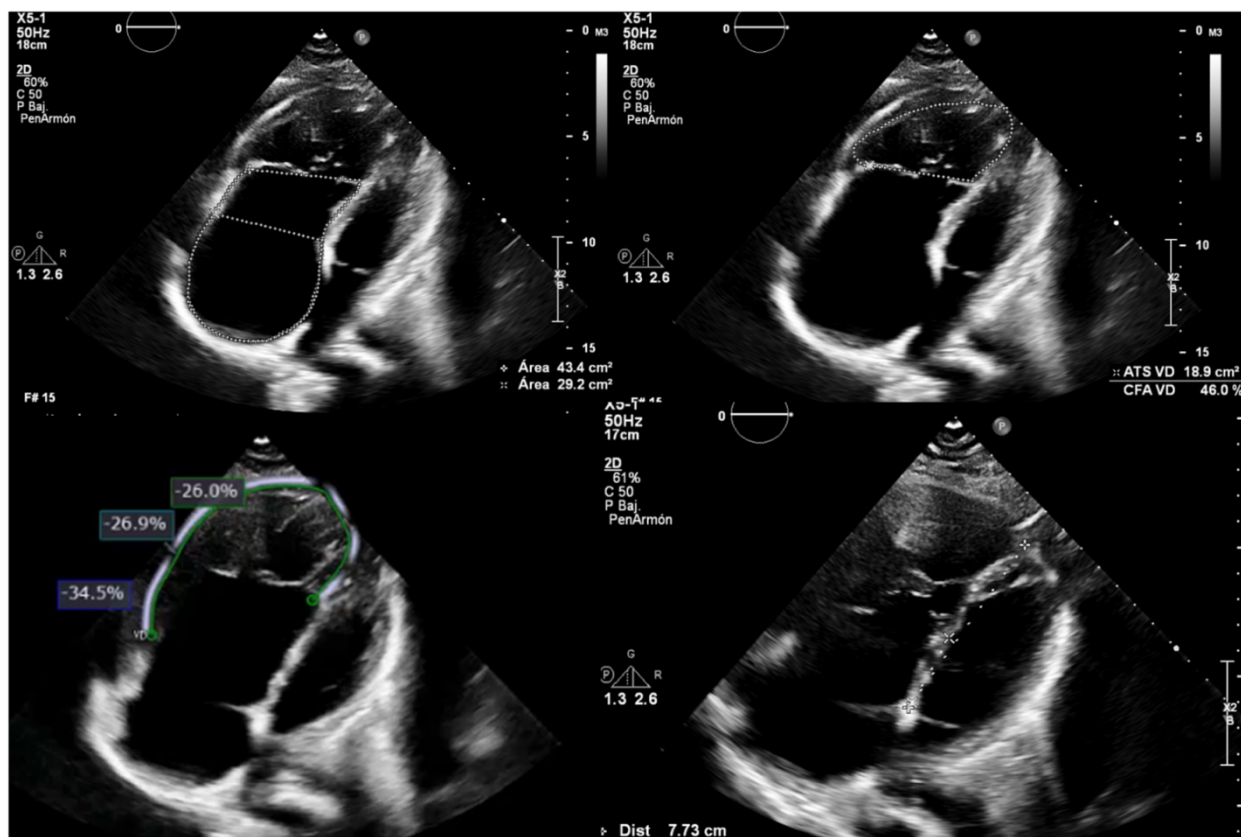
T -8

- ECG ANOMALO -

12 derivaciones; colocación estándar

Unconfirmed Diagnosis







PO.09 | SUCCESSFUL RESYNCHRONIZATION USING LEFT BUNDLE BRANCH AREA PACING AFTER DEVICE-RELATED CONDUCTION BLOCK IN A PEDIATRIC PATIENT

Musa Öztürk¹, İlker Ertuğrul¹, Hayrettin Hakan Aykan¹, Tefik Karagöz¹

¹ Department of Pediatric Cardiology, Hacettepe University Faculty of Medicine

Background and Aim

Transcatheter closure of perimembranous ventricular septal defects poses a risk of injury to the conduction system, especially left bundle branch block (LBBB), which may result in electromechanical dyssynchrony and eventually cardiomyopathy. While cardiac resynchronization therapy (CRT) remains the conventional treatment for dyssynchrony, its application in pediatric patients is often constrained by specific technical challenges. Therefore, this case highlights the effective use of left bundle branch area pacing (LBBaP) as a physiological alternative to reestablish synchrony and treat heart failure in a patient who developed cardiomyopathy following device closure.

Case Presentation

The patient underwent transcatheter closure of a perimembranous VSD at 18 months old. At the 24-month follow-up (6 months after the procedure), the patient showed impaired left ventricular systolic function, along with bundle branch block. A dual-chamber pacemaker with LBBaP was implanted due to a possible procedure-related fascicular block. Post-procedure electrocardiography showed significant QRS shortening (from 160–170 ms to 110 ms), indicating successful capture of the conduction system (Figure 1). The left bundle branch block has been resolved. Echocardiography demonstrated gradual improvement in left ventricular systolic function, with normalization of ejection fraction within nine months. Clinical symptoms fully resolved, and pacemaker settings remained stable over a 12-month follow-up without complications.

Conclusion

This case demonstrates the effectiveness of LBBaP as a viable treatment option for pediatric patients with device-related conduction block and heart failure, offering a physiological alternative to traditional pacing methods.



Keywords: Left bundle branch area pacing(LBBaP), transcatheter VSD closure, conduction block, device-related cardiomyopathy, children

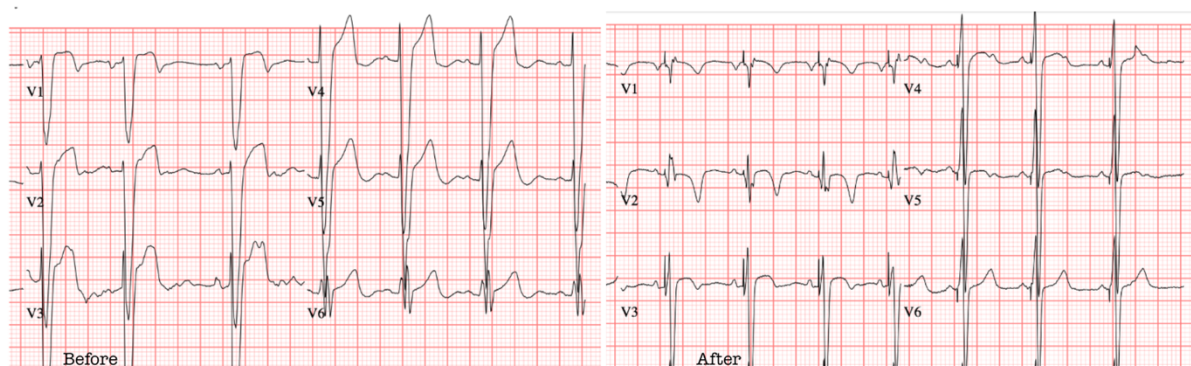


Figure 1: Comparison of chest leads of the patient's pre-procedure and post-LBBaP ECG.

25 mm/s - 10 mm/mV



PO.10 | THE EXPERIENCE OF OPEN WINDOW MAPPING IN PEDIATRIC PATIENTS WITH WOLFF-PARKINSON-WHITE SYNDROME: SINGLE-CENTER TRIAL

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1) Morozovskaya Children's City Clinical Hospital

2) I.V. Davydovsky City Clinical Hospital of the Moscow Department of Health

3) Patrice Lumumba Peoples' Friendship University of Russia

The authors of the study declare no conflict of interest.

Background and Aim. Wolff-Parkinson-White (WPW) syndrome is one of the most common causes of supraventricular tachycardia in children, also associated with a risk of sudden cardiac death. Currently, catheter ablation is considered to be the treatment of choice for WPW syndrome; however, the recurrence rate after ablation of accessory pathways (AP) can reach up to 12%. A special group of patients are those with Ebstein's anomaly. In addition to the aforementioned risks, effective ablation should be performed before surgery of the congenital heart disease. Open window mapping (OWM) appears to be one of the most effective strategies nowadays. Currently, there are no systematic reviews on the use of OWM in children. The aim of this study is to evaluate the efficacy and safety of this approach in pediatric patients.

Materials and Methods. From January 2021 to March 2026, open window mapping was performed in 91 patients aged from 2 to 17 years (mean age $12,8 \pm 3,5$), including 8 patients with Ebstein's anomaly. The minimum weight of the patient undergoing OWM during procedure was 12 kg; the minimum weight of the patient with Ebstein's anomaly was 16 kg. All procedures were performed using the Carto 3 system. The criterion for evaluating the effectiveness of the procedure was the absence of antegrade and retrograde conduction through accessory pathway intraoperatively, as well as the absence of recurrence at 3, 6, and 12 months after the procedure.

Results. Open-window mapping significantly reduced the recurrence rate in patients with right-sided accessory pathways. Patients with Ebstein's anomaly in our study demonstrated zero recurrence rate, despite their accessory pathways used to be broad and oblique. No major complications were observed during the entire follow-up period. From 2021 to 2024, only patients with Ebstein's anomaly or patients undergoing reinterventions were selected as candidates for open-window mapping. From 2025 onwards in our department open-window mapping is performed as a primary procedure for all children with right-sided accessory pathways.

Conclusion. Open window mapping is considered to be a promising option in reducing procedure time, recurrence and complication rates, which also demonstrates extra utility in patients with Ebstein's anomaly.



PO.11 | HIGH POWER SHORT DURATION RADIOFREQUENCY ABLATION OF LEFT ATRIAL FOCAL ATRIAL TACHYCARDIA - A PAEDIATRIC CASE SERIES

Sarah Lengauer¹, Meike Schwendt¹, Isabel Deisenhofer¹, Gabriele Hessling¹, Marta Telishevska¹

¹ Department of Electrophysiology, German Heart Center Munich, Technical University of Munich

Background and Aim:

Focal atrial tachycardia (FAT) is rare in children, predominantly arises from the right atrium, and is often drug-resistant. Conventional radiofrequency ablation (RFA) is safe and effective. We report a paediatric case series of rare left atrial (LA) FAT treated with high-power short-duration (HPSD) RFA. This technique used in adults for pulmonary vein isolation (PVI) offers shorter procedures, more efficient lesions and greater safety. As HPSD also creates shallower lesions, these benefits are particularly valuable in children, given thinner atrial walls and smaller hearts and might reduce the risk of collateral damage.

Methods: We report three pediatric patients aged 10, 12, and 17 with drug-refractory FAT undergoing ablation. None had congenital heart disease. During electrophysiological study, multipolar activation mapping (EnSite Precision or EnSite X; Abbott) with Advisor™ HD Grid confirmed LA FAT. A 4 mm contact-force-sensing irrigated catheter (Tactiflex™ SE) was used with energy settings of 65 W for 6–7 s on the posterior and 10–15 s on the anterior LA wall. Two patients underwent unilateral antral HPSD PVI for FAT near the left or right inferior pulmonary vein; one received focal RFA on the posterior LA roof. A 20 min wait confirmed success.

Results: No complications occurred. FAT was terminated by ablation in all cases. Mean procedural time was 64.3 ± 0.02 min; RF time was 6.8 ± 2.1 min. After the procedure, two patients received apixaban for 3 months, and one received ASA. Follow-up included 24 h Holter monitoring and outpatient visits. Mean follow-up was 65.3 ± 28.1 days, with no FAT recurrences.

Conclusions: In this pediatric case series, HPSD RFA showed excellent safety and efficacy with no complications or recurrence during follow-up. These results align with adult HSPD pulmonary vein isolation studies, which report good procedural and safety outcomes.

Disclosure: No significant financial interests or relationships with related product manufacturers, service providers, or companies.

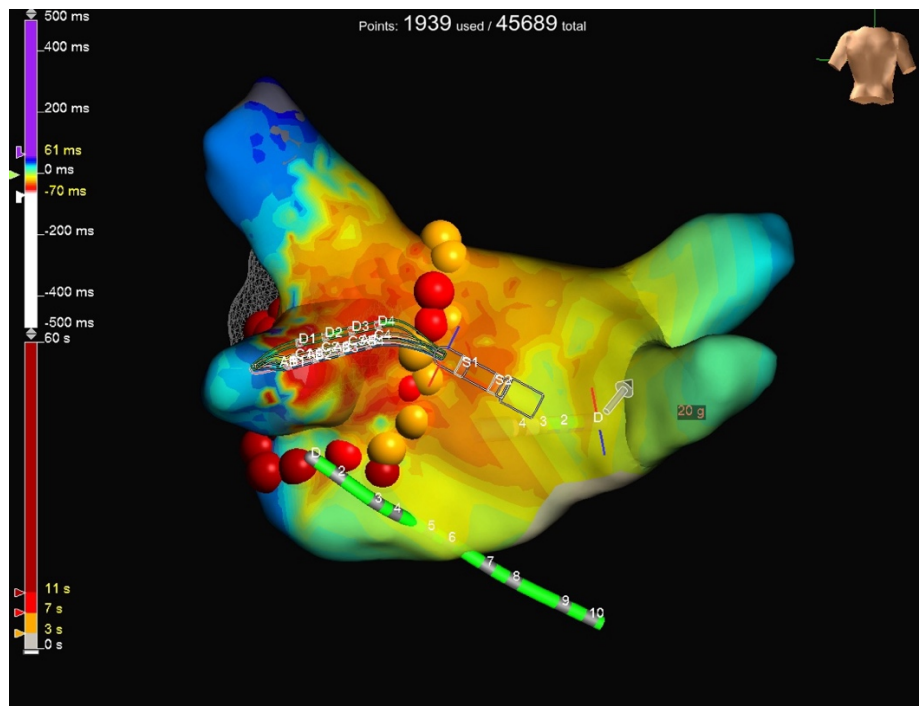


Figure 1: LAT map displaying FAT from the LIPV, ablationsets for HPSP PVI left

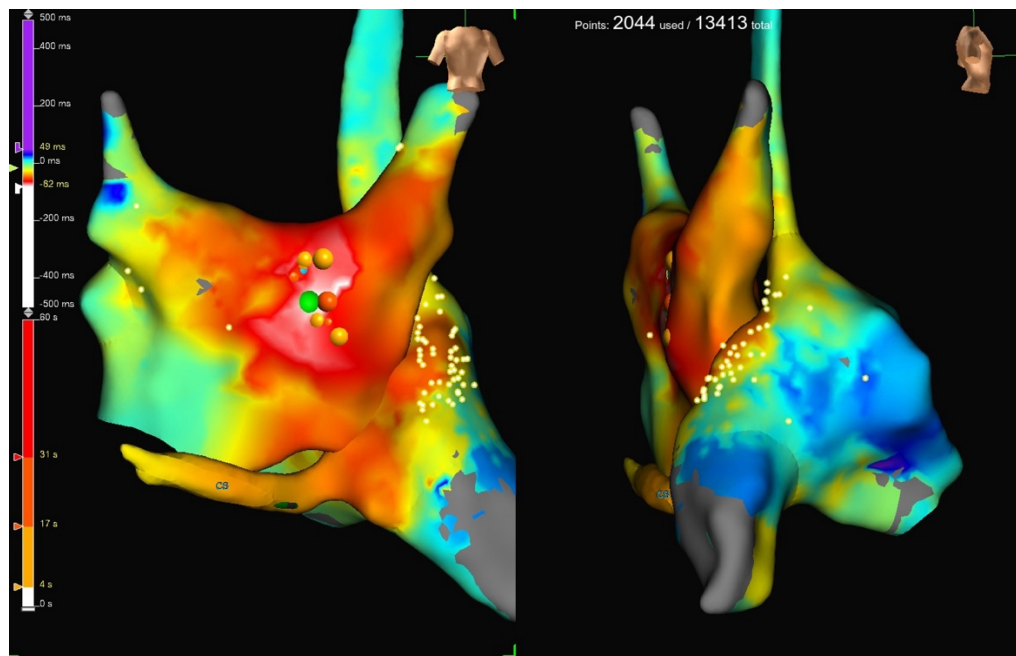


Figure 2: LAT map displaying FAT from the posterior roof near the RSPV, focal HPSP ablation lesion set



PO.12 | IMPACT OF ELECTRODE SIZE ON UNIPOLAR AND OMNIPOLAR VOLTAGE MAPPING IN PEDIATRIC AND ADULT ATRIA: DOES SIZE MATTER?

Mathijs van Schie¹, Paul Knops¹, Manouk Linderhof¹, Nawin Ramdat Misier¹, Vehpi Yildirim¹, Yannick Taverne², Natasja de Groot¹

¹ Cardiology, Erasmus University Medical Center

² Cardiothoracic surgery, Erasmus University Medical Center

Background and Aim: Atrial tachyarrhythmias occur in 60% of adult and 5-15% of pediatric patients with congenital heart disease (CHD). Effectiveness of catheter ablation in these patients is often hampered by altered anatomy and complex arrhythmogenic substrates. Voltage mapping is frequently used during electrophysiological studies to identify arrhythmogenic substrates underlying arrhythmias in pediatric and adult CHD patients. In daily clinic, bipolar electrograms are most commonly used, although omnipolar electrograms can be used to overcome directional influences affecting the bipolar peak-to-peak voltage. Unipolar electrograms are also directional independent, although they are influenced by the amount of cardiac tissue underneath the recording electrodes. It remains unknown whether these types of electrograms are influenced by the different cardiac sizes in pediatric and adult patients.

Methods: Intraoperative bi-atrial epicardial mapping (interelectrode distance 2 mm) was performed in 59 pediatric and 65 adult patients (2.8 [1.0-6.4] vs 44 [35-55] years). Unipolar electrograms were recorded from which omnipolar electrograms were constructed using two orthogonal electrode pairs. On average 5,231±2,229 unipolar and 15,574±7,633 omnipolar potentials were included from pediatric compared to 8,762±2,903 unipolar and 28,647±9,532 omnipolar potentials from adult patients.

Results: In adults, bi-atrial unipolar voltages were larger than omnipolar voltages (5.5 [4.7-6.6] vs 4.9 [4.0-6.5] mV, $P<0.001$), while in pediatrics omnipolar voltages were larger than unipolar voltages (5.3 [4.2-6.6] vs 6.8 [5.5-8.4] mV, $P<0.001$). In both groups, median unipolar voltages linearly correlated with median omnipolar voltages (adults: $R^2=0.981$, pediatrics: $R^2=0.953$). Unipolar voltages were comparable between pediatrics and adults ($P=0.418$), while omnipolar voltages were larger in pediatrics ($P<0.001$). For the entire population, there was a strong inverse exponential relationship between age and the difference of omnipolar and unipolar voltages ($R^2=0.874$).

Conclusions: The different cardiac sizes did not have any impact on unipolar potential voltages. However, the smaller cardiac size relative to the interelectrode distance in pediatrics most likely influenced the omnipolar potential voltages, resulting in larger voltages as compared to adults. Usage of this technique for voltage mapping should therefore be with caution and specific pediatric voltage cut-offs should be considered.



PO.13 | IDENTIFICATION OF SINO-ATRIAL NODE LOW-VOLTAGE AREAS USING UNIPOLAR AND OMNIPOLAR MAPPING IN PEDIATRIC PATIENTS WITH CONGENITAL HEART DISEASE

Mathijs van Schie¹, Jiahao Zheng¹, Manouk Linderhof¹, Lixia Dai¹, Can Zhang¹, Yannick Taverne², Annemien van den Bosch¹, Natasja de Groot¹

¹ Cardiology, Erasmus University Medical Center

² Cardiothoracic surgery, Erasmus University Medical Center

Background and Aims: Sinus node dysfunction (SND) commonly occurs in pediatric congenital heart disease (CHD) patients. Unipolar and omnipolar mapping techniques are available to explore the degree of electrical remodeling in the sino-atrial node (SAN). While each technique offers specific advantages and disadvantages, their roles in SAN mapping in pediatrics are not yet fully understood. We therefore performed high-resolution voltage mapping of the SAN to investigate the interrelationship between unipolar and omnipolar low-voltage areas.

Methods: Epicardial mapping was performed in 45 patients (0.6 [0.4-3.1] years). Potential morphology characteristics were computed, including peak-to-peak voltages and relative R/S ratios. Omnipolar electrograms (EGM) were constructed from orthogonal bipolar EGM pairs. Low-voltage areas (LVA) were defined as unipolar<1mV and omnipolar<0.5mV.

Results: Omnipolar voltages surrounding the SAN were larger than unipolar voltages (6.2 [4.8-7.6] vs 5.2 [3.6-6.3] mV, $p<0.001$). Omnipolar LVAs near the SAN were identified in only 9 (20%) patients, while unipolar LVAs were present in 34 (76%) patients; 8 patients had combined unipolar and omnipolar LVAs. Unipolar LVAs were larger than omnipolar LVAs (6.5 [4.6-10.5] vs 0.7 [0.5-2.2]%, $p<0.001$). At the SAN exit site, the morphology of unipolar potentials consistently consisted of full S-wave potentials, while the morphologies of omnipolar potentials were highly variable (peak dominance ratio: 0.92 [0.89-0.95] vs 0.38 [0.18-0.52], $p<0.001$).

Conclusion: At the SAN area, a considerable amount of LVAs could be identified using unipolar voltage mapping. These unipolar potentials typically consisted of full S-wave potentials, which were easily recognizable but led to an overidentification of LVAs. By utilizing combined voltage mapping, LVAs near the SAN may be more accurately identified and used to identify patients at risk for developing SND.

**PO.14 | NON-TRANSVENOUS ICD IN CHILDREN**

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Introduction

Pediatric patients requiring ICD are burdened with a higher risk of complications, mainly due to limited anatomical dimensions and need for long-term ICD system durability. In patients with no cardiac pacing indication, non-transvenous systems like subcutaneous ICD (S-ICD) and extravascular ICD (EV-ICD) represent alternatives that avoid several disadvantages of transvenous ICD systems, such as intravascular infections and complications associated with lead extraction. In this study, we present our experience with the use of S-ICD and EV-ICD in the pediatric population in Slovakia.

Materials and Methods

All pediatric patients who underwent S-ICD and EV-ICD implantation since the initiation of the non-transvenous ICD program at our center in September 2019 were included in this retrospective analysis. Implantation procedures were performed under general anesthesia. S-ICD was implanted using the two-incision technique with intermuscular can placement and EV-ICD was implanted by substernal tunneling.

Results

From a total of 21 patients, 18 underwent S-ICD and 3 patients EV-ICD implantation. Primary prevention of SCD was initial indication in 11 patients. Three patients underwent replacement of previous ICD system due to lead dysfunction. Ten patients were diagnosed with channelopathy, ten patients had cardiomyopathy and one unknown diagnosis at the time of ICD implantation. Median age was 14.5 years (range 9–18 years), median body weight was 58 kg (range 25.5–80.5 kg), and median body mass index (BMI) was 20 kg/m². No periprocedural complications were observed. Median follow-up duration was 24 months (range 2–78 months). All patients demonstrated normal parameters of respective ICD systems.



Three patients (14,3%) experienced appropriate shock, one of them had VT/VF triggered by inappropriate shock (IAS). 3 patients (14,3%) experienced inappropriate shocks. Four patients (19%) had inappropriate tachycardia detection without shock delivery. Following sensing adjustment and adherence to regime instructions, no further IAS were recorded during follow-up. S-ICD was explanted in one patient during heart transplantation procedure. One patient with EV-ICD needed system removal due to infection and eventual transvenous ICD implantation.

Conclusion

We demonstrated favorable results regarding the safety and efficacy of non-transvenous ICD systems in children. The use of S-ICD and EV-ICD in selected patients represent a relevant therapeutic option that may reduce short-term and, particularly, long-term complications associated with transvenous ICD.

Keywords

Sudden cardiac death, S-ICD, EV-ICD, appropriate shock, complications



PO.15 | UTILITY OF AN AUTOMATIC AND PATIENT-TRIGGERED CARDIAC EVENT RECORDER: FIRST EXPERIENCE IN CHILDREN

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² Inonu University, Department of Pediatric Cardiology, Malatya, Turkey,

Background and Aim: This study aimed to evaluate the diagnostic yield of an event recorder capable of automatic and manual recording in children with possible arrhythmia related symptoms.

Methods: We evaluated retrospectively 81 children monitored with Novacor event recorder system between September 2021 and September 2024(Figure 1). All of the patients performed 12-lead electrocardiogram (ECG), 24-hour-Holter monitoring, and echocardiography before the cardiac event recording.

Results: The mean patient age was 11.30 ± 3.87 years. The indications included palpitation in 59 (72.8%) patients, chest pain and palpitations in 19 (23.5%) patients, palpitation and pre-syncope in 3 (3.7%) patients. Symptoms were identified in 65 children using the Event Recorder, and different types of arrhythmias were detected in 41 (63%) children. Of the 16 patients who were asymptomatic during the enrollment period, 9 (56%) had arrhythmia recorded with automatic arrhythmia detection property only. While 24-hour-Holter monitoring detected a clinical diagnosis in 13 patients (16.7%), event recorder diagnosed the clinical arrhythmia in 41 children (63%) (Figure 2).

Conclusion: We found that the automatic mode of the device provided additional clinical and therapeutic consequences. For this reason, a cardiac event recorder capable of automatically and manually recording may be a more preferable method in children with potentially arrhythmia related symptoms.

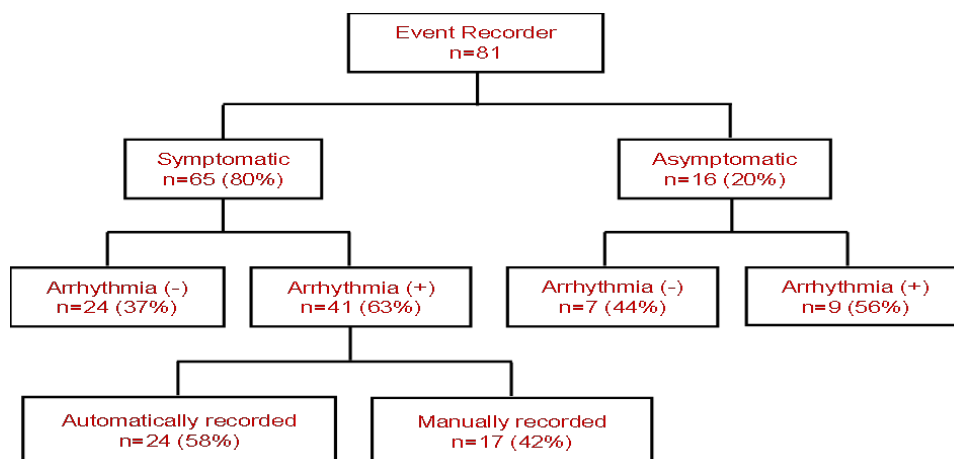
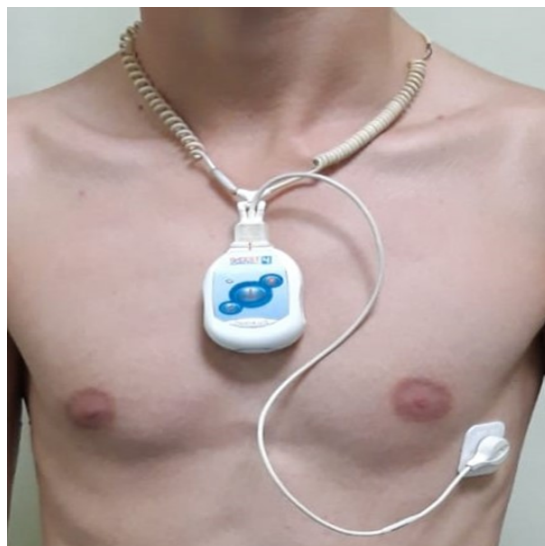


Figure 2. Arrhythmias identified with cardiac event recorder

**PO.16 | LONG-TERM OUTCOMES OF CATHETER ABLATION FOR SUPRAVENTRICULAR ARRHYTHMIAS IN PATIENTS WITH EBSTEIN ANOMALY: A SINGLE-CENTER EXPERIENCE**

Marta Telishevska¹, Sarah Lengauer¹, Miruna Popa¹, Meike Schwendt¹, Isabel Deisenhofer¹, Gabriele Hessling¹

¹ Electrophysiology, German Heart Center Munich, Germany

Background and Aim: Ebstein anomaly (EA) is a rare congenital tricuspid valve malformation associated with supraventricular arrhythmias (SVA), including accessory pathway-mediated atrioventricular reentrant tachycardia (AVRT)/Wolff-Parkinson-White (WPW) pattern, intra-atrial reentrant tachycardia (IART), atrial flutter, and atrial fibrillation (AF). Catheter ablation (CA) is the treatment of choice, but anatomical complexity and prior surgery may increase procedural difficulty. We evaluated long-term outcomes of CA across all SVA subtypes in EA.

Methods: We retrospectively analyzed all EA patients undergoing CA for SVA at our institution between 2011 and 2026. Procedures were performed using high-density three-dimensional electroanatomical mapping. Acute success was defined as non-inducibility at procedure end, and recurrence as documented SVA after a 3-month blanking period.

Results: Forty-five patients underwent 68 ablation procedures. The median age was 42 years (range 9–70), 56% were female, and 16% were pediatric. Prior cardiac surgery was present in 44%, including Cone reconstruction in 7%, and 27% underwent surgery after ablation. Cardiac implantable devices were present in 18%. The most common arrhythmia at first ablation were WPW (40%), typical atrial flutter (20%), atypical atrial flutter/IART (13%), paroxysmal AF (11%), persistent AF (9%), focal atrial tachycardia (4%), and combination arrhythmia (2%); multiple accessory pathways were present in 9%. Acute procedural success was 98.5%. Over a median follow-up of 35 months (range 1–160), recurrence occurred in 38% of patients, while recurrence after the final ablation was 20%.



The highest recurrence rates after the final ablation were observed in persistent and paroxysmal AF (both 50%), highlighting AF as the substrate with the greatest risk of recurrence following CA. Patients underwent 1–5 ablation procedures, with a median of 1. All pediatric patients had WPW and achieved 100% acute success; one child experienced recurrence after the first ablation but remained arrhythmia-free after repeat ablation. Complications occurred in 2.9% of procedures and included one sinus arrest requiring pacemaker implantation and one conservatively managed pericardial effusion. No stroke or procedure-related death occurred.

Conclusions: In this large single-center EA cohort, CA demonstrated high acute success, good safety, and durable long-term arrhythmia control across a broad spectrum of SVAs, including excellent outcomes in children with WPW.



PO.17 | ANTIMICROBIAL ENVELOPE USE FOR PREVENTION OF CIED INFECTION IN ACHD PATIENTS

Holly Daw¹, Laura Bayliss¹, Luke Sevier¹, Vinit Sawhney¹, Martin Lowe¹, Filip Zemrak¹, Vivienne Ezzat¹

¹ Cardiology, Barts Heart Centre

Background: Patients with ACHD are at increased risk of CIED complications, including infection due to many factors including young age at implant and the requirement for multiple procedures over their lifetime. In recent years novel strategies to reduce risk of CIED infection have been introduced including the use of antimicrobial envelopes such as Tyrx (Medtronic). In January 2023 Barts Heart Centre implemented the use of the 'BLISTER' score to direct the use of Tyrx, with a BLISTER score of 6 as the threshold for use (1).

Methods: We reviewed procedural data from all patients with an ACHD diagnosis that underwent CIED procedure in the years 2023 to 2025 at a single tertiary centre. Basic demographics and Tyrx utilisation was recorded and analysed. Comparison was made with the non-ACHD cohort.

Results: 312 ACHD patients underwent CIED procedures during the study period, of which 39% received Tyrx. 50% of Tyrx recipients were female, with a mean age 48 (range 18-82 years). When procedures not normally amenable to Tyrx were excluded the utilisation increased to 57% (Figure 1). Exclusion was applied to leadless pacemakers, SICD, and generator removal. Tyrx utilisation was highest in CRTP and CRTD procedures (Figure 2). In comparison Tyrx use over the same period was 11% for the non-ACHD cohort, with 35% female, mean age 71 (range 17-99 years).

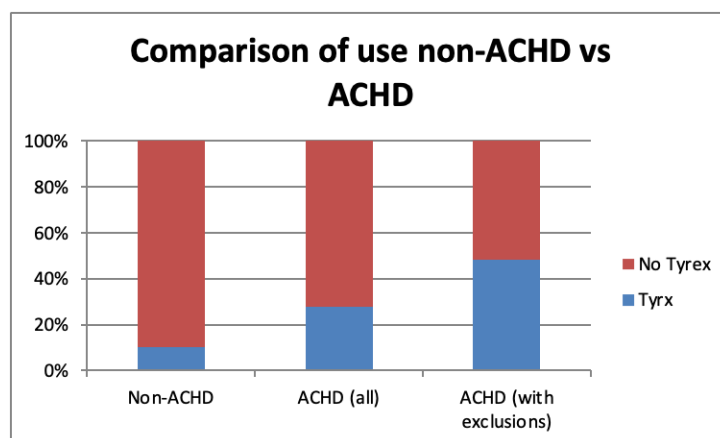


Figure 1: Comparison of Tyrx use in non-ACHD cohort Vs ACHD cohort with and without exclusions for leadless/SICD/generator removal.

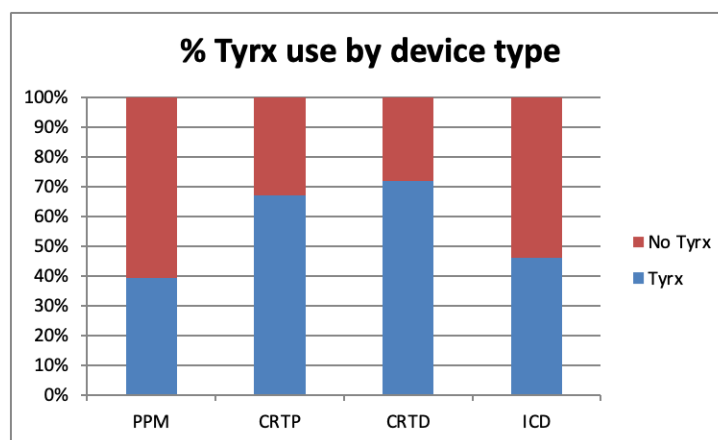


Figure 2: % Tyrx use by device type across the study period.

Conclusions: The use of the BLISTER score to simplify decision-making regarding Tyrx use for CIED procedures allows for comparison between ACHD and non-ACHD cohorts. This data demonstrates that ACHD patients have a significantly higher BLISTER score than a non-ACHD cohort. This is largely attributable to young age and number of previous procedures. As expected for all patients, use in CRTP and CRTD procedures was higher as this is a factor that directly increases BLISTER score. Further work is required to investigate the link between higher BLISTER score and sex in the ACHD cohort.

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- (1) Maclean E, Mahtani K, Honarbakhsh S, Butcher C, Ahluwalia N, Dennis ASC, Creta A, Finlay M, Elliott M, Mehta V, Wijesuriya N, Shaikh O, Zaw Y, Ogbedeh C, Gautam V, Lambiase PD, Schilling RJ, Earley MJ, Moore P, Muthumala A, Sporton SCE, Hunter RJ, Rinaldi CA, Behar J, Martin C, Monkhouse C, Chow A. **The BLISTER Score: A Novel, Externally Validated Tool for Predicting Cardiac Implantable Electronic Device Infections, and Its Cost-Utility Implications for Antimicrobial Envelope Use.** *Circ Arrhythm Electrophysiol.* 2024 Mar;17(3):e012446. doi: 10.1161/CIRCEP.123.012446. Epub 2024 Jan 22. PMID: 38258308; PMCID: PMC10949977.



PO.18 | CIED TYPE AND ACHD COMPLEXITY – EXPERIENCE FROM A SINGLE TERTIARY CENTRE

Holly Daw¹, Charles Butcher¹, Laura Bayliss¹, Luke Sevier¹, Vinit Sawhney¹, Martin Lowe¹, Filip Zemrak¹, Vivienne Ezzat¹

¹ Cardiology, Barts Heart Centre

Background: The term ACHD covers a wide range of diagnoses, a proportion who will require a CIED (cardiac implantable device). We describe a large tertiary centre cohort; basic demographics and device type implanted by ACHD complexity are presented.

Methods: Retrospective, electronic patient record, data was collected for all patients seen in a dedicated CIED clinic between 2022 and 2025. Those with an ACHD diagnosis or congenital complete heart block (cCHB) were included. ACHD complexity was categorised according to American heart association guidelines (1)

Results: 804 patients were included in the analysis. 49% were female with an average age of 44 (range 16-88 years). 90% of patients had an ACHD diagnosis (10% cCHB). Of those with ACHD, 48% had simple lesions, 28% moderate and 24% complex, (*Table 1a*). Across all groups, pacemaker was the most common implanted device (50%), of which 9% were leadless (*Table 1b*). The highest proportion of high energy devices (ICD and CRTD) was seen in those with moderate lesions (28%). 11% of all high energy devices were SCD. Implantable monitors (ICM) made up 27% of implanted devices with the greatest proportion in simple lesions (*Figure 1*).

Table 1a

	Total (n=804)
Age at last appointment (SD)	44 (17)
Sex F n (%)	390 (49)
Patients with ACHD n (%)	721 (90)
Complexity of CHD Lesion	
Complex n (%)	176 (24)
Moderate n (%)	199 (28)
Simple n (%)	346 (48)
Patients with isolated cCHB n (%)	83 (10)

Table 1b

Division by device type	Total (n=804)
Loop recorder n (%)	196 (24)
Pacemaker n (%)	404 (50)
- Of which leadless	37 (5)
Cardiac Resynchronisation Therapy Pacemaker n (%)	46 (6)
Cardiac Resynchronisation Therapy Defibrillator n (%)	45 (6)
Cardioverter Defibrillator n (%)	113 (14)
- Of which SCD	18 (2)

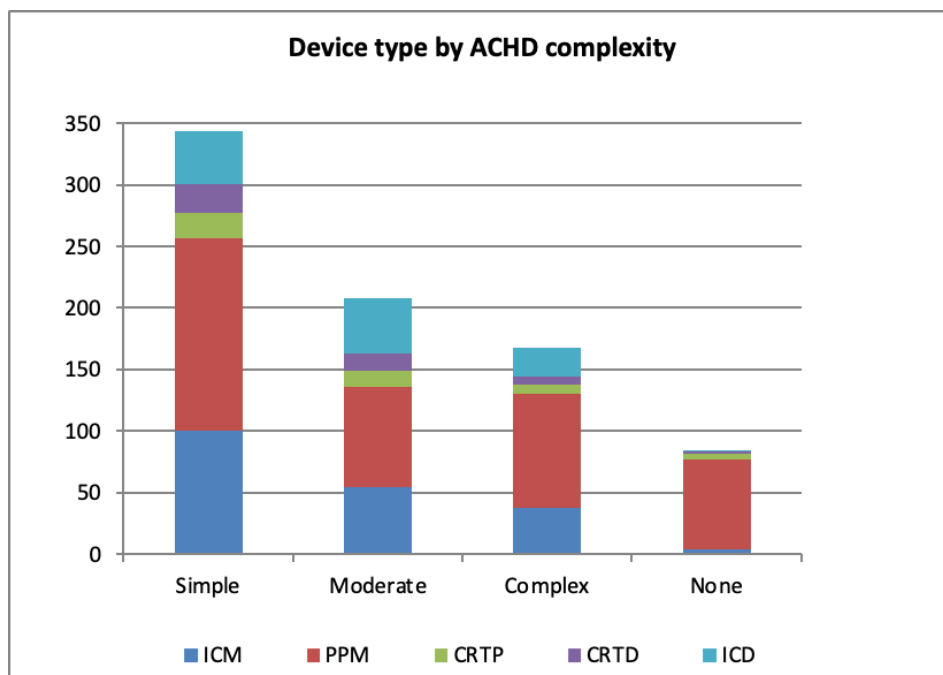


Figure 1: Device type by ACHD complexity (none = isolated cCHB).

Conclusions: These data show the complexity of ACHD patients requiring CIED follow-up. The majority have pacemakers, and the highest proportion of patients with defibrillators are in those with moderate ACHD lesions. Further work is needed to better describe outcomes in this growing complex cohort of patients with both ACHD and CIED.

References:

- (1) **2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines.** Karen K. Stout, MD, FACC, Chair, Curt J. Daniels, MD, Vice Chair, Jamil A. Aboulhosn, MD, FACC, FSCAI, Biykem Bozkurt, MD, PhD, FACC, FAHA, Craig S. Broberg, MD, FACC, Jack M. Colman, MD, FACC, Stephen R. Crumb, DNP, AACC, Joseph A. Dearani, MD, FACC, Stephanie Fuller, MD, MS, FACC, Michelle Gurvitz, MD, FACC, Paul Khairy, MD, PhD, Michael J. Landzberg, MD, FACC, Arwa Saidi, MB, BCH, FACC, Anne Marie Valente, MD, FACC, FAHA, FASE, and George F. Van Hare, MD, FACC. *Circulation*

**PO.19 | THE MANY FACES OF POSTERIOR FASCICULAR VENTRICULAR TACHYCARDIA:
FROM NEONATES TO TEENAGERS, NOT ONE-SIZE-FITS-ALL****Erin Skelly¹, Dipika Menon¹**

¹ Saint Louis University Department of Pediatrics, SSM Health Cardinal Glennon Children's Hospital

Introduction:

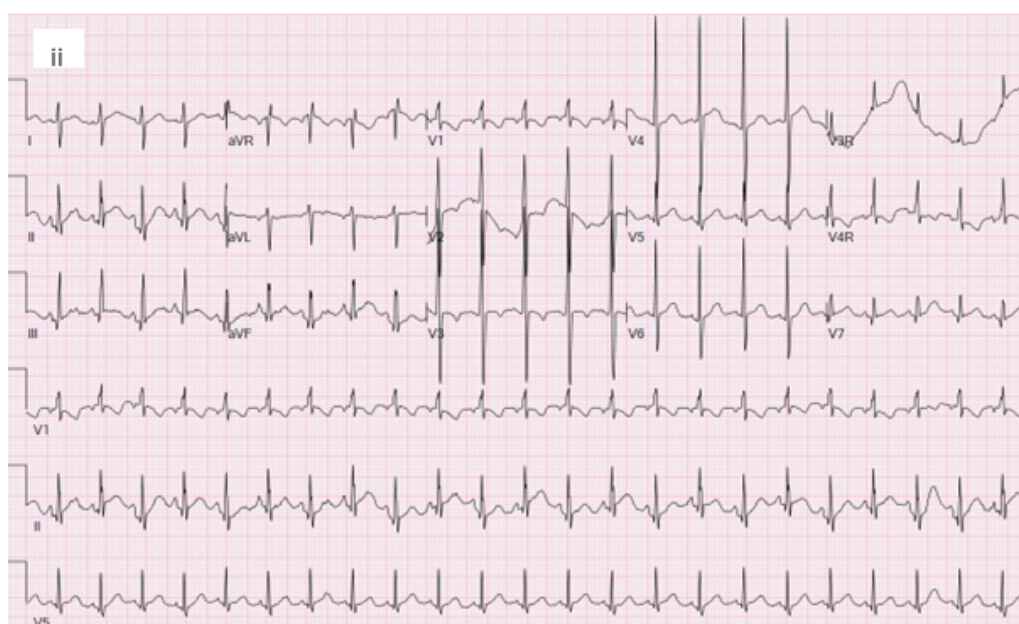
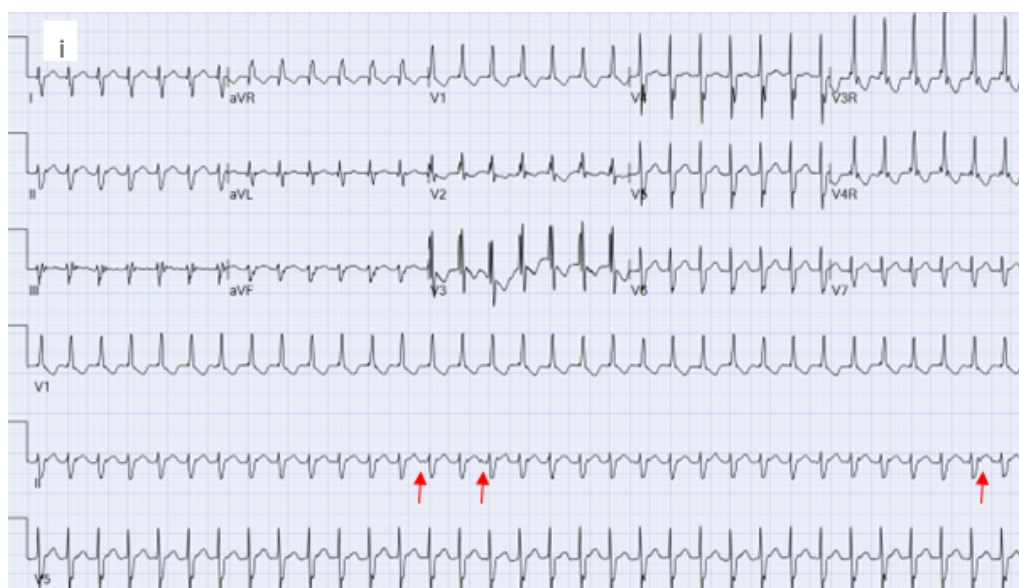
Belhassen's ventricular tachycardia (VT) or idiopathic left posterior fascicular VT is a reentrant arrhythmia originating from the left posterior fascicle demonstrating a characteristic right bundle branch block pattern and left axis deviation. It represents about 10-15% of all idiopathic left VTs and mainly affects males aged 15-40 years. Given that the reentrant circuit comprises a verapamil-sensitive zone, these arrhythmias are recognized as being uniquely sensitive to calcium channel blockers, especially verapamil.

Description:

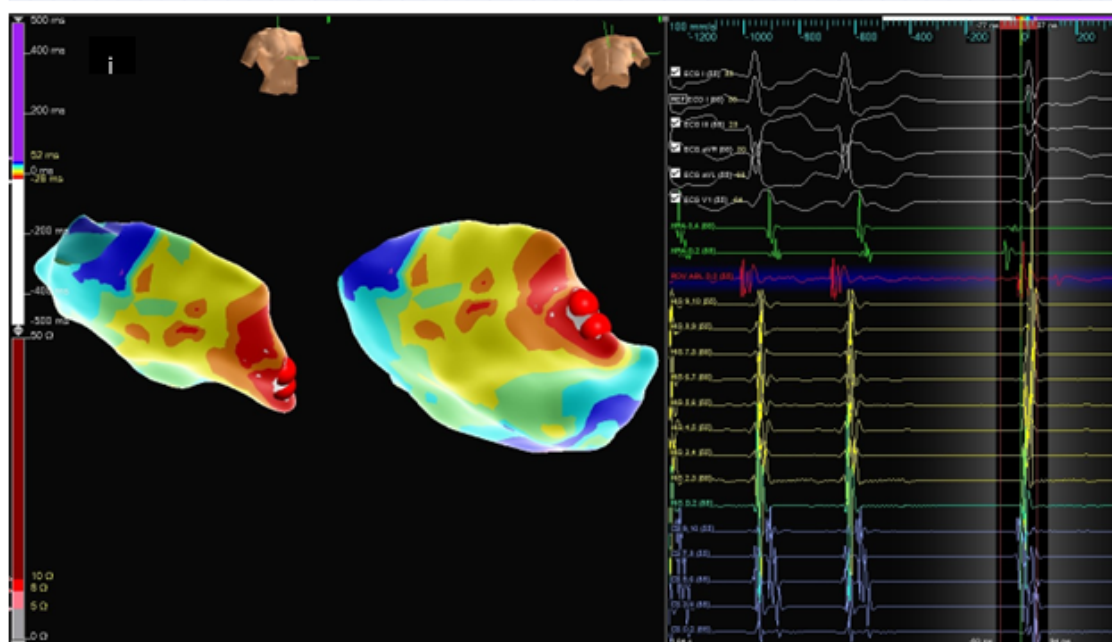
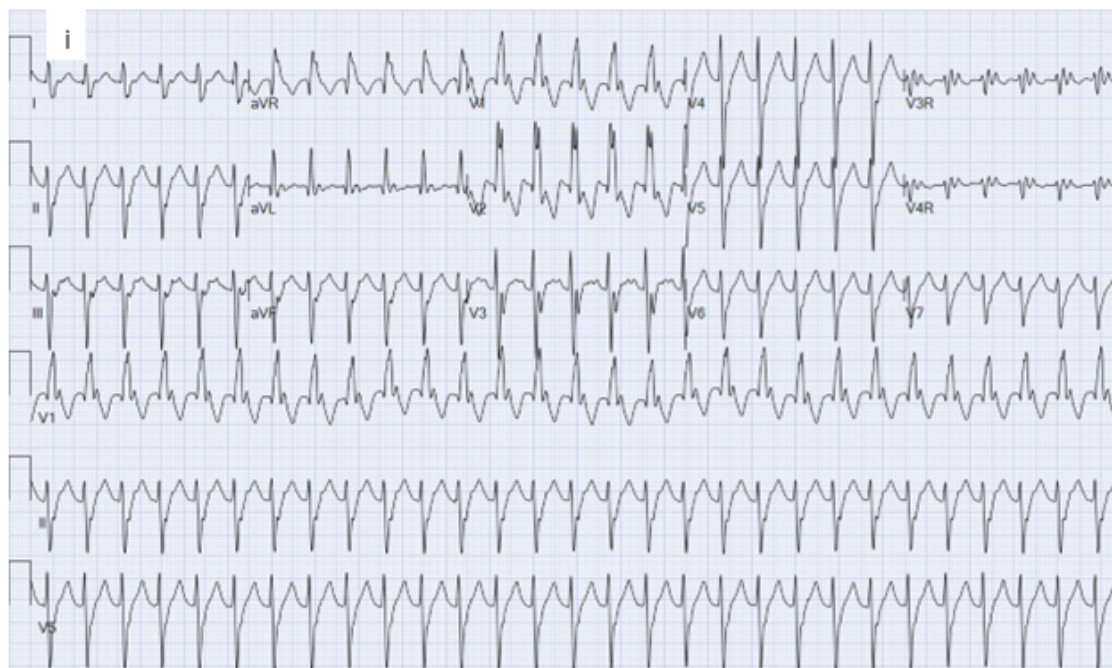
We present a case series of three pediatric patients diagnosed with idiopathic left posterior fascicular VT. Our patients were diagnosed at 14 days (Patient A), 4 years (Patient B), and 16 years (Patient C). Two were male and one was female (Patient B). Treatment response varied in our patients – Patient A converted to sinus rhythm with an amiodarone bolus and is currently maintained on oral amiodarone with no further recurrence. Patient B was initially cardioverted at an outside facility. She was started on verapamil, but experienced multiple breakthrough episodes of tachycardia despite dose optimization. Sotalol was then added to her medical regimen, and she ultimately underwent successful ablation of the substrate with no further recurrence. Patient C had a more classic presentation as a teenager and converted to sinus rhythm with a single dose of IV verapamil. He was started on oral verapamil which was continued until he underwent successful ablation of the substrate. All patients had echocardiograms with normal cardiac anatomy and systolic function.

Conclusion:

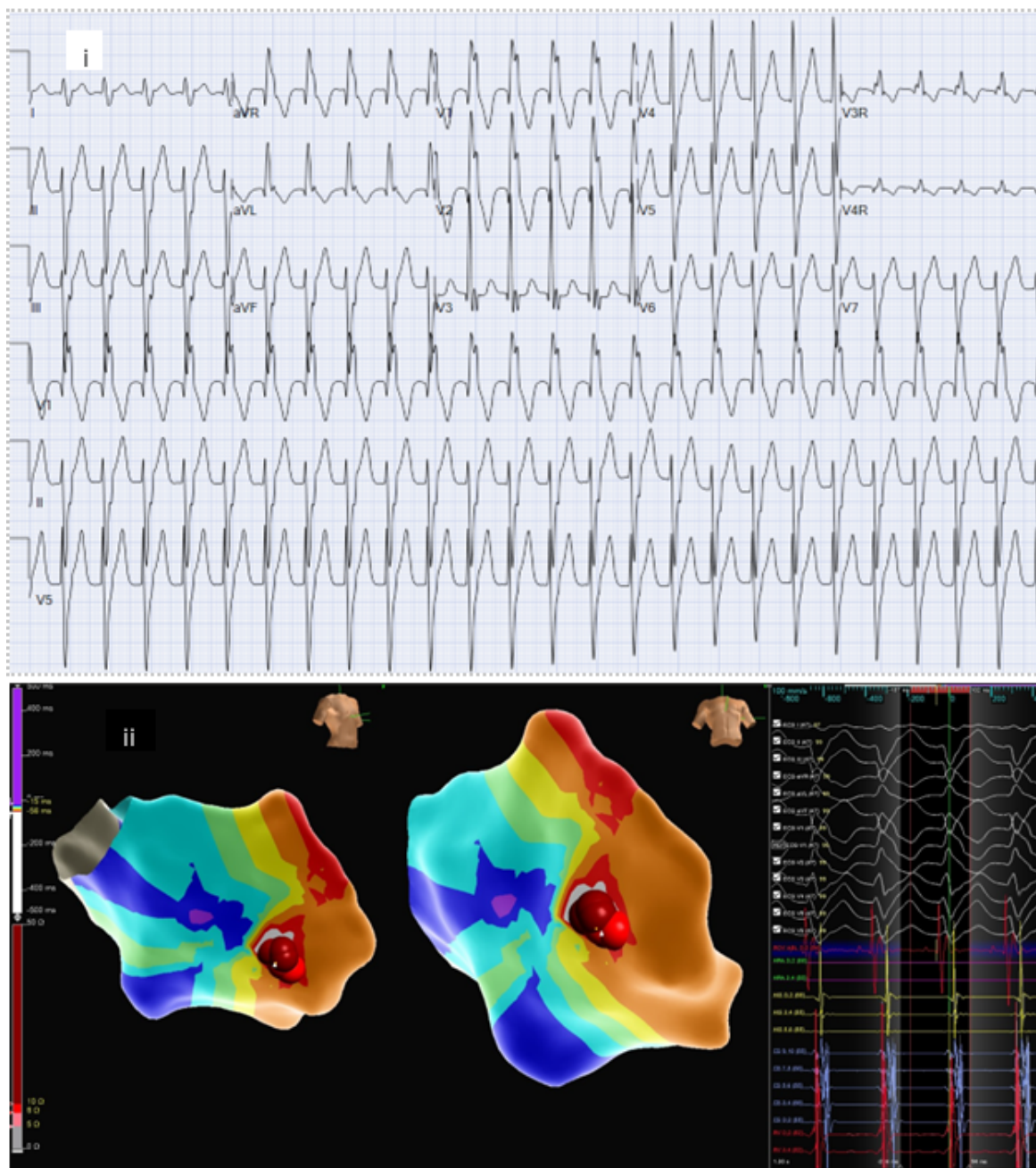
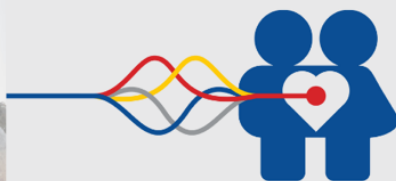
Our case series demonstrates the variable age of presentation of left posterior fascicular VT with a rare presentation in a 14-day old neonate. To our knowledge, this is the youngest patient reported with posterior fascicular VT. Our case series describes the variability in response to antiarrhythmic agents and highlights the successful use of alternate agents, especially in neonates where calcium channel blockers are typically avoided. Catheter ablation targeting the earliest Purkinje potential was effective in our patients demonstrating the efficacy of ablation as well as the safety even in younger patients who are < 25 kg.



Patient A with EKG showing tachycardia with RBB pattern and left axis deviation with VA dissociation (red arrows) consistent with posterior fascicular VT and QRS duration of 76 ms (i) and EKG obtained post conversion with amiodarone bolus, now with sinus rhythm and QRS duration of 68 ms (ii)



Patient B with wide complex tachycardia with RBB pattern and left axis deviation consistent with posterior fascicular VT and QRS duration of 136 ms (i) and activation map showing site of ablation (red dots) and termination of VT during ablation with intracardiac electrogram (ii)



Patient C with wide complex tachycardia with RBB pattern and left axis deviation consistent with posterior fascicular VT and QRS duration of 142 ms (i) and activation map showing site of ablation (red dots) with intracardiac electrogram (ii)



PO.20 | CLINICAL SPECTRUM, DIAGNOSTIC CHALLENGES, AND PHENOTYPIC OVERLAP OF RYR2-RELATED CHANNELOPATHIES IN PEDIATRIC PATIENTS: A CASE SERIES WITH FOCUS ON SUSPECTED CALCIUM RELEASE CHANNEL DEFICIENCY SYNDROME (CRDS)

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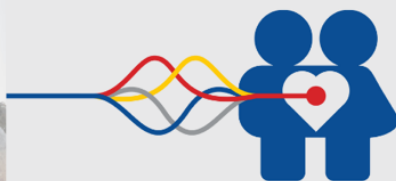
Background:

The cardiac ryanodine receptor type 2, encoded by the *RYR2* gene, is essential for calcium-induced calcium release in cardiomyocytes¹. Although *RYR2* variants are classically associated with catecholaminergic polymorphic ventricular tachycardia, loss-of-function variants have recently been linked to calcium release channel deficiency syndrome, a distinct and underrecognized arrhythmogenic disorder². We present three pediatric patients with *RYR2*-related arrhythmic phenotypes, highlighting clinical heterogeneity.

Case Presentations:

Case 1: A 16-year-old patient presented with sudden syncope during acute emotional stress and was found to be in asystole on emergency department arrival, requiring cardiopulmonary resuscitation. Resuscitation was complicated by two ventricular fibrillation episodes requiring defibrillation. Family history revealed sudden cardiac death in two siblings at approximately 18 years of age. Echocardiography showed a structurally normal heart. Genetic analysis identified a heterozygous *RYR2* variant, c.6515T>A, with documented loss-of-function effects on RyR2 channel activity. Calcium release channel deficiency syndrome was diagnosed. Propranolol was initiated, with no recurrent malignant arrhythmia during follow-up.

Case 2: A 12-year-old boy presented with exertional irregular heartbeats. Baseline Holter monitoring was normal. Exercise stress testing revealed rare premature ventricular complexes at 177 bpm, absent at peak exercise. Genetic analysis identified a heterozygous likely pathogenic *RYR2* variant, c.2104G>T. After propranolol initiation, repeat exercise testing showed complete suppression of exercise-induced ventricular ectopy.



Case 3: A 17-year-old girl with type 1 diabetes mellitus presented with exertional palpitations, dyspnea, and intermittent presyncope. Holter monitoring showed a low premature ventricular complex burden of approximately 1.3%. Echocardiography revealed mild ascending aortic dilatation and trivial aortic regurgitation. Exercise testing demonstrated monomorphic premature ventricular complexes beginning at 150 bpm and persisting throughout maximal exertion without progression. Genetic testing revealed a heterozygous *RYR2* variant of uncertain significance, c.980C>G. Verapamil partially reduced ventricular ectopy.

Conclusion:

This case series illustrates the broad phenotypic spectrum of *RYR2*-related arrhythmia syndromes in children, ranging from exercise-induced ectopy to aborted sudden cardiac arrest. *RYR2* variants should be considered in children with unexplained ventricular arrhythmias, exertional ectopy, syncope, or sudden cardiac arrest. Clinical phenotype, family history, exercise testing, and genetic interpretation are essential for diagnosis, treatment selection, and risk stratification.



PO.21 | WENCKEBACH PERIODICITY IN A PEDIATRIC PATIENT WITH COEXISTING TYPICAL AND ATYPICAL AVNRT: A CASE REPORT

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Background: Atrioventricular nodal reentrant tachycardia (AVNRT) is the most common supraventricular tachycardia in children¹. The coexistence of typical and atypical forms of AVNRT in the same procedure is rarely described in the literature². Here, a patient is presented in whom typical, atypical, and AVNRT with Wenckebach periodicity were induced in the same procedure.

Case summary: A 13-year-old girl underwent electrophysiological study (EPS). At first, non-sustained narrow-QRS tachycardia with an atrial-to-ventricular ratio greater than 1 and intermittent AV Wenckebach periodicity was repeatedly observed, initially raising suspicion for atrial tachycardia. However, reproducible adenosine-sensitive termination, induction of a long-RP tachycardia by ventricular pacing, and a ventricular overdrive pacing response supported (atypical) AVNRT. After cryoablation catheter positioning, typical AVNRT with a VA interval of 0 ms was also induced and terminated immediately during initial cryoapplication in the slow pathway region. Post-ablation programmed stimulation, both at baseline and under isoprenaline, failed to reinduce tachycardia.

Conclusion: This case illustrates an unusual pediatric presentation of AVNRT with multiple electrophysiological phenotypes during a single procedure, including typical AVNRT, atypical AVNRT, and A>V tachycardia with Wenckebach periodicity. Recognition of these rare manifestations is essential to avoid misdiagnosis as atrial tachycardia, prevent inappropriate ablation targeting, and ensure safe, effective slow pathway modification.





PO.22 | TRANSTHORACIC ECHOCARDIOGRAPHY-GUIDED TRANSSEPTAL PUNCTURE IN PEDIATRIC PATIENTS: A SINGLE-CENTER EXPERIENCE

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Background

In pediatric patients with left-sided accessory pathways, catheter ablation via the transseptal approach increasingly relies on intracardiac echocardiography (ICE) for guidance¹. However, ICE requires additional vascular access, extends procedure time, carries mechanical risks, and poses significant cost and availability challenges². This study evaluated the safety and feasibility of transthoracic echocardiography (TTE) as an accessible alternative for guiding transseptal puncture in a pediatric population.

Methods

We retrospectively analyzed 54 patients aged 0–18 years who underwent TTE-guided transseptal catheter ablation at Ankara Etlik City Hospital between November 2022 and April 2026. Demographic data, accessory pathway risk category, lesion location, procedural success, complication rates, and recurrence were assessed.

Results

Of 54 patients (62.3% male), the most common presenting symptom was palpitation (60.4%). Manifest pre-excitation was present in 62.2% and high-risk accessory pathways in 27%. Ablation was successful in 48 patients (88.9%), with a median procedure duration of 166 minutes. Median total radiation dose was 57.5 mGy. TTE-guided transseptal access was achieved successfully in all 54 patients (100%). One patient required conversion to a transaortic approach due to an epicardial pathway at the aortomitral continuity. Two patients (7.5%) experienced recurrence, both successfully treated with a repeat procedure.

Conclusion

TTE-guided transseptal puncture achieved a 100% access success rate in this pediatric cohort, comparable to published ICE-guided series. TTE guidance streamlined needle positioning by reducing the number of SVC-to-fossa ovalis dropping maneuvers and enabled real-time fine-tuning of catheter tip placement relative to the fossa ovalis margins. With a low complication profile and no requirement for additional vascular access, TTE represents a feasible, cost-effective, and readily available imaging modality for transseptal access in pediatric electrophysiology.



PO.23 | APPROACH TO EPICARDIAL PERMANENT PACEMAKER PLACEMENT IN THE PAEDIATRIC POPULATION

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Background: Permanent pacemaker insertion in paediatrics is complicated by smaller size, challenging vascular access, growth potential and congenital heart disease which may contraindicate transvenous systems. Epicardial systems minimise lead fracture/dislodgement, venous occlusion and tricuspid valve damage however there is no standardised approach to insertion in children. We describe our local approach.

Methods: Historically, our centre placed both the cathode and anode simultaneously on the right ventricle as it is the most easily accessed via lower sternotomy. Recently, changes were made to provide better synchrony and pump efficiency by placement on the left ventricle (LV) apex (Figure 1). Via left anterior thoracotomy, the cathode is placed on the LV apex in a site free from fibrosis/fat/infarction then tested unipolar; ideally where a narrow QRS is generated. Optimal cathode placement provides a safeguard for future unipolar pacing if required. The circuit is completed by attaching the lead to muscle/skin/surgical retractors. Adequacy of cathode lead placement is established before placing the anode, providing timely identification of a suboptimal lead. Once a good threshold is achieved on unipolar testing, the anode is affixed and the lead is tested bipolar. An elevated threshold implies the anode lead is suboptimal and requires revision. A high output test is undertaken to ensure no phrenic nerve capture. Use of steroid-eluting leads reduces inflammation and protects from exit block and threshold elevation. The pulse generator is placed in the subrectal sheath area with the redundant length of lead left in the chest cavity outside the pericardium allowing for somatic growth and reducing the risk of cardiac strangulation, coronary compression and bow stringing. The remaining length is placed behind the pulse generator in a smooth coil.

Results: This stepwise technique, used at our centre since 2021, has provided a systematic approach to epicardial pacemaker lead placement to the LV apex, better synchrony and pump efficiency (by placement on LV apex), and optimised lead parameters by efficient and timely identification in theatre of suboptimal electrode button placement.

Conclusions: Whilst controlled comparison of other techniques is challenging, we propose an efficient, systematic, standardised approach to epicardial bipolar lead placement in children.

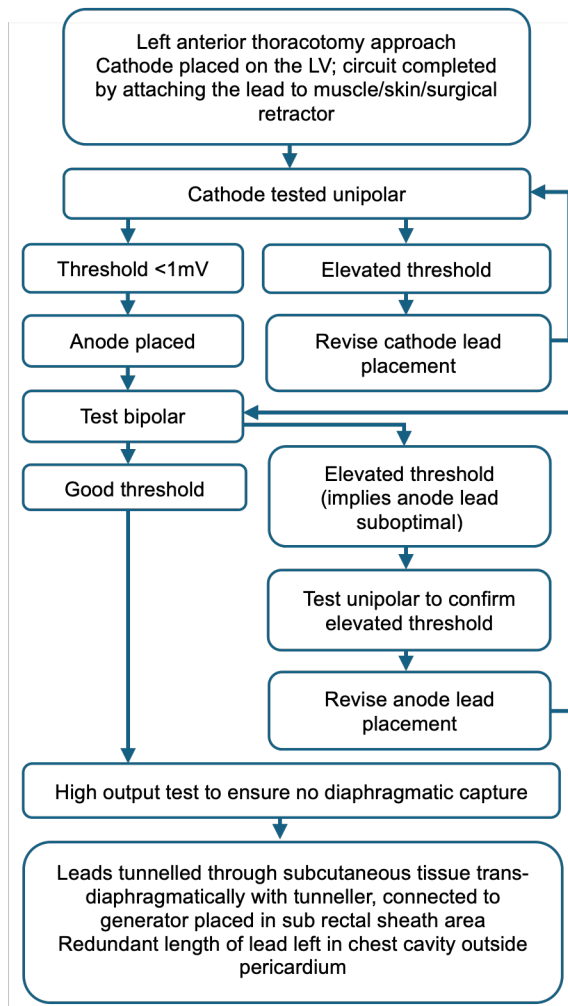


Figure 1: Systematic approach to epicardial pacemaker lead placement

**PO.24 | SCAR-RELATED VENTRICULAR ECTOPY IN A CHILD: REVERSAL OF VENTRICULAR DYSFUNCTION AFTER CATHETER ABLATION****Neeraj Nirmal¹, Camille-Marie Go-Cacanindin¹, Charalampos Kotidis², Vinay Bhole¹**¹ Paediatric Cardiology, Birmingham Children's Hospital² Paediatric Cardiology, East Midlands Congenital Heart Centre**Background:**

Scar-related ventricular arrhythmias are recognised in patients with congenital heart disease (CHD), most commonly following surgical repair. However, post-infarction myocardial scar causing frequent premature ventricular complexes (PVCs) in childhood is exceptionally rare. We present a unique case of high-burden scar-related PVCs resulting in left ventricular (LV) dysfunction, successfully treated with catheter ablation.

Case Presentation:

A 10-year-old boy (25 kg) with complex CHD, including TGA, ventricular septal defect (VSD), and coarctation of the aorta (CoA), underwent arterial switch operation, VSD closure, and CoA repair. His postoperative course was complicated by coronary artery injury requiring surgical revision. Subsequent cardiac catheterisation demonstrated myocardial infarction involving the inferior and septal LV walls, mild circumflex artery narrowing, and mild LV dysfunction. He later developed frequent monomorphic PVCs with a burden of 43%, associated with progressive LV systolic dysfunction. Imaging showed unobstructed epicardial coronaries, reduced LV ejection fraction (LVEF 35%), regional wall motion abnormalities, and a transmural scar in the mid-inferior LV.

Electrophysiology Study and Ablation:

An electrophysiology study was performed under general anaesthesia. Venous access was obtained via the left femoral vein because of right femoral vein occlusion. Electroanatomical mapping using CARTO (Biosense Webster) demonstrated low-voltage scar in the inferior and septal LV. Activation mapping identified the earliest activation of the clinical PVC at the mid-septum. Initial radiofrequency ablation (RFA) from the right ventricular septal aspect was unsuccessful, suggesting an intramural or left-sided substrate. LV access was achieved via a patent foramen ovale. Further mapping revealed earlier pre-potentials on the LV anterior septum, and targeted irrigated-tip RFA resulted in immediate PVC suppression and elimination.



Outcome:

PVC burden decreased from 43% to 1% following ablation. Follow-up demonstrated significant recovery of LV function, with LVEF improving from 35% to 45%.

Conclusion:

This case highlights the rare occurrence of post-infarction scar-related PVCs in a child with repaired TGA. The combination of young age, small body size, and infarct-mediated arrhythmogenic substrate underscores its uniqueness. It demonstrates the value of detailed electrophysiological mapping in identifying complex intramural circuits and the effectiveness of catheter ablation in reversing PVC-induced cardiomyopathy.

**PO.25 | ATROPINE AS A THERAPY APPROACH TO REDUCE THE FREQUENCY OF RESPIRATORY SPELLS WITH SINUS ARREST****Till Schöchtner¹, Andreas Tulzer¹, Christoph Prandstetter¹, Florian Putre¹**¹ Children's Heart Center Linz, Kepler Universitätsklinikum Linz**Background and Aim:**

Breath-holding spells and reflex anoxic seizures are common benign paroxysmal events in infancy and early childhood and are generally associated with an excellent long-term prognosis. However, severe cardioinhibitory forms with prolonged asystolic pauses are uncommon, particularly in very young infants. We report a 4-month-old infant with recurrent cyanotic respiratory spells associated with prolonged asystole and describe the successful use of atropine as a non-invasive therapeutic approach.

Methods:

A 4-month-old female infant was referred for evaluation of recurrent cyanotic spells that had increased in frequency over four weeks. ECG monitoring at an outside hospital documented asystolic pauses of up to 5 seconds. Further evaluation included a 72-hour Holter-ECG, ECG, echocardiography, neurological assessment, and clinical follow-up. Family history was negative for cardiovascular disease and sudden cardiac death. Developmental history and neurological examination were age appropriate.

Results:

The 72-hour Holter ECG demonstrated recurrent bradycardia with asystolic pauses lasting up to 8 seconds. Echocardiography showed no structural heart disease apart from a hemodynamically insignificant patent foramen ovale. Neurological assessment was unremarkable. Based on the suspected vagally mediated cardioinhibitory mechanism, oral atropine was initiated at 5 µg/kg/day in three doses. Following treatment initiation, no further respiratory spells or asystolic episodes occurred during inpatient monitoring. Given the presumed functional and potentially transient nature of the condition, the patient's young age, and the absence of persistent bradycardia outside the episodes, primary pacemaker implantation was not pursued. Home monitoring, parental cardiopulmonary resuscitation training, and close pediatric cardiology follow-up were arranged.



Conclusions:

This case highlights the importance of rhythm evaluation in infants presenting with recurrent cyanotic spells, even in the absence of structural heart disease. Severe vagally mediated cardioinhibitory episodes with prolonged asystole may occur at a very young age. Oral atropine may represent an effective non-invasive treatment option in selected patients and may allow avoidance of permanent pacing. Despite the severity of presentation, the overall long-term prognosis of reflex cardioinhibitory events in infancy is generally favorable.

**PO.26 | SMALLER HARDWARE WHEN THE SMALLEST PATIENTS NEED PACING: EARLY MULTICENTER EXPERIENCE FROM THE NEOPACE STUDY**

Paola Ferrari¹, Giovanni Malanchini¹, Davide Akira Garattini¹, Cristina Leidi¹, Charalampos Kotidis², Vinay Bhole³, Camille-Marie Go³, Christophe Vô⁴, Shubhayan Sanatani⁵, Sakethram Saravu Vijayashankar⁵, Luca Tomasi⁶, Joaquin Osa⁷, Francesca Papesso⁸, Paolo De Filippo¹

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Background: Permanent pacing in neonates presents substantial challenge due to extremely small body size, anatomic variability and limited device options; even the smallest conventional pacemakers remain disproportionally large for newborns < 3 kg. To overcome these limitations, a novel pediatric-sized implantable pulse generator, derived from a modified leadless pacemaker subassembly and connected to bipolar epicardial leads, has been introduced. Evidence for its clinical use in this population is still limited. We here propose the first report from its international use.

Methods: NeoPACE study is a multicenter observational registry collecting real-world data on neonates requiring permanent pacing and implanted with the new Implantable Pulse Generator device. Eleven consecutive patients were enrolled across seven international centres between March 2023 and October 2025. Demographic data (mean gestational age 33.9 ± 3.0 weeks, mean weight 1901.8 ± 564.3 g), clinical history and procedural details were collected. Follow-up was available for all patients, with a median duration of 8.1 months (IQR 3.0–13.1, 1 - 22.7 months).



Results: The main indication for pacing was congenital AV block (10 patients, 90.9%, including 5 cases attributed to maternal circulating antibodies for autoimmune disease), one patient had severe sinus node dysfunction (9.1%). Every patient received a single chamber epicardial system with ventricular lead. Mean body weight at time of implant was $1901,8 \pm 564,3$ g. Procedural success was achieved in all cases. Median age at implantation was 40 days (0 - 95 days), including one newborn implanted at birth. Mean procedure time was 106.6 ± 44.9 minutes. At implant, mean capture threshold was 0.75 ± 0.50 V x 0.40 ms, R-wave amplitude 10.1 ± 5.2 mV, and impedance 785.5 ± 328.7 Ω . Electrical performance remained stable during follow-up. No acute complications or procedure-related mortality occurred. Adverse events were reported in 2 patients (18.2%), both related to epicardial lead dysfunction. Two required surgical revision (at 5 and 15 months). In 2 other patients, a progressive increase in capture threshold up to 1.88 V x 0.4 ms was observed. One was successfully managed by device reprogramming; while the other underwent battery replacement 10 months after implant. All patients remained on VVI pacing (10 at 110 bpm, 1 at 80 bpm, 1 at 70 bpm as lower rate), with 99-100% ventricular pacing in 9 cases (81.8%). All patients demonstrated appropriate growth and device tolerance during follow-up. No skin erosions or pocket infections was observed.

Conclusions In this multicenter experience, implantation of the novel implantable pulse generator was feasible and acutely safe. Mid-term performance was stable. These early results confirm feasibility of the use of the device and underscore the need for continued long term evaluation and device refinement, specifically tailored to this highly vulnerable population.

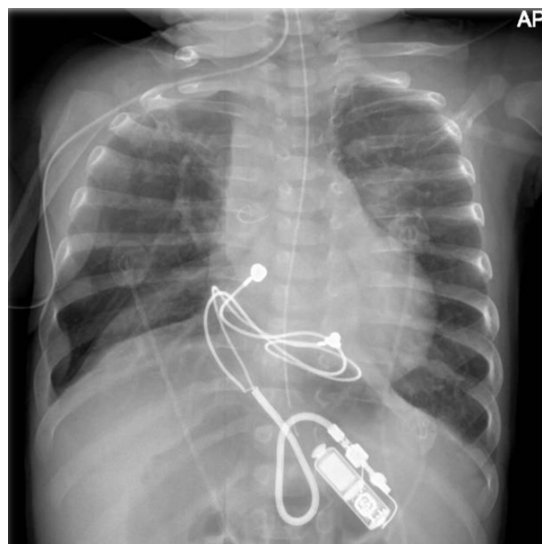




Table 1. Baseline Characteristics, Procedural Data, and Outcomes of the NeoPACE Cohort

Category	Variables	Results
Patient Characteristics	Number of patients	11
	Gestational age at birth, mean $\pm$ SD	33.9 $\pm$ 3.0 weeks
	Birth weight, mean $\pm$ SD (g)	1901.8 $\pm$ 564.3
	Age at implant, median (IQR), days	40 (30–48)
	Age range at implant, days	0–95
Indications for Pacing	Congenital atrioventricular block	10 (90.9%)
	– associated with maternal autoantibodies	5 (45.5%)
	Severe sinus node dysfunction	1 (9.1%)
Device and Implant Characteristics	Pacing system	Single-chamber epicardial VVI
	Ventricular lead configuration	Single bipolar epicardial lead
	Procedural success	11/11 (100%)
	Procedure duration, mean $\pm$ SD (min)	106.6 $\pm$ 44.9
	Implant at birth	1 patient
Electrical Parameters at Implant	Capture threshold, mean $\pm$ SD (V $\times$ ms)	0.75 $\pm$ 0.50 V $\times$ 0.40 ms
	R-wave amplitude, mean $\pm$ SD (mV)	10.1 $\pm$ 5.2
	Lead impedance, mean $\pm$ SD (Ω)	785.5 $\pm$ 328.7
Follow-Up	Median follow-up duration, months (IQR)	8.1 (3.0–13.1)
	Follow-up range, months	1–22.7
	Ventricular pacing percentage	99–100% in 9 patients (81.8%)
	Lower pacing rate	110 bpm (10 pts); 80 bpm (1 pt); 70 bpm (1 pt)
Complications and Reinterventions	Acute complications and mortality	None
	Adverse events overall	2 patients (18.2%)
	Lead-related dysfunction	2 patients
	Surgical lead revision	2 cases (at 5 and 15 months)
	Increased capture threshold requiring intervention	2 cases (up to 1.88 V $\times$ 0.4 ms)
	– managed by reprogramming	1 case
	– requiring generator replacement	1 case (after 10 months)
Clinical Outcomes	Growth and somatic development	Appropriate in all patients
	Device tolerance	Good; no skin erosions
	Pocket infections	None



PO.27 | LONG-TERM OUTCOME OF ACCESSORY PATHWAY CATHETER ABLATION IN PEDIATRIC POPULATION WITH VENTRICULAR PREEXCITATION. SINGLE CENTER EXPERIENCE.

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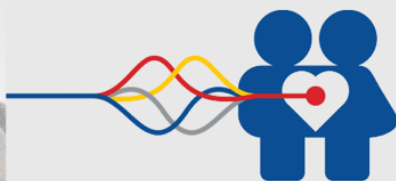
Background and Aim: Radiofrequency catheter ablation (RFCA) of accessory pathways (AP) is first-line therapy in the pediatric population with symptomatic and/or high-risk ventricular preexcitation (PE). This study aimed to determine long-term ablation success and predictors of recurrence.

Methods: This retrospective single-center study included 185 consecutive patients (mean age 16 years) with PE who underwent RFCA under general anesthesia at one tertiary care center over a 10-year period (2015–2024). Fluoroscopy and a 3D electroanatomic mapping system were used in all patients. Non-irrigated ablation catheters were used initially. No "consolidation" RF pulses (anatomically guided additional applications) were delivered at the site of AP conduction block. After ablation, patients were observed at least 30 minutes and tested for residual conduction using intravenous isoprenaline and adenosine. After discharge, patients were followed (median 21 months) for AP conduction and/or recurrent AP-mediated tachycardias via at least annual clinical examination, including standard ECG and 24-h ambulatory ECG monitoring. Factors analyzed for association with recurrence were age, weight, AP localization, number of RF applications, time from RF onset to AP conduction block, and use of an irrigated-tip catheter.



Results: The 185 patients underwent 213 ablation procedures over 10 years. Acute success with bidirectional AP conduction block was achieved in 182 patients (98.4%). Of 41 recurrences, 22 occurred within 24 hours post-ablation, and delayed conduction block was observed in 3. The final recurrence rate was 20.8% (38/182). However, 12 patients showed only asymptomatic, intermittent, low-risk PE, leaving clinically relevant recurrences in 26/182 patients (14.3%). Factors associated with recurrence were: (1) irrigated catheter deployment after failed non-irrigated attempts ($p=0.0003$); (2) ablation at the site of mechanical block ($p=0.0065$); (3) delivery of ≥ 3 RF pulses to achieve AP block (trend, $p=0.07$); and (4) AP location other than left free wall (trend, $p=0.06$). Twenty-eight patients with symptomatic and/or permanent PE underwent a successful second RFCA. Overall long-term clinical success was achieved in 98.3%.

Conclusion: In this consecutive pediatric cohort with ventricular PE, the acute success rate was 98.4%. The overall recurrence rate after first RFCA was 21%, 14% being clinically relevant. With repeat ablations, the long-term clinical success rate reached 98.3%.



PO.28 | REVERSIBLE COMPLETE AV BLOCK AS AN UNUSUAL MANIFESTATION OF “HOT PHASE” ARRHYTHMOGENIC CARDIOMYOPATHY: CASE REPORT

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Background and Aim

Arrhythmogenic cardiomyopathy (ACM) is a genetic myocardial disease associated with ventricular arrhythmia and sudden cardiac death. Although typically chronic, some patients develop acute myocarditis-like “hot-phase” episodes. We report a rare pediatric case of genetically confirmed ACM presenting with transient complete atrioventricular block during a hot-phase episode, a rare manifestation of the disease.

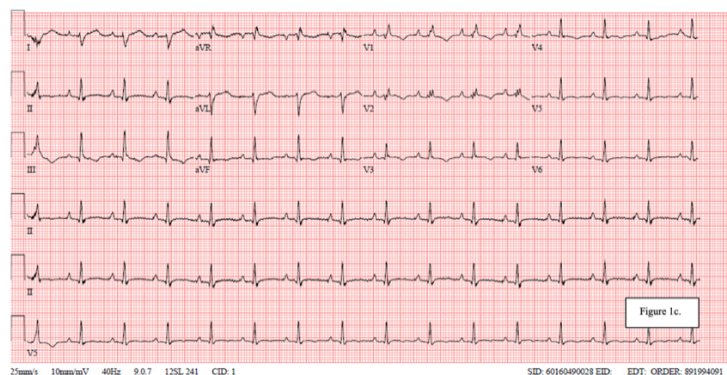
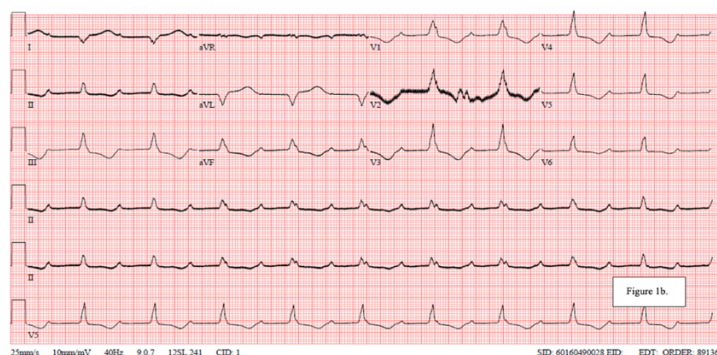
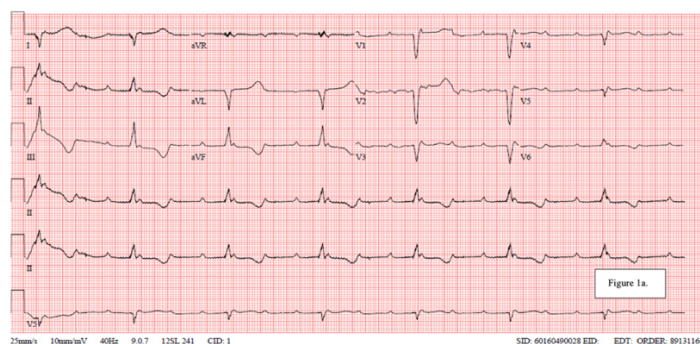
Case Report

A 12-year-old girl with a family history of sudden cardiac death in a sibling, was diagnosed with arrhythmogenic cardiomyopathy carrying a homozygous *DSG2* mutation during family screening and presented with presyncope and chest pain. Following marked troponin elevation, she was referred to our center, where bradycardia and progressive atrioventricular block were identified on the ECG (**Figure 1a**). On admission, inflammatory markers and viral studies were negative, while cardiac enzymes were profoundly elevated. Cardiac MRI revealed extensive right ventricular fibrosis and dyskinesis, newly developed mid-septal edema (T2 hyperintensity), and patchy biventricular late gadolinium enhancement (**Figure 2**), findings that were consistent with an acute inflammatory episode. During intensive care follow-up, the heart rate decreased to 40 bpm, and serial ECGs demonstrated progressive conduction system involvement with bifascicular block and QRS widening (**Figure 1b**). Temporary external pacing was initiated due to the risk of hemodynamic deterioration. Prednisolone therapy was initiated for hot-phase myocardial inflammation and presenting with conduction system involvement. By day three, a decline in cardiac enzyme levels was accompanied by recovery of sinus rhythm (**Figure 1c**), and pacing support was successfully discontinued. Given her high-risk profile, an ICD was surgically implanted on day 8. The patient recovered uneventfully and was discharged after the normalization of cardiac enzymes and tapering of steroid therapy (**Figure 3**).



Conclusion

This case highlights transient high-degree atrioventricular block as a rare manifestation of the hot phase of ACM. Clinicians must recognize that inflammatory episodes can affect the cardiac conduction system, particularly in genetically confirmed diseases. Such manifestations may indicate a more aggressive phenotype, necessitating prompt diagnosis and treatment. While conduction abnormalities can resolve with anti-inflammatory therapy, evaluation of arrhythmic risk and device therapy remains critical. Recognizing these phases is essential for optimizing therapeutic strategies and preventing adverse outcomes.



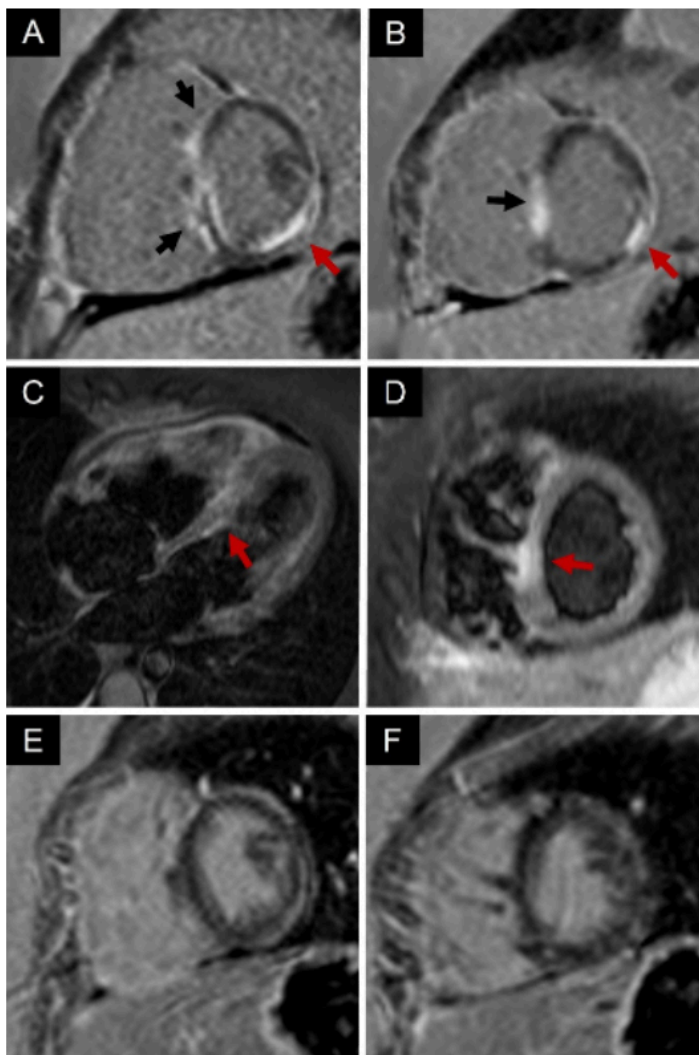


Figure 2: Short-axis PSIR images obtained at the basal-to-midventricular junction (A) and midventricular level (B) show extensive patchy late gadolinium enhancement (LGE), most pronounced within the septum (black arrows) and inferolateral wall (red arrows), extending to the full myocardial thickness. There is also diffuse LGE involving the right ventricle. Four-chamber (C) and short-axis (D) T2-weighted fat-suppressed images demonstrate accompanying septal myocardial edema (arrows). On the corresponding short-axis PSIR images from 2024 (E, F), no extensive septal LGE was present, apart from subepicardial enhancement involving the anterolateral and inferolateral segments.

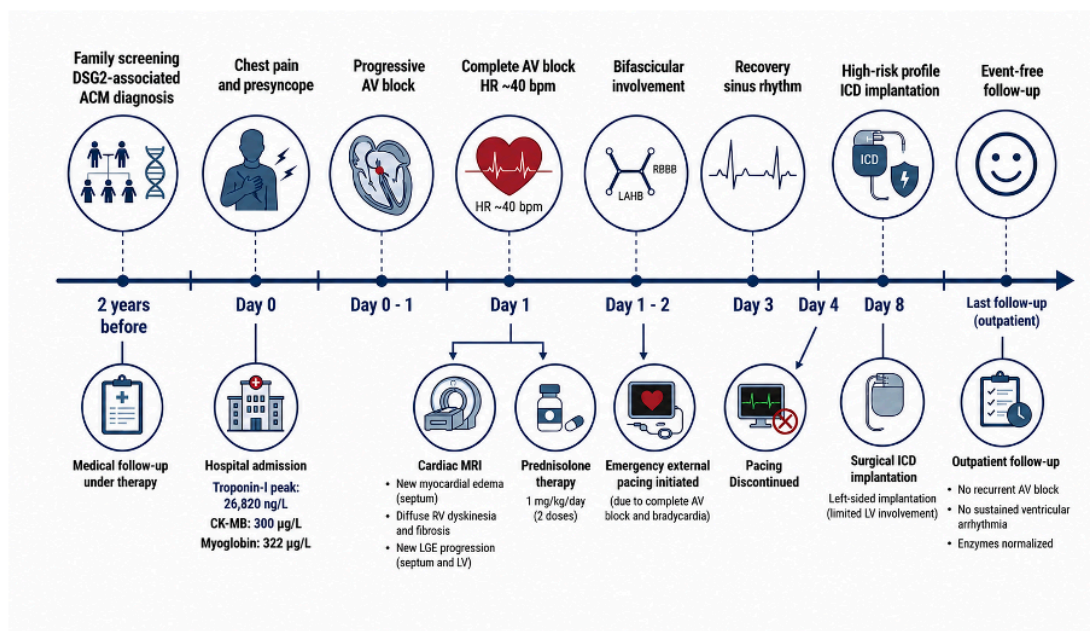
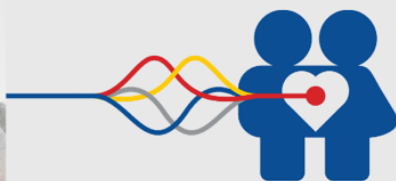


Figure 3: Clinical timeline of hospitalization showing the evolution of atrioventricular conduction disturbances, cardiac MRI findings, corticosteroid treatment, temporary external pacing support, recovery of sinus rhythm, and subsequent surgical ICD implantation. (ACM, arrhythmogenic cardiomyopathy; AV, atrioventricular; RBBB, right bundle branch block; LAHB, left anterior hemiblock; LGE, late gadolinium enhancement; RV, right ventricle; LV, left ventricle; ICD, implantable cardioverter-defibrillator; HR, heart rate.)

**PO.29 | ATRIAL TACHYCARDIA IN AN INFANT WITH UNREPAIRED TETRALOGY OF FALLOT – IS EARLY ARRHYTHMOGENESIS A REASONABLE EXPLANATION?****Lilia Bardarska¹, Elissaveta Levunlieva², Dimitar Pechilkov², Anna Kaneva-Nentcheva²**¹ Pediatric Cardiology, National Heart Hospital, Sofia, Bulgaria² Pediatric Cardiology, National Heart Hospital, Sofia, Bulgaria

Background: Arrhythmias are rare in unrepaired tetralogy of Fallot (ToF) patients before the first year of life mainly associated with electrolyte imbalance or disease progression.

Case Summary: A 2.5-month-old boy diagnosed as a newborn with ToF was scheduled for palliative surgery because of hypoxic spells. Modified Blalock-Taussig (BT) shunt was performed under general anesthesia. On the 17th postoperative day, irregular narrow complex tachycardia was observed and persisted after adenosine-induced transient AV block (Figure 1 and 2). Sinus rhythm was restored with amiodarone. The child experienced another tachycardia episode 1.5 months after the surgery suppressed again by intravenous amiodarone infusion and dose adjustments (Figure 3). Corrective surgery was performed at the age of 11 months. No arrhythmia was documented during the early and intermediate postoperative period. Amiodarone therapy was withdrawn 6 months after the surgical repair and no tachycardia recurrence was documented on follow-up ECGs (Figure 4) and Holter monitoring.

Discussion: Tachyarrhythmias are rare in infants with ToF despite in the perioperative period and occasionally reported after BT shunt. Few studies suggest arrhythmogenic remodeling caused by the disease itself.

Take home messages: Our case highlights the importance of arrhythmia in ToF patients. Remodeling could begin as early as infancy and is not directly linked with the hemodynamic state.



Disease Timeline

Time	Age	Event
Feb. 2025		Full term. Normal postnatal adaptation
March 2025	1 month	Cardiac murmur. Diagnosis: Tetralogy of Fallot
March-April	1-1.5 months	Hypoxic spells
April 2025	1.5 month	Echo+CT: hypoplastic pulmonary arteries with McGoon index 130-140%. Small aorto-pulmonary collaterals.
May 2025	2.5 months	Modified Blalock-Taussig anastomosis 3.5 mm
May 2025	2.5 months (17 th postoperative day)	Irregular narrow complex tachycardia
June 2025	4 months	Amiodarone dose reduction (5 mg/kg)
July 2025	5 months	Arrhythmia recurrence. Sinus rhythm restored with intravenous amiodarone.
August 2025	6 months	Amiodarone dose adjustment (5 mg/kg)
August – December 2025	6-10 months	Stable condition. No hypoxic spells. No arrhythmia. SpO2 80-90%
January 2026	11 months	Corrective surgery. No documented arrhythmia.
Present	1 year 6 months	Therapy withdrawn, no arrhythmia on ECG and Holter

Figure 1: Narrow complex tachycardia 190-200 bpm. Limb leads only.

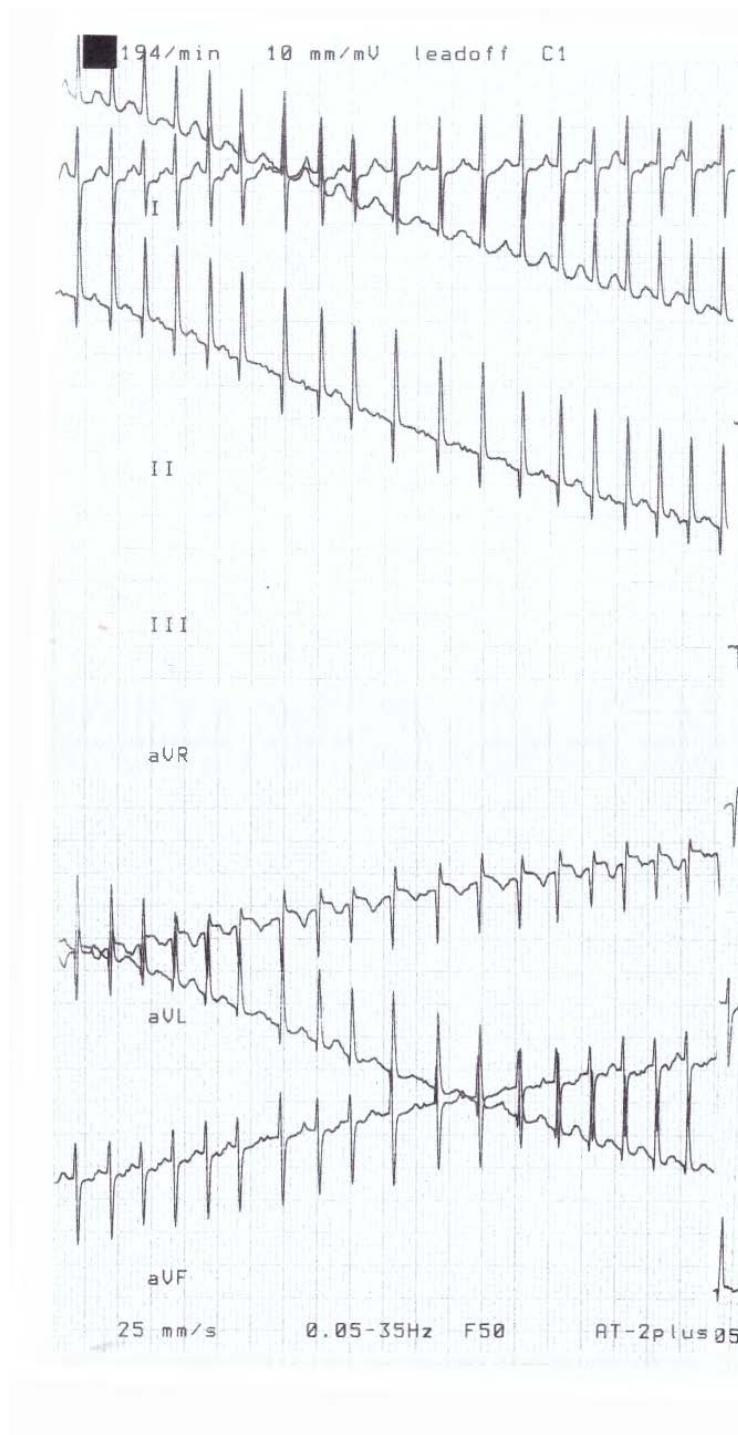


Figure 2: Adenosine bolus

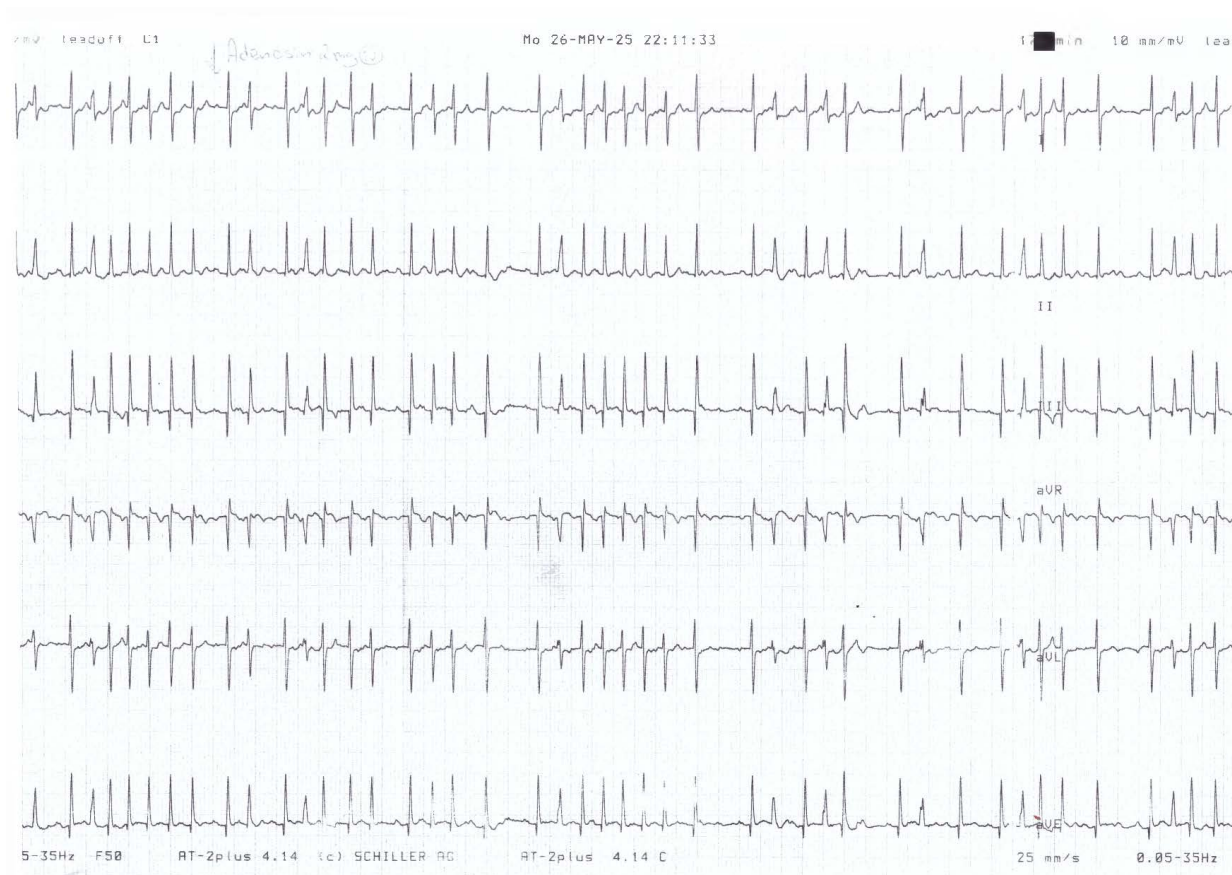


Figure 3: Irregular tachycardia interrupted by single sinus beats (arrow) – limb leads



PO.30 | EVALUATION AND MANAGEMENT OF CLINICALLY SIGNIFICANT NEONATAL TACHYARRHYTHMIAS

Lilia Bardarska¹, Anna Kaneva-Nentcheva¹, Dimitar Petchilkov²

¹ Pediatric Cardiology, National Heart Hospital, Sofia, Bulgaria

² Pediatric Cardiac Intensive Care Unit, National Heart Hospital, Sofia, Bulgaria

Background: Tachyarrhythmias during the neonatal period are uncommon, yet they represent potentially life-threatening conditions. This study aimed to analyze the clinical presentation, arrhythmia types, therapeutic responses, maternal risk factors, and long-term recurrence rates in a cohort of neonates.

Methods: We conducted a retrospective, descriptive case series study of all newborns with tachyarrhythmias admitted to our department between 2011 and 2022. Clinical histories, management strategies, and outcomes were reviewed.

Results: Twenty-five neonates (56% male) were identified for the studied period. Supraventricular tachycardia (SVT) was the sole arrhythmia observed. In seven cases (28%), tachycardia had been identified and managed prenatally. Structural heart disease was diagnosed in 28% of patients. Maternal risk factors (diabetes, infection, arterial hypertension) were present in 28% of cases. The majority of patients (88%) presented with clinical symptoms of heart failure, and 28% required intensive care (median stay: 3.7 days). Amiodarone was the primary antiarrhythmic agent used (n=18), proving effective as monotherapy in 39% of cases; the remainder required combination therapy. Electrical cardioversion was utilized in six patients, successfully restoring sinus rhythm in three of them (50%). Adverse events related to high-dose amiodarone were documented in six neonates. Sinus rhythm was achieved in 84% of patients. Although 36% experienced recurrence during the first year of life, only 16% exhibited persistent arrhythmia after the age of one.

Conclusions: While clinically significant neonatal tachyarrhythmias are rare, they often require intensive care, multi-drug management in the acute phase, but the long-term prognosis remains favorable. Spontaneous resolution is common after infancy, and catheter ablation remains a viable option for persistent cases once the patient reaches an appropriate weight.



Table 1. Demographic and clinical characteristics	
Birth weight, g	3123 (2100-3900)
Prenatal diagnosis, n (%)	7 (28)
Structural heart disease, n (%)	7 (28)
Congenital heart defects, n	6
Cardiomyopathy, n	1
Symptoms	
None, n (%)	3 (12)
Heart failure, n (%)	22 (88)
Impaired LV Function, n (%), mean EF (%)	8 (32), (45.5)
Hospital stay, mean (distribution)	14.2 days (3-57)
ICU stay n (%), days mean (distribution)	7 (28), 3.7 days (1-7)
Sinus rhythm restored, n (%)	21 (84)
Recurrence during the first year, n (%)	9 (36)
Recurrence after the age of one, n (%)	4 (16)

Table 2. Treatment Regimens									
In-Hospital	Vagal man.	Adenosine	Amiodarone	Digoxin	Beta blocker	Amio+Dig	Amio+BAB	Amio+Flec	DC Cardioversion
N of patients	3	8	7	2	1*	4	4	1	6**
Dose	NA	0.1-0.3 mg/kg	10-28 mg/kg loading	30-35 mcgr/kg loading	1-3 mg/kg	10-30 mg/kg+ 30-35 mcgr/kg	10-30 + 1-3 mg/kg	15+5 mg/kg	1-2 J/kg
Post discharge	No therapy	Amiodarone	BAB	Digoxin	Flecainide	Amio+BAB	Amio+Dig		
N of patients	1	9	2	2	1	9	1		
Dose	NA	5-10 mg/kg	1-3 mg/kg	10 mcgr/kg	5 mg/kg	5-16+1-3 mg/kg	10 mg/kg+ 10 mcgr/kg		

**PO.31 | LONG-TERM OUTCOMES OF SLOW PATHWAY LATE ACTIVATION MAPPING–
GUIDED CRYOABLATION FOR ATRIOVENTRICULAR NODAL REENTRANT TACHYCARDIA****Wei-Chieh(Wayne) Tseng¹, Shuenn-Nan Chiu¹, Chun-Wei Lu¹, Mei-Hwan Wu¹**¹ Pediatric Cardiology, National Taiwan University Children's Hospital**Background and Aim:**

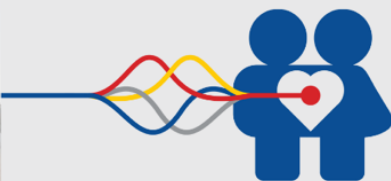
Slow pathway late activation mapping (SPLAM) has been shown to facilitate localization of optimal ablation sites during cryoablation for atrioventricular nodal reentrant tachycardia (AVNRT). However, the long-term outcomes of this novel mapping strategy remain unclear. This study aimed to evaluate the long-term efficacy and safety of SPLAM-guided AVNRT cryoablation.

Methods:

All consecutive patients who underwent AVNRT cryoablation between January 2017 and December 2025 were retrospectively enrolled. Conventional procedures performed between January 2017 and May 2020 served as the control group, whereas procedures using SPLAM to identify the wave-collision region after June 2020 comprised the study group. Patients with moderate or severe congenital heart disease complexity, as well as simple congenital defects requiring surgical intervention, were excluded.

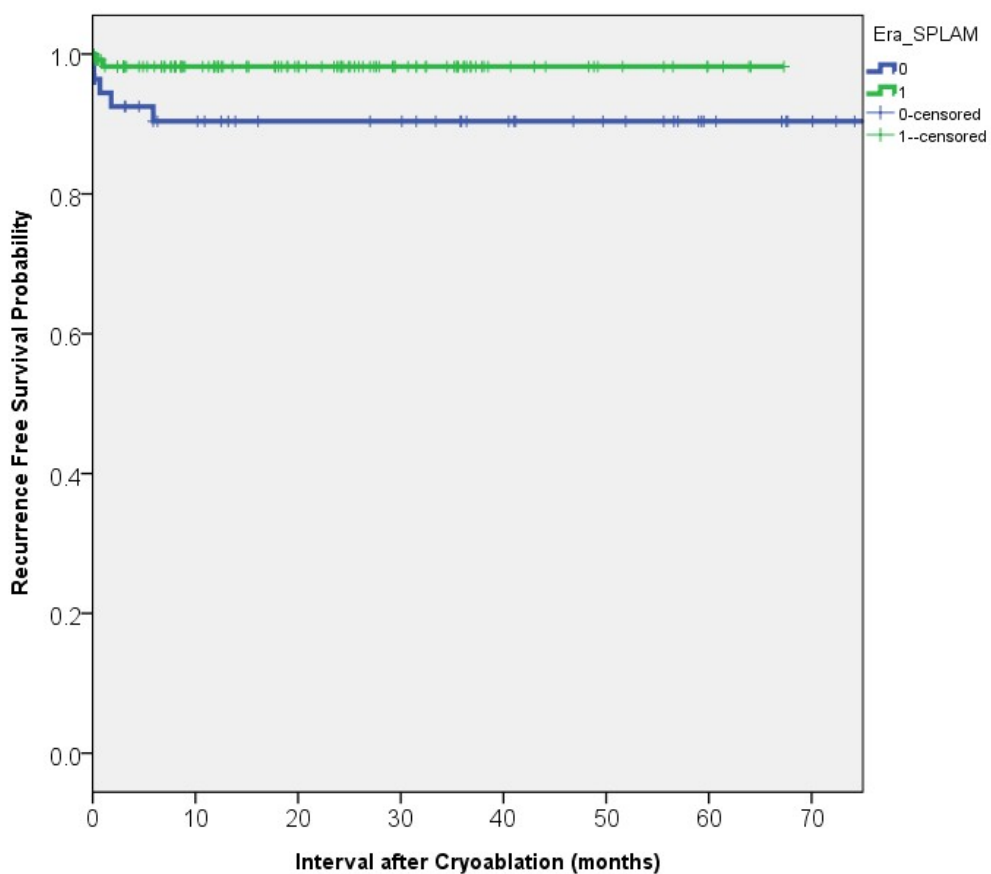
Results:

A total of 193 AVNRT cryoablation procedures were performed during the study period. After exclusion criteria were applied, 182 procedures were included for analysis, including 59 in the control group (30 females, 29 males) and 123 in the study group (64 females, 59 males). The mean age was 15.1 years in the control group and 16.3 years in the study group ($p = .328$). Mean procedure duration was 97.1 minutes in the control group and 105.3 minutes in the study group ($p = .374$). Excluding 9 procedures in which multiple arrhythmia mechanisms were ablated during the same procedure (2 in the control group and 7 in the study group), the mean procedure duration was 97.2 minutes in the control group and 99.8 minutes in the study group, respectively ($p = .754$). Acute procedural success was achieved in 100% of patients in both groups. Kaplan–Meier survival analysis demonstrated 1-year and 5-year recurrence-free survival rates of 90.4% and 90.4% in the control group, compared with 98.2% and 98.2% in the study group, respectively (log-rank test, $p = .023$). No patient developed atrioventricular block during follow-up.



Conclusions:

SPLAM-guided cryoablation enables more precise localization of slow pathway ablation targets in AVNRT. This strategy was associated with significantly lower long-term recurrence rates while maintaining excellent procedural safety.





PO.32 | PHENOTYPE OF TRUNCATING FILAMIN C VARIANT CARDIOMYOPATHY IN CHILDHOOD: A TWO-FAMILY CASE SERIES

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¹Paediatric Inherited Cardiac Conditions Team, Royal Brompton and Harefield Hospitals, London

Background and aim:

This case series describes 6 paternally-inherited Filamin C genotype-positive patients from two families, diagnosed in childhood. Their clinical course has been documented to understand similarities and differences compared to adult counterparts.

Methods:

Data was retrospectively collected from an electronic patient record on patient demographics, genetic variant, age and mode of first pathological finding, serial investigation data, medical therapy and ICD status.

Results:

All FLNC variants were truncating. Two patients from Family 1 had pathogenic FLNC c.7731C>A variants and four from Family 2 had pathogenic FLNC c.4127+1G>T variants. One patient also carried a paternally-inherited a VUS in DES.

Three patients were symptomatic at diagnosis. One experienced chest pain during a myocarditis-like presentation with oedema and inferolateral subepicardial enhancement, one dizziness and the third retrospectively reported episodes of dizziness whilst fishing.

The first pathological finding in all patients was fibrosis identified on cMRI between the ages of 9 and 17. The most striking finding was the development and progression of fibrosis on CMR. Most patients initially had normal studies before developing sometimes rapidly progressive fibrosis.

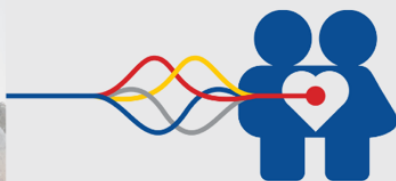


Abnormalities on baseline ECG, echo, Holter and exercise testing did not reliably correlate with degree of fibrosis on MRI or predict ventricular arrhythmias. One patient had NSVT and increasing ventricular ectopy during exercise correlating with more extensive mid-wall fibrosis. Another received an appropriate ICD shock for VT despite no preceding concerns on ECG, Holter or exercise testing. Signal averaged ECG performed later in his clinical course showed abnormal values. There was no extracardiac phenotype in our patient group.

Four of six patients underwent ICD implantation. One has received both appropriate and inappropriate shocks. Only one patient is treated with a beta blocker and none developed ventricular dysfunction.

Conclusion:

Our paediatric patients demonstrated an early scar-positive phenotype with fibrosis developing from late childhood and preceding any chamber dilatation, ventricular dysfunction, significant arrhythmia or symptoms. These findings suggest that truncating FLNC variants may produce clinically important arrhythmogenic substrates during childhood and adolescence and CMR is important to identify fibrosis preceding the development of the classic arrhythmic and structural changes seen in adult cohorts.

**PO.33 | LONG-TERM OUTCOMES AND APPROPRIATE ICD THERAPIES IN PEDIATRIC HCM: ARE WE MISSING HIGH-RISK PATIENTS?**

Michele Lioncino¹, Massimo Stefano Silvetti¹, Giulia Garosi¹, Sara Di Marzio¹, Nicoletta Cantarutti², Marianna Cicienia¹, Fabio Anselmo Saputo¹, Rachele Adorisio², Fabrizio Drago¹

¹ Paediatric Cardiology and Cardiac Arrhythmias Complex Unit, Bambino Gesù Children's Hospital IRCCS, Piazza Sant'Onofrio, 00165 Rome, Italy.

² Advanced Cardiovascular Therapy Unit, Bambino Gesù Children's Hospital IRCCS, Piazza Sant'Onofrio, 00165 Rome, Italy.

Background: Implantable cardioverter defibrillators (ICDs) are recommended for sudden cardiac death prevention in children with hypertrophic cardiomyopathy (HCM). However, pediatric data on long-term efficacy and device-related complications remain limited.

Methods: We retrospectively analyzed children with HCM undergoing ICD implantation at a tertiary center. Appropriate therapies were defined as ICD shocks delivered for ventricular tachycardia (VT) or ventricular fibrillation (VF). The predictive performance of the HCM Risk-SCD-Kids score was evaluated using receiver operating characteristic analysis. Progression of hypertrophy was assessed as the difference between baseline and follow-up left ventricular maximal wall thickness z-scores.

Results Twenty-eight children underwent ICD implantation at a median age of 13 years. Twenty-six patients (93%) received an ICD for primary prevention and 2 (7%) for secondary prevention. Ten patients received transvenous ICDs and eighteen subcutaneous ICDs. Median follow-up was 3.8 years (IQR 1.4–5.7).

Seven patients (25%) experienced appropriate ICD therapies. Five episodes were triggered by VF and two by sustained VT. Patients receiving appropriate therapies were younger at implantation (11.0 vs 15.0 years, $p=0.042$), despite longer follow-up duration (7.3 vs 2.3 years, $p=0.021$). Kaplan-Meier analysis demonstrated a significantly higher incidence of appropriate ICD therapies among secondary prevention patients (log-rank $p=0.0005$). Of note, both patients implanted for secondary prevention exhibited exercise-induced ECG abnormalities and advanced diastolic dysfunction, whereas their estimated HCM Risk-Kids score was $<6\%$.

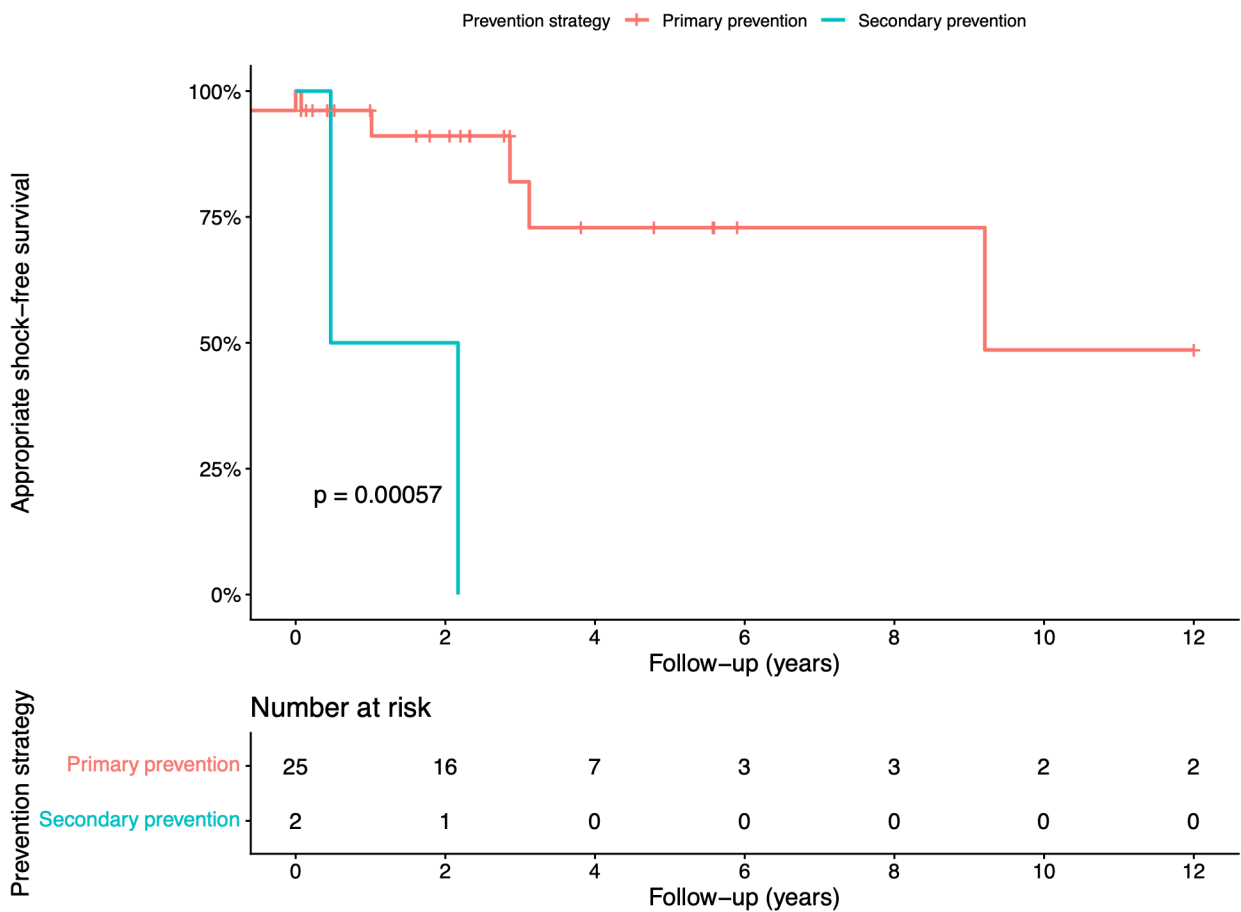
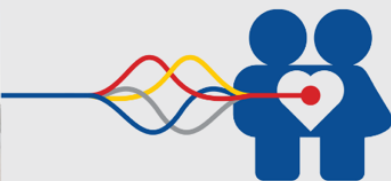


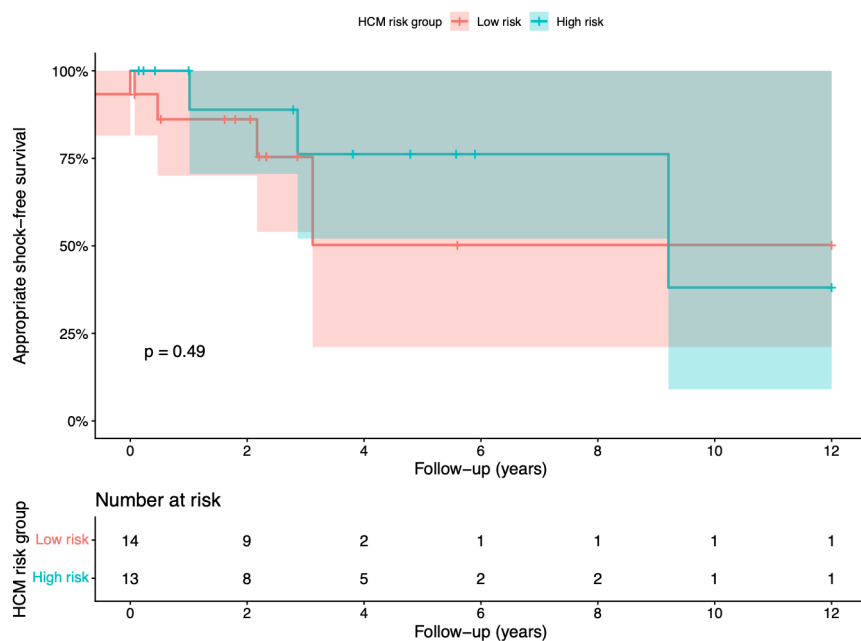
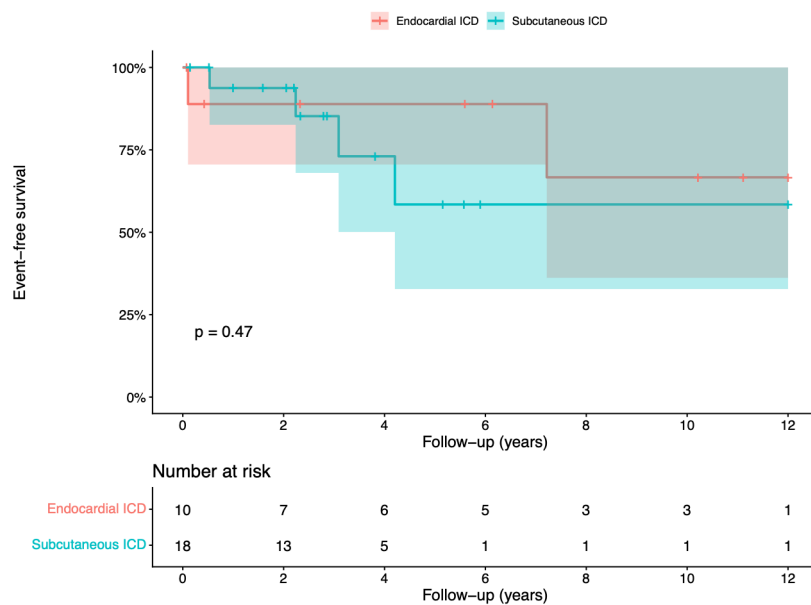
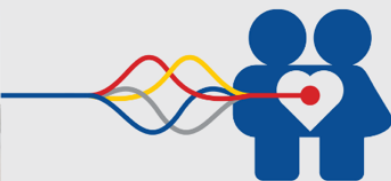
The HCM Risk-Kids score showed limited ability to identify patients experiencing appropriate therapies (AUC 0.50, 95% CI 0.22–0.78) and freedom from appropriate shocks did not differ between patients with HCM Risk-Kids $\geq 6\%$ and $< 6\%$ (log-rank $p=0.49$). Device-related complications occurred in six patients (21%), including five inappropriate shocks and one infection with erosion requiring reintervention. Complication-free survival was not significantly different between transvenous and subcutaneous ICD recipients (log-rank $p=0.47$). Progression of hypertrophy, assessed by changes in left ventricular wall thickness z-score over time, was not associated with device-related complications ($\beta = -0.04$, $p=0.54$)

Conclusion ICDs provided potentially life-saving therapies in 25% of our cohort. Guideline-recommended risk stratification tools seem to show limited ability to predict appropriate ICD interventions. The presence of ischemic ECG abnormalities and advanced diastolic dysfunction may provide incremental information beyond conventional risk models.

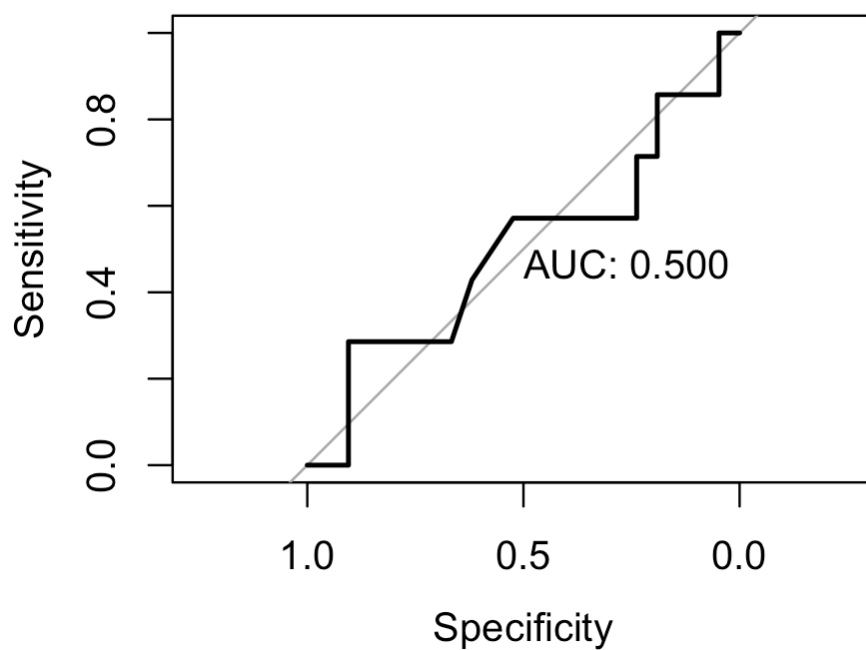


Variable	No appropriate shock (n=21)	Appropriate shock (n=7)	P value
Male sex, n (%)	15 (71.4)	3 (42.9)	0.362
Symptoms, n (%)			0.017
• None	20 (95.2)	4 (57.1)	
• Syncope	0 (0.0)	2 (14.3)	
• Neurally mediated syncope	1 (4.8)	0 (0.0)	
Height, cm	158.0 [150.0–166.0]	150.5 [146.8–155.0]	0.220
Weight, kg	52.0 [45.0–70.0]	41.0 [36.0–52.0]	0.089
Age at implant, years	15.0 [12.0–16.0]	11.0 [10.5–12.5]	0.042
Subcutaneous ICD, n (%)	15 (71.4)	3 (42.9)	0.362
Fluoroscopy time, min	1.05 [0.0–8.55]	9.0 [5.8–12.5]	0.198
Procedure time, min	91.5 [80.0–109.3]	105.0 [90.0–120.0]	0.684
ICD complication, n (%)	4 (19.0)	2 (28.6)	1.000
Pocket infection, n (%)	1 (4.8)	0 (0.0)	1.000
Lead erosion, n (%)	1 (4.8)	0 (0.0)	1.000
Inappropriate shock, n (%)	3 (14.3)	2 (28.6)	0.776
HCM Risk-Kids score at implant	8.7 [7.6–10.7]	9.0 [6.4–10.8]	1.000
HCM Risk-Kids score at follow-up	6.3 [4.6–10.2]	9.0 [7.7–9.9]	0.309
Maximal wall thickness at implant, mm	24.0 [21.0–27.0]	23.0 [16.5–27.0]	0.440
LV wall thickness z-score at implant (Boston)	11.0 [7.7–12.7]	15.0 [10.1–19.5]	0.301
Maximal wall thickness at follow-up, mm	27.0 [24.8–30.0]	26.5 [24.3–31.0]	0.927
LV wall thickness z-score at follow-up (Boston)	11.0 [9.7–13.5]	13.5 [7.9–18.2]	0.567
Left atrial diameter at implant, mm	32.0 [28.0–37.0]	29.0 [28.0–33.5]	0.506
Left atrial diameter z-score at implant (Detroit)	1.33 [0.95–2.21]	1.00 [0.88–2.15]	0.657
NSVT at implant, n (%)	6 (28.6)	3 (42.9)	0.815
Family history of SCD, n (%)	12 (60.0)	4 (57.1)	1.000
Syncope at implant, n (%)	1 (4.8)	1 (14.3)	1.000
LGE at implant, n (%)			0.662
• Absent	2 (10.0)	0 (0.0)	
• Present	14 (70.0)	2 (100.0)	
• Extensive	4 (20.0)	0 (0.0)	
End-stage disease at implant, n (%)	1 (4.8)	1 (14.3)	1.000
Apical aneurysm at follow-up, n (%)	0 (0.0)	1 (14.3)	0.151
High HCM risk score, n (%)	10 (47.6)	3 (42.9)	1.000
Massive HCM, n (%)	7 (33.3)	4 (57.1)	0.503





ROC curve of HCM Risk kids in predicting appropriate ICD therapies





PO.34 | HIGH-GRADE ATRIOVENTRICULAR BLOCK AS A MANIFESTATION OF CORONARY ISCHEMIA CAUSED BY AORTIC ROOT THROMBOSIS IN A CHILD WITH HYPOPLASTIC LEFT HEART SYNDROME

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Background

Atrioventricular (AV) block in children with congenital heart disease is most commonly associated with surgical injury, inflammation, or primary conduction system disease. Myocardial ischemia resulting in high-grade AV block is rare in the pediatric population. We report a case of transient high-grade AV block caused by coronary malperfusion secondary to thrombus formation in the native aorta of a child with hypoplastic left heart syndrome (HLHS).

Case Presentation

A 3-year-old boy with HLHS after staged Norwood-Sano and bidirectional Glenn palliation was transferred to our pediatric intensive care unit because of recurrent arrhythmias, elevated cardiac biomarkers, and angina-like symptoms. Prior to admission, he had experienced a transient loss of consciousness requiring bystander cardiopulmonary resuscitation.

During intensive monitoring, the patient developed intermittent high-grade AV block with pauses of up to six seconds, necessitating temporary pacing.

Echocardiography identified a thrombotic structure within the native aortic root near to the coronary artery origins while laboratory investigations revealed markedly elevated troponin levels.

Following multidisciplinary discussion, cardiac catheterization with local thrombolysis was performed. During the intervention, presumed embolization of thrombotic material into the right coronary artery resulted in acute hemodynamic collapse and cardiac arrest. After prolonged cardiopulmonary resuscitation, veno-arterial extracorporeal membrane oxygenation (VA-ECMO) was established. Coronary angiography confirmed right coronary artery obstruction, which was successfully treated with thrombectomy, local thrombolysis, and coronary stent implantation.



Results

Cardiac function gradually recovered, allowing successful ECMO explantation after four days. No further AV conduction abnormalities occurred during the subsequent hospital course. Serial electrocardiograms demonstrated restoration of stable sinus rhythm without recurrence of AV block. Cardiac biomarkers decreased progressively, and echocardiography showed improved ventricular performance. The patient was discharged home in stable clinical condition under dual antiplatelet and anticoagulation therapy.

Conclusion

This case highlights acute AV block as a rare presentation of myocardial ischemia in a child with HLHS. In patients with complex congenital heart disease, new-onset conduction abnormalities should raise suspicion of coronary compromise, particularly when accompanied by elevated cardiac biomarkers and ischemic symptoms. Prompt recognition and aggressive multidisciplinary management may be lifesaving and can result in complete recovery of AV conduction despite severe ischemic injury.



PO.35 | A RARE CASE OF PARA-HISIAN ATRIOVENTRICULAR RE-ENTRANT TACHYCARDIA PRESENTING AS PERMANENT JUNCTIONAL RECIPROCATING TACHYCARDIA

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Background and Aim: Classical para-hisian pathways usually present with short RP tachycardia. There are rare reported permanent junctional reciprocating tachycardia (PJRT) arising from decremental retrograde accessory pathways. We report an unusual case of manifest para-Hisian pathway presenting as PJRT. Correct diagnosis is important because ablation targets often lie in close proximity to the AV conduction system.

Methods: A 14-year-old girl presented with incessant narrow-complex tachycardia since infancy, resulting in tachycardia-induced ventricular dysfunction. She was initially diagnosed as PJRT and managed medically.

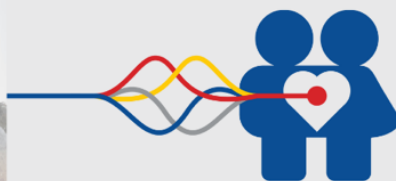
Prior electrophysiology study in demonstrated a septal re-entrant long-RP tachycardia. Mid-septal ablation terminated the tachycardia but resulted in first-degree AV block with early recurrence. A repeat study in 2017 with extensive cryomapping failed to perturb the arrhythmia and ablation was deferred.

She remained symptomatic and we repeated EP study. Baseline ECG demonstrated persistent pre-excitation with fused AV signals seen deep inside the coronary sinus. Multiple pacing maneuvers were analyzed.

Key findings were long VA interval during echo beats, earliest echo beat mapping to the non-coronary cusp region, intermittent loss and subsequent definitive elimination of antegrade pre-excitation and persistence of retrograde conduction despite loss of antegrade pathway conduction.

Ablation targeting the retrograde decremental pathway resulted in elimination of antegrade pre-excitation with no further recurrence during follow up.

Discussion and Conclusion: This case illustrates the diagnostic challenges posed by rare decremental retrograde pathways in children, made even more difficult by prior ablations. Careful analysis of pacing maneuvers, echo beats, pre-excitation behaviour, and bundle branch block responses was essential in establishing the diagnosis. A retrograde aortic approach was utilized to perform a safe ablation while minimizing the risk of AV nodal injury.

**PO.36 | DIASTOLIC FUNCTION IN PEDIATRIC LONG QT PATIENTS**

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Background: Channelopathies such as Long QT syndrome (LQTS) predispose to ventricular arrhythmias and sudden cardiac death. Although LQTS is traditionally classified as a purely electrical disorder, evidence from animal models and adult cohorts suggests subclinical mechanical dysfunction. Systematic data on pediatric populations remain absent. This study aims to evaluate mechanical myocardial alterations in pediatric LQTS patients.

Methods: We performed a retrospective analysis in a single tertiary referral center. Over an observational period from 2017 to 2025, standardized echocardiographic studies were performed in 86 pediatric LQTS patients (m=45, f=41) during routine medical evaluation. The cohort comprised LQTS1 (n=30), LQTS2 (n=12), LQTS3 (n=4), rare mutations (n=4), and clinical LQTS diagnoses (n=36). Patients with structural heart disease, additional mutations causing functional impairment, or repetitive arrhythmias were excluded. Echocardiograms assessed morphology, chamber size, and systolic function. Diastolic function was evaluated using E/A ratio, septal E/E', mitral annular E/E', and left atrial size. All values were reanalyzed by a single person.

Results: Diastolic dysfunction was documented in 33 of 86 patients (38%) at least once during the study. Genotype-specific prevalence of diastolic impairment was observed in 10 LQTS1 patients (33%), 5 LQTS2 patients (42%), 3 LQTS3 patients (75%), 1 LQTS4 patient, and 14 patients with a clinical Long QT diagnosis (39%). A severely prolonged QTcB interval (>500 ms) was present in 13 patients, 6 of whom (46%) demonstrated concomitant diastolic dysfunction.

Conclusion: Diastolic dysfunction is present in a substantial proportion of pediatric LQTS patients. This mechanical impairment does not solely depend on the degree of QTc prolongation. These findings support the hypothesis of an underlying electromechanical aspect in LQTS that manifests early in childhood.

**PO.37 | CARDIAC ARREST IN A PATHOLOGIC KCND3 VARIANT****Jason Cooper¹, Ann Goggins¹, Brendan Burke¹, Joel Temple¹**¹ Nemours Cardiac Center, Thomas Jefferson University, Philadelphia, USA

Clinical presentation: An 18-year-old male with a 916 G>A (G306S) heterozygous de novo gain of function variant in the KCND3 gene presented with a sudden cardiac arrest. The patient has known epilepsy, autism, and chronic difficult to control atrial fibrillation with rapid ventricular response. Baseline ECG was notable only for a diffuse early repolarization pattern (Figure 2). His atrial fibrillation was initially managed with atenolol and flecainide. Flecainide was discontinued given inadequate rhythm control. Two days after flecainide discontinuation the patient suffered an out-of-hospital sudden cardiac arrest. The patient's loop recorder captured polymorphic ventricular ectopy that degenerated into polymorphic ventricular tachycardia (PMVT) then ventricular fibrillation (VF) (Figure 1). He received appropriate resuscitation and was transferred to our hospital. He was cannulated onto ECMO, and his recovery was complicated by two similar events. Based on the limited literature available, quinidine gluconate was started with improvement of his early repolarization pattern and excellent rhythm control. The patient had cardiac and neurologic recovery to baseline. A transvenous dual-chamber ICD was placed for secondary prevention without complication.

Discussion: The KCND3 gene encodes the alpha subunit of the transient outward potassium current (I_{to}). Both gain of function and loss of function variants are associated with increased risk of sudden death. Gain of function variants are associated with Brugada syndrome, early repolarization syndrome, atrial fibrillation, developmental delay, and epilepsy. Flecainide is a weak inhibitor of the I_{to}. Given the close relationship between his arrest and flecainide cessation, it is possible that flecainide was suppressing ventricular arrhythmias. That said, coincidental disease progression cannot be ruled out.

Conclusion: To our knowledge, this is the first reported arrest with documented arrhythmia in a patient with a gain of function variant in the KCND3 gene. Apart from short runs of PMVT and ST changes prior to the event, there were no early warning signs of disease progression. After initiating quinidine, the patient demonstrated improvement of the early repolarization pattern (Figure 2). The role of flecainide in his event remains unclear, but the close relationship to his arrest suggests flecainide may provide some protection to this patient population.

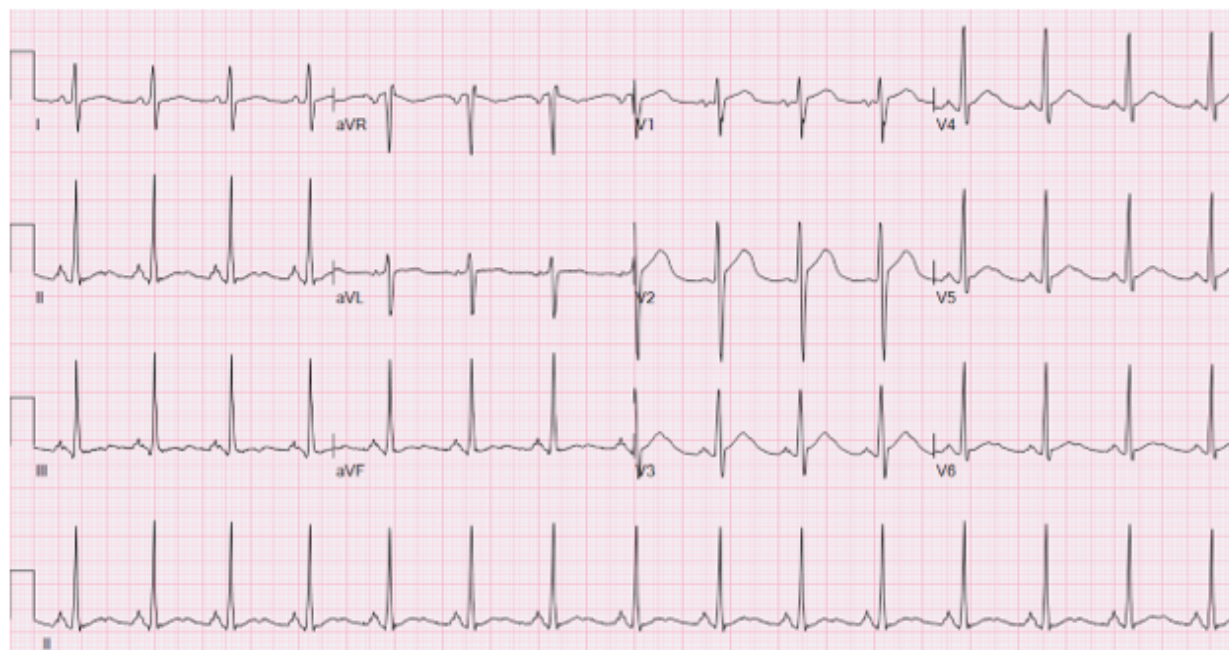


 **Episode Detail:** Tachy (ID# 6171)





Figure 2: Top image in blue is the patient's baseline ECG prior to quinidine initiation. Bottom image in red is the patient's ECG after quinidine initiation.



**PO.38 | SAFETY OF TRANSSEPTAL PUNCTURE IN LEFT-SIDED ABLATIONS IN CHILDREN:
A SINGLE-CENTER EXPERIENCE OF 301 CASES**

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Introduction: Transseptal puncture (TSP) is the standard approach for accessing left-sided arrhythmogenic substrates in pediatric patients. Children, however, are particularly vulnerable to the stochastic effects of ionizing radiation. It is therefore of paramount importance to adhere to the ALARA (As Low As Reasonably Achievable) principle, which aims to keep radiation exposure as low as possible without compromising procedural success. This study sought to evaluate the safety, efficacy, and procedural characteristics of TSP in a large pediatric cohort, with a particular focus on an ALARA-compliant workflow.

Methods: A total of 301 pediatric patients who underwent electrophysiology study and radiofrequency (RF) catheter ablation requiring TSP at our center were retrospectively reviewed. All procedures were performed under the guidance of three-dimensional (3D) electroanatomic mapping to minimize overall radiation exposure; however, the transseptal puncture itself was carried out using conventional techniques under fluoroscopy guidance alone.

Results: The mean age of the study population was 12.15±4.0 years, with a mean body weight of 48.22±19.47 kg. Structurally normal hearts were present in 99% of cases. The primary indications were manifest accessory pathways (55.5%) and concealed accessory pathways (36.2%). TSP was successfully performed in 99.3% of patients (299/301). In the two cases where TSP was unsuccessful, a transaortic approach was employed and ablation was completed successfully. The mean total ablation procedure time was 92.69±40.03 minutes. Notably, the mean fluoroscopy time was recorded at 52.74±81.14 seconds, reflecting the ultra-low radiation workflow. The overall acute ablation success rate was 96.7%. No major or minor complications, including pericardial effusion or tamponade, were observed across the entire cohort.



Conclusion: Our findings demonstrate that transseptal puncture in the pediatric population is an exceptionally safe and effective procedure, achieving a success rate exceeding 99% with zero complications. Furthermore, this study provides evidence that TSP can routinely be performed with ultra-low fluoroscopy times (mean <1 minute), even when conducted under fluoroscopy guidance alone, when supported by 3D electroanatomic mapping. These results represent a successful real-world application of the ALARA principle in pediatric electrophysiology.

Keywords: Pediatric electrophysiology; Transseptal puncture; Catheter ablation; ALARA principle; Fluoroscopy reduction; 3D electroanatomic mapping.

**PO.39 | NON-INVASIVE ELECTROCARDIOGRAPHIC IMAGING TO ASSESS VENTRICULAR ACTIVATION DURING LEFT BUNDLE BRANCH AREA PACING IN PEDIATRIC PATIENTS**

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Background and Aim

Conduction system pacing (CSP), particularly Left Bundle Branch Area Pacing (LBBAP), has emerged as an alternative to cardiac pacing in children. However, the lack of validated ECG criteria for LBB capture in children remains a challenge, thus the need for using novel non-invasive tools. Electrocardiographic Imaging (ECGI) is a non-invasive cardiac mapping tool that reconstructs biventricular activation from a single heartbeat using high-density body surface potentials. This study aims to evaluate ECGI-derived metrics as an intraoperative assessment tool during LBBAP implantation in pediatric patients.

Methods

Thirteen pediatric patients (58.3% male, 8.9±4.9 years, 35.5±20.2 Kg) undergoing LBBAP implantation were prospectively included. Cardiac diagnosis included: 4 patients with structurally normal hearts and CHB and 9 with CHB and structural cardiac abnormalities; 12 with CHD and 1 with hypertrophic cardiomyopathy. 5 patients had previous epicardial cardiac devices; 2 RV, 2 LV, 1 epicardial CRT. Simultaneous ECG recordings and ECGI maps were acquired during the LBBAP procedure. Patients were divided in 2 groups based on baseline QRS: group 1 included patients with a normal QRS duration (n=4); group 2 included those having intrinsic bundle branch block or were paced (n=9). Results were compared between groups using non-parametric related-samples Wilcoxon Signed Rank Test.

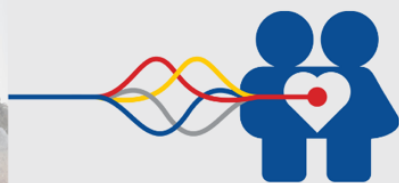


Results

There was a non-statistically significant reduction in QRS duration (134.1 ± 33.2 vs 117.5 ± 17.3 ; $p=0.09$), ECGI duration (122.3 ± 25.0 vs 118.3 ± 19.4 ; $p=0.42$) and TAT (77.8 ± 32.2 vs 69.0 ± 13.6 ; $p=0.34$) across all patients when comparing baseline and final position. When comparing between groups, QRS duration (89.3 ± 6.3 vs 109.3 ± 22.7 ; $p=0.1$), ECGI duration (97.2 ± 17.3 vs 106.7 ± 26.3 ; $p=0.45$) and TAT (35.0 ± 4.1 vs 58.7 ± 5.9 ; $p=0.06$) were non-statistically significantly prolonged for patients in Group I, whereas all parameters were significantly shortened in Group II: QRS duration (151.0 ± 19.6 vs 120.6 ± 15.6 ; $p=0.01$), ECGI duration (133.4 ± 19.4 vs 123.4 ± 14.4 ; $p=0.1$) and TAT (96.9 ± 15.2 vs 73.6 ± 13.8 ; $p=0.02$).

Conclusions

ECGI metrics indicate that LBBP improves significantly all ventricular activation times measured using QRS duration, ECGI duration and TAT when performed in patients with previous pacing or intrinsic bundle branch block. Despite the limited sample size, these findings support the use of ECGI as a real-time assessment tool during LBBP implantation in pediatric patients.

**PO.40 | TRANSIENT AV BLOCK POST TRANSCATHETER ASD DEVICE CLOSURE****Natasha Donald¹, Leonie Wong², Carles Bautista²**¹ Paediatric Cardiology, Royal Brompton Hospital² Paediatric Cardiology, Royal Brompton Hospital, Guy's and St Thomas' NHS Foundation Trust, London**Background and aim:**

Atrioventricular block (AVB) is an uncommon but recognised complication of transcatheter ASD closure. Whilst most cases are transient, persistent conduction disease and late progression to complete heart block have been reported. We describe a case of a 7 year old boy who developed early complete AV block recovering at day 2 post ASD device closure.

Methods:

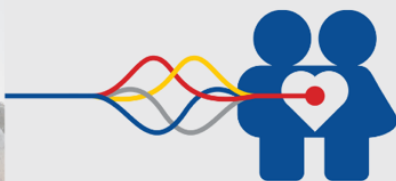
Data was retrospectively collected from an electronic patient record including demographics, imaging findings, procedural details, serial ECG, echocardiography and follow up ambulatory monitoring.

Results:

A 7 year old boy (26kg, 134.5cm) underwent transcatheter device closure of a large secundum ASD. ASD measured up to 28mm with a deficient posterior rim. A 24 mm Figulla Flex II occluder was initially deployed but failed to completely occlude the defect and was replaced with a 27mm device. Wenckebach conduction was observed intraoperatively. Indexed device size for height was 0.2mm/cm. Aspirin was commenced post procedure.

Complete AV block developed following device implantation with a haemodynamically stable junctional escape rhythm at 80-90 bpm. Prednisolone (2 mg/kg/day) was commenced and device removal was considered. However, gradual recovery of conduction occurred over the subsequent 2 days progressing through marked first-degree AV block (maximum PR interval 440ms) before ultimate recovery of normal 1:1 AV conduction at 42 hours. Follow-up echocardiography demonstrated stable device position and preserved biventricular function. Follow up 72hr Holter ECG showed sinus rhythm throughout with no recurrent AV block.

From the existing literature, high risk factors in this case included large device size, deficient posterior rim and intraprocedural conduction disturbance. Protective factors included a device/weight ratio <1.5mm/kg, older age and absence of pre-existing conduction disease.



Conclusion:

This case demonstrates transient complete AV block following ASD device closure in a child with multiple recognised risk factors. Intraprocedural Wenckebach conduction may represent an early marker of AV node injury. Despite progression to complete AV block, conduction recovered fully with conservative management and corticosteroid therapy avoiding device explant. Predictors of early recovery and the role of corticosteroid therapy in avoiding device explant remain poorly defined. Further cohort studies are needed to develop an evidence-based approach to these cases.



PO.41 | PULSED FIELD ABLATION FOR ATRIAL FIBRILLATION IN ADULT CONGENITAL HEART DISEASE: LESSONS FROM SEVEN CASES

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¹ Department of Pediatric Cardiology, Sakakibara Heart Institute

Background and Aim:

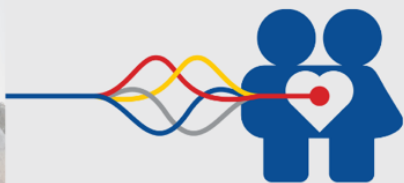
Pulsed field ablation (PFA) has emerged as a nonthermal ablation modality with potential safety advantages for atrial fibrillation (AF). However, experience in adult congenital heart disease (ACHD) remains limited, particularly in complex or cyanotic patients. We reviewed our early experience with PFA in ACHD patients to evaluate procedural feasibility, recurrence patterns, and safety signals relevant to patient selection.

Methods:

We retrospectively analyzed seven ACHD patients who underwent PFA for AF or atrial tachyarrhythmia at our institution. Clinical background, prior ablation history, ablation strategy, acute procedural success, arrhythmia recurrence, repeat procedures, and periprocedural complications were reviewed. Particular attention was paid to two patients undergoing PFA after prior cryoballoon ablation.

Results:

The median age was 53 years, and five patients were female. Underlying diagnoses included repaired atrial septal defect, congenital aortic stenosis after Ross-Konno operation, congenital atrioventricular block with pacemaker implantation, congenital mitral regurgitation, transposition of the great arteries after Rastelli repair, unrepaired tricuspid atresia with cyanosis, and repaired ventricular/atrial septal defects. PFA strategies included pulmonary vein isolation, posterior wall isolation, and additional substrate modification according to arrhythmia mechanism. Acute target lesion creation was achieved in all evaluable cases. During a median follow-up of 9 months, late recurrence occurred in one patient, while early recurrences were mainly atrial tachycardia. In one redo case after cryoballoon ablation, reconnection of the left pulmonary veins and residual anterior/carina potentials were identified, and PFA with posterior wall isolation resulted in freedom from recurrence. In contrast, a cyanotic unrepaired tricuspid atresia patient, despite preserved pulmonary vein and posterior wall isolation, developed marked hemolysis followed by acute kidney and liver injury requiring intensive care and haptoglobin administration.



Conclusions:

PFA was feasible in ACHD patients and may be particularly useful in post-cryo redo procedures by allowing targeted lesion optimization. However, recurrence in ACHD may reflect atrial substrate beyond pulmonary vein reconnection. Cyanotic ACHD patients may be vulnerable to clinically significant hemolysis-related organ injury, highlighting the need for careful patient selection and strict postprocedural monitoring.



PO.42 | ELECTRICAL CAPTURE, MECHANICAL RESPONSE AFTER UPGRADE FROM DUAL-CHAMBER PACING TO LEFT BUNDLE BRANCH PACING: A PROSPECTIVE ECHOCARDIOGRAPHIC AND ELECTRICAL ASSESSMENT IN CORRECTED CONGENITAL HEART DISEASE

Yazmin Binti Yusoff Azudin¹, Dr. Hasri Samion², Esther Mathias Ajik²

¹

² Paediatric and Congenital Heart Centre,

Introduction: Chronic right ventricular pacing may lead to electrical and mechanical dyssynchrony, contributing to ventricular dysfunction and heart failure progression. Left bundle branch area pacing (LBBAP) has emerged as a physiological pacing strategy that restores ventricular activation through the native conduction system. We report the initial experience at Institute Jantung Negara (IJN) evaluating electrical, echocardiographic, and biomarker changes following upgrade from dual-chamber pacing to LBBAP.

Methods: Three patients with chronic dual-chamber pacemakers underwent successful upgrade to LBBAP and were prospectively evaluated. Electrical assessment included QRS duration and left ventricular activation time (LVAT). Echocardiographic parameters assessed before and after implantation included aortic-pulmonary pre-ejection delay, left ventricular posterior wall-mitral inflow delay, electromechanical delay (EMD), electrosystolic delay (ESD) and left ventricular ejection fraction (LVEF). NT-proBNP levels were measured as a marker of neurohormonal response.

Results: LBBAP implantation was successful in all patients without procedural complications. Mean QRS duration decreased from 170.3ms to 144.0ms, indicating improved electrical synchrony. Aortic-pulmonary pre-ejection delay improved in two patients (61ms to 43ms and 72ms to 43ms), while left ventricular posterior wall-mitral inflow delay improved in two patients (74ms to 40ms and 69ms to 0ms). NT-proBNP levels decreased in all patients, from 73 to 30 pg/ml, 848 to 377 pg/ml, and 269 to 202 pg/ml, respectively. LVEF improved from 44% to 56% in one patient, while modest reductions were observed in the remaining two patients. EMD and ESD demonstrated variable responses following upgrade.

Conclusion: In this initial IJN experience, upgrading from dual-chamber pacing to LBBAP was feasible and associated with improved electrical synchrony and favorable reductions in NT-proBNP. Improvements in selected echocardiographic measures of mechanical synchrony suggest a potential physiological benefit of LBBAP. Larger studies with longer follow-up are warranted to further define the impact of LBBAP on ventricular mechanics and cardiac function.

**PO.43 | A UNIQUE AND RARE CLINICAL PRESENTATION OF SEVERE EBSTEIN'S ANOMALY ASSOCIATED WITH A LEFT-SIDED ACCESSORY PATHWAY**

Khalid Dagreri¹, Rayan Halabi¹, Saleh Al Ghamdi¹, Sara Shehab¹, Lamia Al Shengeiti², Ahmed Al Fagih

¹ Pediatric Cardiac Electrophysiology,

² Adult cardiac electrophysiology,

Objective:

This report highlights a rare clinical association between Ebstein's anomaly and a left-sided concealed accessory pathway (AP), with a focus on its clinical and electrophysiological characteristics.

Methods:

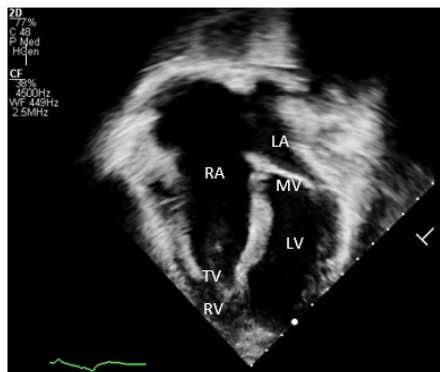
A 12-year-old girl with a diagnosis of Ebstein's anomaly was planned for surgical intervention. Despite experiencing frequent palpitations, her 12-lead ECG showed sinus rhythm. An electrophysiological study confirmed the presence of a left-sided concealed AP. Radiofrequency ablation was performed using an irrigated-tip catheter via trans-septal access.

Results:

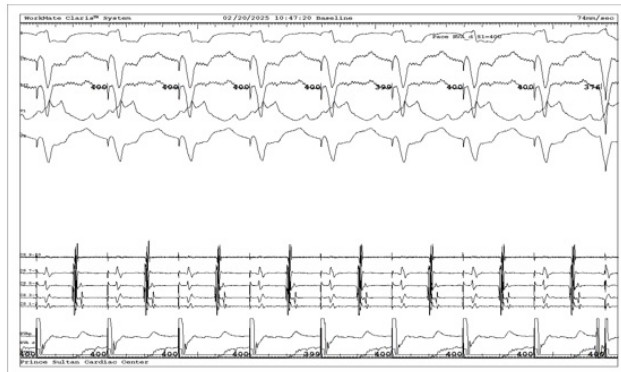
The electrophysiological study identified the earliest activation at the left posterior-lateral mitral annulus, confirming the AP location. Successful radiofrequency ablation resulted in complete elimination of the retrograde conduction through the pathway.

Conclusion:

This case underscores the rarity of a left-sided concealed AP in the context of Ebstein's anomaly. It emphasizes the importance of a comprehensive electrophysiological evaluation in complex cases and demonstrates the effectiveness of radiofrequency ablation in managing arrhythmias associated with congenital heart defects.



A) Trans-Thoracic echo (4-Chamber view) labelled



B) Intra-cardiac tracing during ventricular pacing showing eccentric retrograde activation



C) Intra-cardiac tracing during tachycardia showing eccentric retrograde activation



D) Post ablation intra-cardiac tracing during ventricular pacing showing VA dissociation



PO.44 | FIRST REPORTED CASE OF PULSED FIELD ABLATION FOR FOCAL ATRIAL TACHYCARDIA IN A POST FONTAN PALLIATION

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² pediatric cardiology,

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Introduction

Atrial tachyarrhythmias are a common and challenging late complication in patients with Fontan circulation. Extensive atrial scarring, prior surgical incisions, and altered hemodynamics create a complex arrhythmogenic substrate, making catheter ablation technically demanding and potentially risky. Pulsed field ablation (PFA) has emerged as a non-thermal, myocardial-selective ablation technology with an excellent safety profile in large atrial fibrillation cohorts, including the MANIFEST-PF study of more than 40,000 patients. However, its use in Fontan patients remains extremely limited, and reported experience has been confined to atrial fibrillation rather than atrial tachycardia.

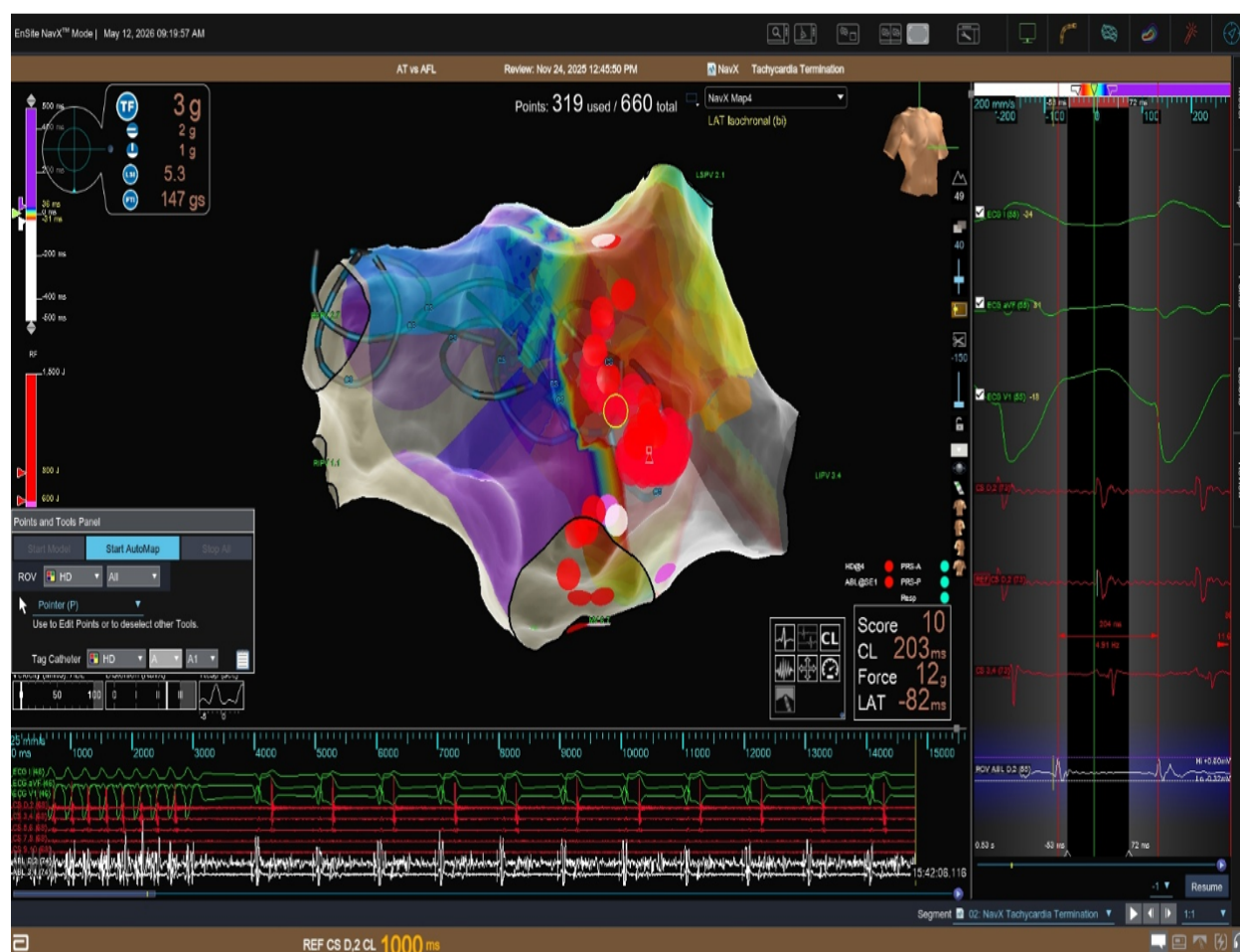
Case Presentation

We describe a 42-year-old patient with extracardiac Fontan circulation who presented with recurrent, drug-refractory atrial tachycardia. The procedure was performed under general anesthesia using a carefully planned multimodality approach tailored to Fontan anatomy. High-density electro-anatomical mapping revealed multiple atrial tachycardia mechanisms. Two arrhythmias were successfully treated using conventional radiofrequency ablation with confirmed conduction block. A third tachycardia, involving a slow-conduction channel near the left upper pulmonary vein, was treated using pulsed field ablation. PFA resulted in immediate arrhythmia termination without hemodynamic instability, collateral injury, or procedural complications. Post-ablation testing demonstrated complete non-inducibility, with preserved Fontan pathway integrity on intracardiac echocardiography.



Conclusion

To our knowledge, this is the **first reported case of pulsed field ablation for atrial tachycardia in a patient with Fontan circulation**, and the first reported outside North America. This case highlights the feasibility and potential safety advantages of PFA in complex congenital heart disease and supports its consideration as an adjunctive tool for atrial tachyarrhythmia ablation in high-risk Fontan patients.



Three-dimensional voltage map of the left atrium in anterior view demonstrating the ablation lesions created using pulsed field ablation (PFA). A linear lesion is seen extending from the right superior pulmonary vein toward the anterior left atrial wall (baffle region). Intracardiac electrograms illustrate the ongoing tachycardia, followed by its termination to a ventricular paced rhythm after PFA applications.



**PO.45 | TRANSHEPATIC VENOUS ACCESS FOR ABLATION OF A LEFT-SIDED
ACCESSORY PATHWAY IN A CHILD WITH SINGLE-VENTRICLE PHYSIOLOGY AND
BILATERAL FEMORAL VEIN OCCLUSION**

**Camille Marie Go¹, Vinay Bhole, Charalampos Kotidis, Krishnakumar Pillai, Neeraj
Nirmal**

¹ Cardiology, Birmingham Women and Children's NHS foundation Trust

Background and Aim:

Catheter ablation in children with complex congenital heart disease may be challenging when conventional venous access routes are unavailable. Transhepatic venous access is a potential alternative but remains infrequently reported in pediatric electrophysiology. We describe successful radiofrequency ablation of a left-sided accessory pathway using exclusive transhepatic venous access in a young child with single-ventricle physiology and bilateral femoral vein occlusion.

Methods:

A 3-year-old girl with unbalanced atrioventricular septal defect, hypoplastic left ventricle, double-outlet right ventricle, malposed great arteries, and previous staged single-ventricle palliation presented with recurrent supraventricular tachycardia despite antiarrhythmic therapy. Escalation of antiarrhythmic treatment resulted in symptomatic bradycardia requiring implantation of an epicardial pacemaker before bidirectional cavopulmonary shunt surgery. During planned pre-Fontan cardiac catheterization and electrophysiology study, complete bilateral femoral venous occlusion was identified. Following multidisciplinary review and ultrasound confirmation of suitable hepatic venous anatomy, percutaneous transhepatic venous access was obtained. A three-dimensional electroanatomical mapping system was used to identify the arrhythmia substrate and guide radiofrequency ablation. Epicardial pacing leads were present at the time of the procedure.

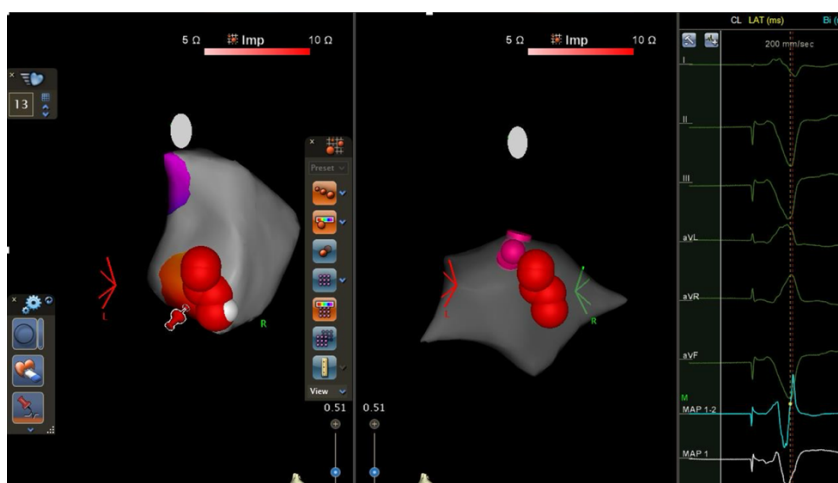
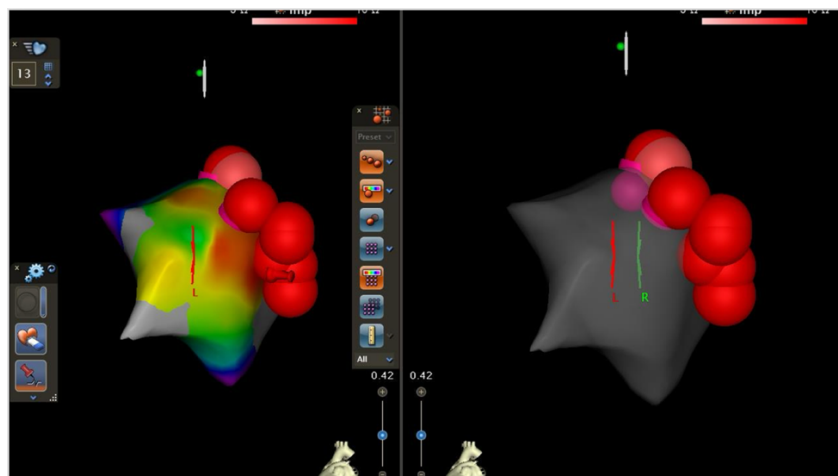
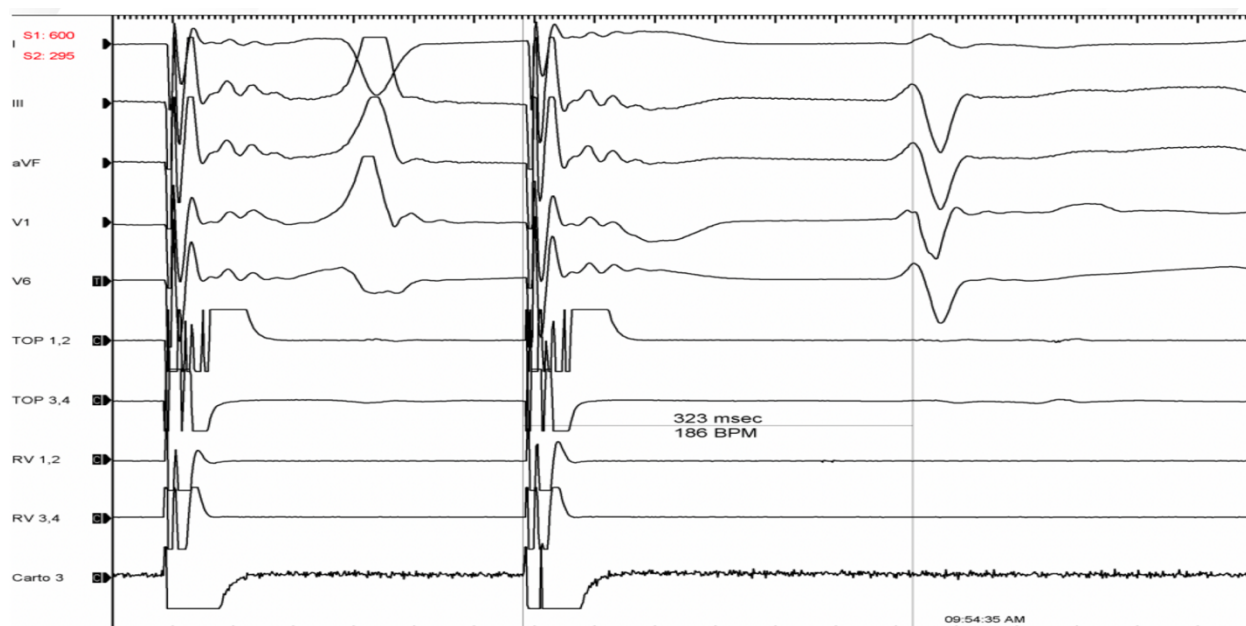
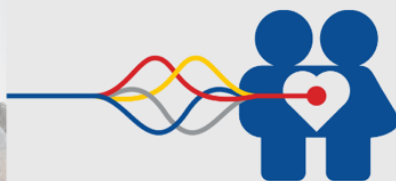
**Results:**

Baseline electrocardiography demonstrated subtle ventricular pre-excitation suggestive of a left-sided accessory pathway. Transhepatic venous access was successfully established without complication. Mapping identified the earliest atrial activation along the left atrioventricular valve annulus, consistent with a left-sided accessory pathway. Radiofrequency energy application resulted in immediate loss of pre-excitation and elimination of accessory pathway conduction. No arrhythmia was inducible following ablation. The hepatic access tract was closed percutaneously without complication. There were no procedural adverse events, including bleeding, hepatic injury, or haemodynamic instability. At 12-month follow-up, there was no recurrence of supraventricular tachycardia or ventricular pre-excitation, antiarrhythmic therapy had been successfully discontinued, and the patient subsequently underwent Fontan completion without arrhythmic complications.

Conclusions:

Transhepatic venous access provides a safe and effective alternative route for catheter ablation in children with complex congenital heart disease and obstructed conventional venous access. Three-dimensional electroanatomical mapping facilitates successful ablation in complex cardiac anatomy. To our knowledge, this represents one of the youngest reported patients to undergo successful accessory pathway ablation using exclusive transhepatic venous access. Early definitive arrhythmia management before Fontan completion may improve procedural feasibility and reduce future arrhythmia burden.





**PO.46 | SERIAL ELECTROCARDIOGRAPHIC REAPPRAISAL AND GENETIC TESTING SUGGEST A CARDIAC-PREDOMINANT ANDERSEN-TAWIL PHENOTYPE IN AN ADOLESCENT ATHLETE**

Daniele Garozzo¹, Cristina Raimondo¹, Marco Perrone², Francesco Flore¹, Daniela Righi¹, Corrado Di Mambro¹, Massimo Stefano Silvetti¹, Fabrizio Drago¹

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² Division of Cardiology and CardioLab, Department of Clinical Sciences and Translational Medicine University of Rome Tor Vergata e Clinical Pathways and Epidemiology Unit, Bambino Gesù Children's Hospital IRCCS., Division of Cardiology and CardioLab, Department of Clinical Sciences and Translational Medicine University of Rome Tor Vergata e Clinical Pathways and Epidemiology Unit, Bambino Gesù Children's Hospital IRCCS.

Background and Aim: Complex ventricular ectopy in adolescents with a structurally normal heart may conceal an inherited arrhythmia syndrome. We report an asymptomatic competitive athlete in whom serial electrocardiographic reinterpretation and genetic testing shifted the diagnostic framework from apparently idiopathic ventricular ectopy to a cardiac-predominant Andersen-Tawil phenotype.

Methods: We retrospectively reviewed serial electrocardiograms, exercise tests, 24-hour Holter recordings, echocardiography, cardiac magnetic resonance findings, genetic data, and response to antiarrhythmic therapy.

Results: Sports screening revealed baseline ventricular bigeminy. Initial Holter monitoring documented about 6% premature ventricular complexes (PVCs), mainly monomorphic, with rare dimorphic couplets and one 7-beat episode of nonsustained ventricular tachycardia. Exercise testing showed isolated PVCs with right bundle branch block morphology and superior axis, consistent with fascicular origin, organizing into bigeminy at low workloads, suppressing at peak exercise, and reappearing during recovery. Cardiac magnetic resonance showed a structurally normal heart without myocardial scar. After nadolol, PVC burden decreased to 4%, but the phenotype remained complex, with persistence of the fascicular morphology and emergence of a second outflow-tract morphology with left bundle branch block pattern, together with frequent couplets, triplets, and a short accelerated idioventricular rhythm during recovery. Serial electrocardiograms later disclosed prominent U waves and dynamic terminal repolarization abnormalities.



Interpretation of the corrected QT interval was limited by fusion of T and U waves, so the abnormality was considered mainly at the level of the terminal T-U complex. Genetic testing identified a maternally inherited heterozygous KCNJ2 c.443G>C p.Arg148Thr missense variant, absent from population databases and classified as likely pathogenic. Combined flecainide and nadolol markedly simplified the phenotype, with no inducible arrhythmias on repeat exercise testing and only four isolated PVCs on follow-up Holter monitoring.

Conclusions: In an adolescent athlete with a structurally normal heart, complex multiform ventricular ectopy should prompt reassessment for an inherited arrhythmia syndrome when accompanied by prominent U waves and terminal T-U abnormalities. In the present case, the combined genetic, electrocardiographic, and arrhythmic profile raised suspicion of a KCNJ2-related Andersen-Tawil phenotype despite the absence of overt extracardiac manifestations. This diagnostic reappraisal supported family evaluation and a more targeted therapeutic strategy.

**PO.47 | ANDERSEN-TAWIL SYNDROME IN A MOTHER-SON PAIR: DIAGNOSTIC DELAY, CASCADE SCREENING, AND POSSIBLE PHENOTYPIC MODIFICATION BY DUAL CHANNELOPATHY****Bilgehan Betül Biçer¹, Tevfik Karagöz¹, İlker Ertuğrul¹**¹ Pediatric Cardiology, Hacettepe University Faculty of Medicine**Background and Aim**

Andersen-Tawil syndrome (ATS) is a rare autosomal dominant channelopathy characterised by periodic paralysis, ventricular arrhythmias, and craniofacial/skeletal dysmorphism. Genetically confirmed cases remain limited in the literature, and diagnosis is frequently delayed because of variable expression and incomplete presentation of the classic triad. We aimed to describe the clinical, electrophysiological, and genetic characteristics of a mother-son pair with ATS and to highlight diagnostic challenges, variable clinical expression, and the potential implications of dual channelopathy.

Methods

Clinical assessment, electrocardiography, Holter monitoring, exercise testing, dysmorphology evaluation, and next-generation sequencing were performed in both individuals.

Results

The proband, a 15-year-old male, presented with recurrent hypokalemic paralysis over several years but received a definitive diagnosis only after approximately six years, during which multiple emergency admissions and repeated neuroimaging investigations were performed without diagnostic clarification. He demonstrated frequent premature ventricular contractions, nonsustained ventricular tachycardia, and classic ATS electrocardiographic findings including prominent U waves. Exercise testing did not induce ventricular tachycardia, and premature ventricular contractions were not suppressed during exercise. Beta-blocker therapy resulted in satisfactory control of ventricular arrhythmias and clinical improvement. His mother reported similar episodes of paralysis during childhood and adolescence that resolved in adulthood and displayed comparable craniofacial and skeletal features consistent with ATS. Although asymptomatic from a cardiac perspective, electrocardiographic and Holter evaluation demonstrated prominent U waves and rare polymorphic premature ventricular contractions.



She had never undergone cardiological or genetic assessment, and cascade screening following the proband's diagnosis identified the same pathogenic KCNJ2 variant (c.199C>T; p.Arg67Trp), confirming her diagnosis. The proband also carried a likely pathogenic SCN4A variant associated with hypokalemic paralysis.

Conclusions

This family highlights the diagnostic delay frequently encountered in ATS and the importance of recognising subtle clinical features. Identification of the mother's diagnosis following evaluation of her son underscores the value of cascade screening. The mother and son demonstrated variable clinical expression despite sharing the same pathogenic KCNJ2 variant, illustrating the heterogeneous presentation of ATS. The additional SCN4A variant may have contributed to phenotypic variability. Early recognition, genetic confirmation, and tailored management are essential for affected patients and their families.

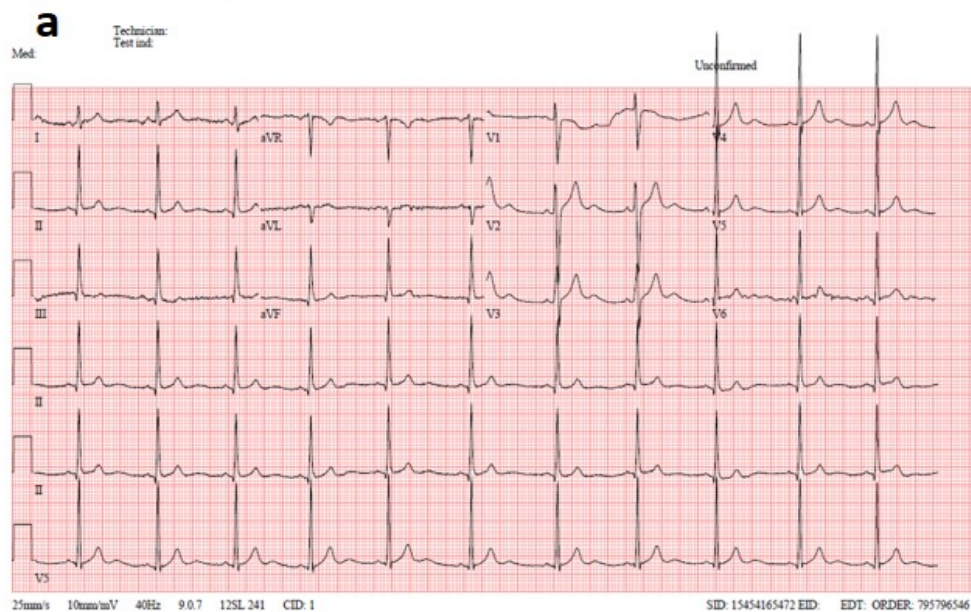




24-JUN-2009 (15 yr)
Male
Room:
Loc: 19

Vent. rate 68 BPM
PR interval 110 ms
QRS duration 82 ms
QT/QTc 354/378 ms
P-R-T axes 21 78 33

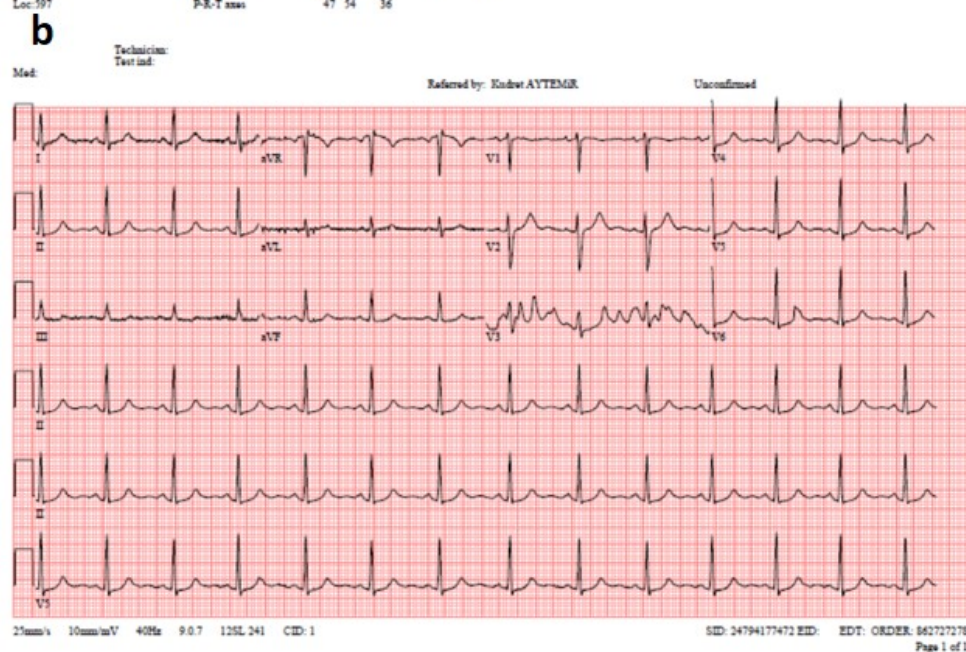
*** Pediatric ECG analysis ***
Normal sinus rhythm
Normal ECG



27-SEP-1990 (33 yr)
Female
Room:
Loc: 597

Vent. rate 81 BPM
PR interval 140 ms
QRS duration 82 ms
QT/QTc 402/467 ms
P-R-T axes 47 54 36

Normal sinus rhythm
Nonspecific ST abnormality
Abnormal ECG





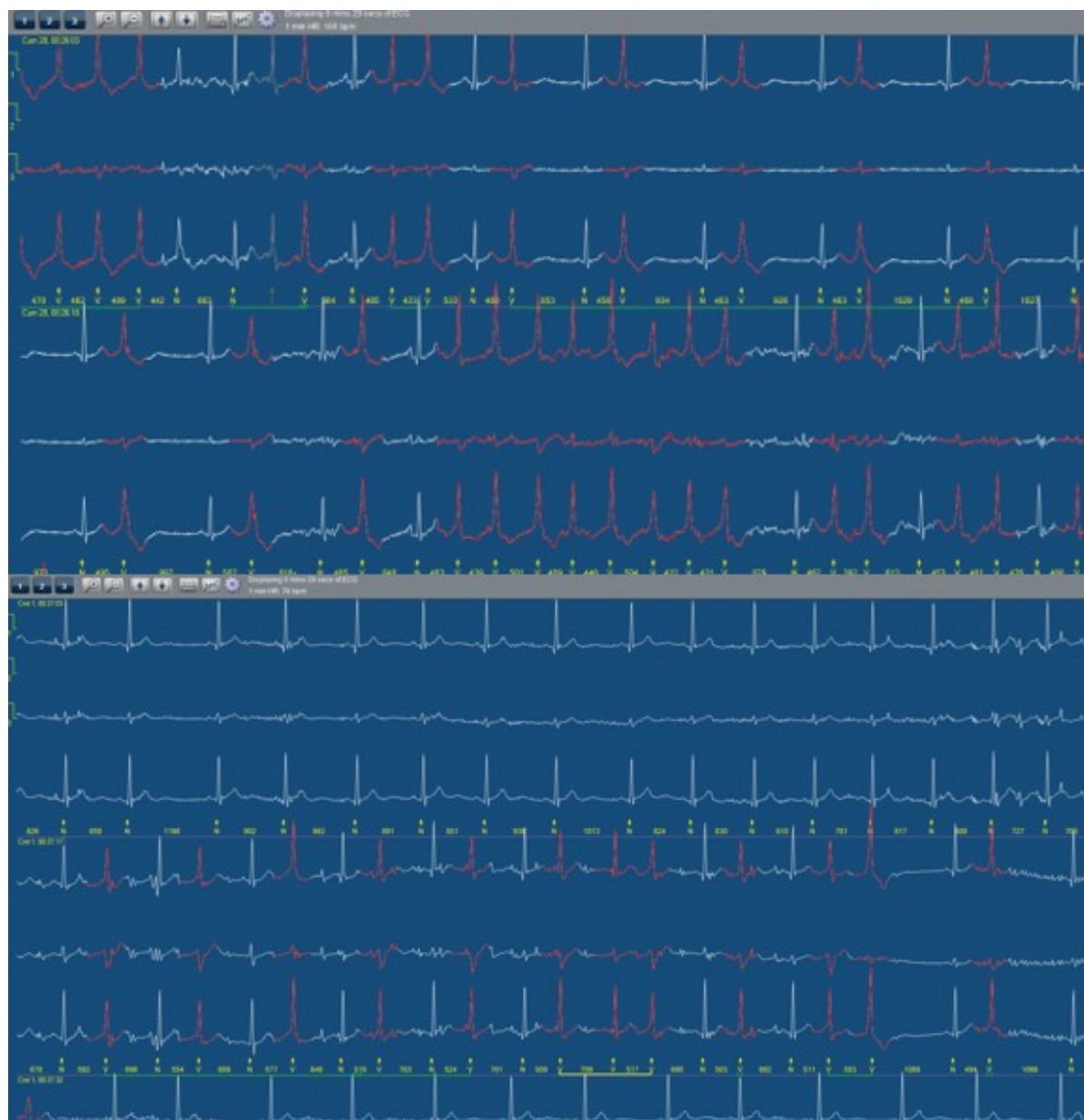
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PO.48 | COMPARISON OF NOVEL MAPPING TECHNIQUES FOR RF ABLATION OF THE SLOW PATHWAY IN THE TREATMENT OF AVNRT IN CHILDREN: RESULTS FROM AN INTERNATIONAL MULTICENTRE STUDY

Tomasz Książczyk¹, Radosław Pietrzak¹, Leonie Wong², Will Regan³, Uładzislau Shyshko⁴, Jan Till², Eric Rosenthal³, Maria Prieto⁵, Eva Sellas Lleo⁶, Klaudia Obsznajczyk-Kowalewska¹, Ferran Roses-Noguer⁷

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⁵ Abbott Medical, Abbott Medical

⁶ Electrophysiology, J&J MedTech

⁷ Department of Paediatric Cardiology, Vall d'Hebron University Campus, Barcelona

Background:

In recent years, new mapping techniques, using concepts such as, low-voltage bridge (LVB), collision point, and pivot point, have been developed to facilitate slow-pathway ablation for the treatment of AVNRT. The aim of this study was to compare the performance of these mapping methods in predicting the target for radio-frequency ablation of the slow pathway in children.

Methods:

This is an international, multicentre, retrospective study analysing paediatric typical slow-fast AVNRT ablations from January 2020 to December 2025. 3D maps of the triangle of Koch were reanalysed to assess LVB, collision, and pivot points relative to successful RF sites. Primary analyses used a frequentist GLMM (binomial, logit) with Laplace approximation. Final inference employed Bayesian logistic GLMMs with weakly regularizing Student-t priors.



Results:

Four European centres submitted 138 paediatric patients-studied with either a Carto (Biosense Webster, J&J), or Abbot mapping system (either Precision or EnSite X). In the Bayesian GLMM, the posterior mean success probabilities were: LVB 0.69 (95% CrI 0.61-0.78), collision 0.17 (0.10-0.24), and pivot 0.25 (0.16-0.35). Direct (pairwise) comparisons showed that LVB performed much better than two other methods: LVB versus collision OR 11.40 (95% CrI 5.27-21.09) and LVB versus pivot OR 6.71 (3.25-12.15). Within-method comparisons indicated little or no difference between the 3D mapping systems.

Conclusions:

Our study identifies the LVB as the strategy with the highest probability of overlap with the successful spot for the slow pathway modification in children with AVNRT. The interaction between the mapping systems and mapping strategy showed no major difference

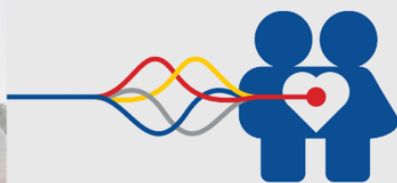
**PO.49 | PULSED-FIELD-ABLATION OF A VENTRICULAR TACHYCARDIA IN A 8 YEAR OLD PATIENT AFTER REPAIR OF FALLOT-TETRALOGY****Sophia Klehs¹, Christian Paech¹, Ingo Daehnert¹, Roman Gebauer¹**¹ Department for Pediatric Cardiology, University of Leipzig - Heart Center

Background: Ventricular Tachycardia (VT) in adult patients after repair of Fallot-Tetralogy (rTOF) are common. Here we present a case of a 8 year old patient (25 kg) after rTOF presenting with VT and treatment with pulsed-field-ablation (PFA).

Case report: Initially the patient with TOF and single coronary ostium (right coronary artery arising from the left sinus) was corrected with RVOT-patch with 7 months. One year later the patient required re-operation for subaortal obstruction, including implantation of a 12-mm Contegra® conduit, resulting in two outflow tracts of the right ventricle.

The first sustained VT occurred at the age of 8 years (monomorph, CL 240 ms) and was cardioverted. Electrophysiological study was performed using CARTO® (Optrell®) without induction of VT. Isthmus 3 showed slow conduction, but because of normal voltage no ablation was performed in Isthmus 3. After another episode of VT, a re-procedure showed low-voltage in Isthmus 3 and again slow-conduction (CARTO® – Decanav®). The clinical VT could be induced and was overpaced by hemodynamic instability. Pace map showed high concordance (94%) to induced VT by pacing from a small healthy area lateral between the native RVOT and the Conduit. Coronary angiography showed close proximity of the right coronary artery to this critical isthmus, so no ablation using Radiofrequency was performed in this area. In a re-procedure the critical isthmus was ablated using PFA (NavX®-Tactiflex Duo®). Coronary angiography before and after each pulse revealed coronary spasm, so Nitroglycerin was injected systemically and intracoronary with recovery after several minutes. Additionally RF-ablation of Isthmus 3 was performed. After the ablation no monomorphic VT was inducible, however a new polymorphic VT was induced. Implantation of an ICD was discussed with the parents. They decided at this time against an ICD, but a re-procedure with re-assessment of VT inducibility was scheduled 4 months after (08/2026).

Conclusion: Pulsed field ablation with parallel intracoronary Nitroglycerin-injection is a strategy in VT-ablation after rTOF in case of close proximity of the critical isthmus and a coronary artery. However, proarrhythmic effects of this ablation method, potentially mediated by transient ischemia through the spasm of the coronary artery, are possible.

**PO.50 | HOMOZYGOUS TAF1A VARIANT CAUSING SEVERE PEDIATRIC DILATED CARDIOMYOPATHY WITH REFRACTORY MULTIFOCAL ATRIAL TACHYARRHYTHMIAS IN TWO COUSINS FROM A CONSANGUINEOUS FAMILY**

Betul Guzelhan¹, Damla Yildiz², Pelin Ozlem Simsek Kiper², Eda Utine², Ilker Ertugrul¹, Tevfik Karagoz¹

¹ Pediatric Cardiology, Hacettepe University Faculty of Medicine

² Pediatric Genetics, Hacettepe University Faculty of Medicine

Background and Aim: TAF1A encodes a subunit of the RNA polymerase I complex critical for ribosomal RNA synthesis. Biallelic TAF1A variants cause a cardiac-specific ribosomopathy; all previously reported cases involved compound heterozygous mutations. Refractory multifocal atrial tachyarrhythmias have been described as sentinel events in TAF1A-related cardiomyopathy, yet the full electrophysiological phenotype remains incompletely characterised. We report the first familial case of homozygous TAF1A-related dilated cardiomyopathy (DCM), with particular focus on the arrhythmia burden and its clinical implications.

Methods: Two first cousins from a consanguineous Turkish family presenting with unexplained early-onset DCM underwent clinical evaluation, including echocardiography, cardiac magnetic resonance imaging, serial Holter monitoring, and electrophysiological assessment.

Results: Whole-exome sequencing revealed a homozygous TAF1A missense variant (NM_005681.4: c.941A>G, p.His314Arg) in both cousins; all four parents were confirmed heterozygous carriers. The proband presented at age 2.5 years with DCM (LVEF 37%), progressing to severe systolic dysfunction (LVEF 17%) by age 6. Her cousin presented at age 8 with severely impaired left ventricular function. Both patients developed refractory multifocal atrial tachyarrhythmias unresponsive to multiple antiarrhythmic agents (carvedilol, amiodarone, lidocaine, sotalol, propafenone, propranolol, and flecainide) and not amenable to catheter ablation due to multifocal origin. Progressive electromechanical deterioration was complicated by intracardiac thrombus formation and cardioembolic events. Both required extracorporeal membrane oxygenation as a bridge to transplantation and died during circulatory support (day 181 and day 79, respectively).



Conclusions: This is the first report of homozygous TAF1A-related DCM, expanding the genotypic spectrum of cardiac ribosomopathies. Refractory multifocal atrial tachyarrhythmias with cardioembolic complications, evolving alongside progressive ventricular dysfunction, constitute a distinctive electromechanical phenotype that may serve as a clinical red flag for TAF1A variants. Early recognition, aggressive rhythm monitoring, and timely transplant listing are essential.



PO.51 | CARDIAC ARREST DUE TO BRADYCARDIA-DEPENDENT TORSADES DE POINTES IN PEDIATRIC IDIOPATHIC COMPLETE ATRIOVENTRICULAR BLOCK

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¹ Department of Pediatrics and Adolescent Medicine, Academic Feldkirch Teaching Hospital, Feldkirch, Austria,

² Department of Pediatrics and Adolescent Medicine, Dornbirn Hospital, Dornbirn, Austria,

Introduction

Complete atrioventricular block (CAVB) in children may be complicated by bradycardia-dependent QT prolongation and Torsade de pointes (TdP), a potentially underrecognized mechanism of syncope and cardiac arrest in this population. We report a previously healthy eight years old child presenting with cardiac arrest as the first manifestation of idiopathic CAVB.

Case Description

An eight-year-old previously healthy girl presented after an abrupt syncopal episode with transient cyanosis and brief seizure-like activity. ECG revealed complete proximal third-degree AV block with a junctional escape rhythm (HR 49/min) and a borderline prolonged QTc of 455 ms (Figure 1). Echocardiography showed a structurally normal heart with preserved biventricular function. Electrolytes were unremarkable. Over the next 24 hours, QTc prolonged to 618 ms without requiring intervention (Figure 2). During resistance against peripheral intravenous cannulation, the patient developed tachycardia, which degenerated into polymorphic ventricular tachycardia (Figure 3). CPR was initiated immediately. ROSC was achieved after seven minutes by cardiac defibrillation. Electrolytes including magnesium and potassium were administered, and a temporary transvenous pacing lead was placed via the right internal jugular vein, achieving hemodynamic stabilization under VVI pacing at 80 bpm. Comprehensive etiologic workup — including infectious serologies, autoimmune screening, metabolic evaluation, and genetic testing — was unrevealing. The patient was referred for permanent pacemaker implantation. Following initiation of permanent ventricular pacing, QTc normalized with no further ventricular arrhythmias. An ICD was deemed unnecessary. At follow-up, the patient remained asymptomatic with stable pacing parameters, complete ventricular pacing dependence, and preserved biventricular function.

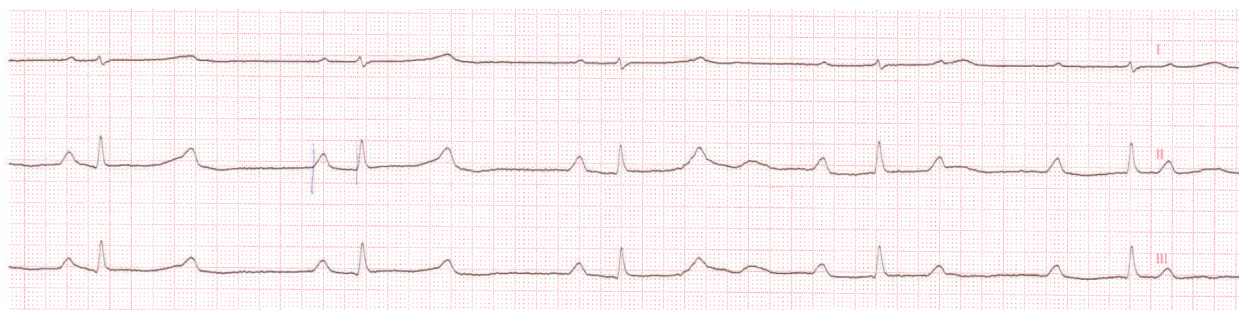


Figure 1: ECG on Admission showing CAVB with borderline QTc Prolongation; QTc: 455 ms.

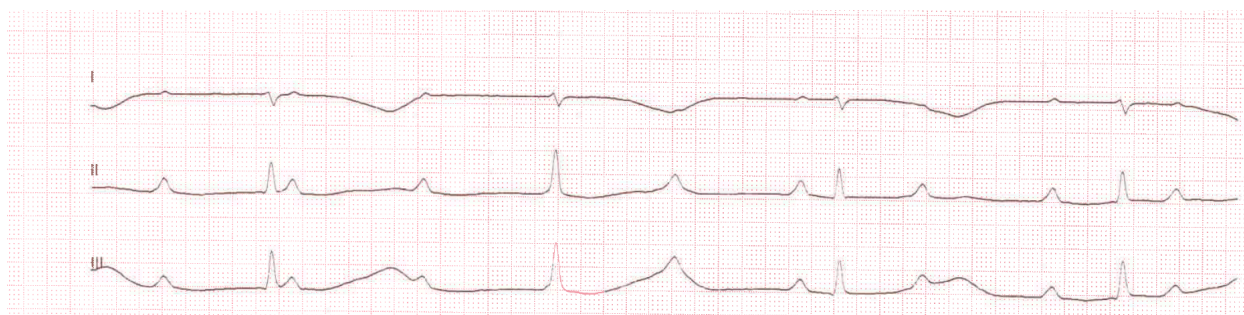


Figure 2: ECG 24 hours after admission showing CAVB with significant QTc Prolongation; QTc: 618 ms.

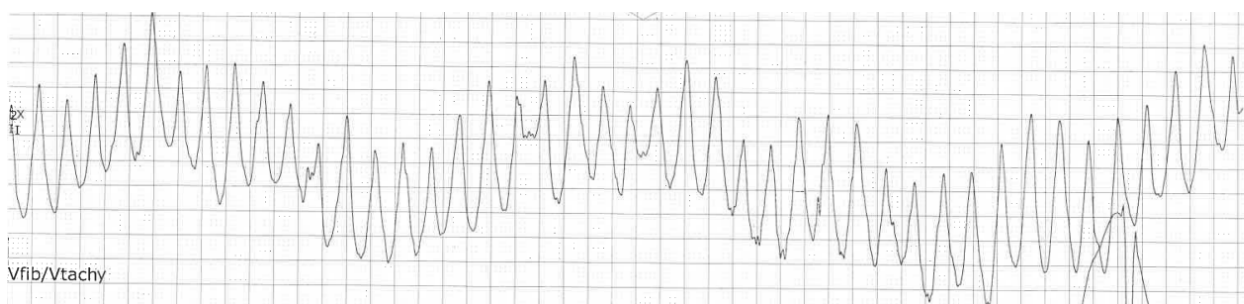
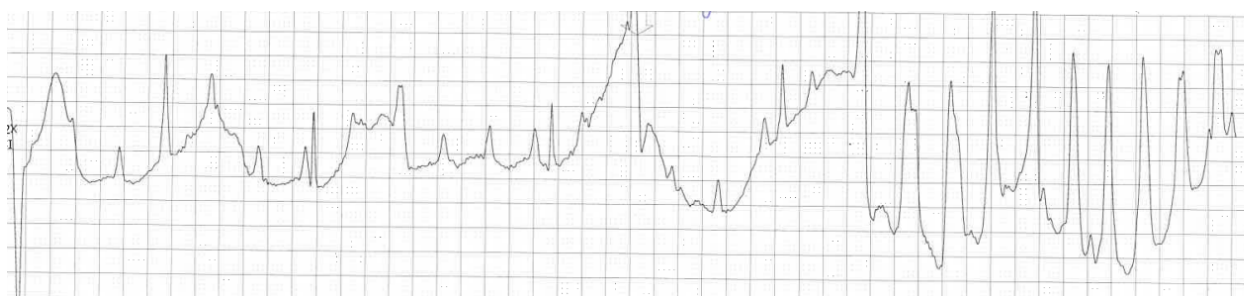


Figure 3: Telemetry recording showing the transition from CAVB to polymorphic ventricular tachycardia



Conclusion

Although bradycardia due to CAVB may be hemodynamically tolerated, associated QTc prolongation significantly increases the risk of life-threatening tachyarrhythmias, particularly TdP. Acute management should include immediate discontinuation of QTc-prolonging medications and maintenance of magnesium and potassium in the upper normal range. For recurrent or refractory TdP, increasing heart rate via transvenous overdrive pacing or intravenous isoproterenol is a critical short-term intervention. Hemodynamically unstable TdP requires immediate unsynchronized defibrillation. Early recognition of bradycardia-induced QTc prolongation should prompt timely evaluation for permanent pacing¹⁻⁵.

This case further supports current recommendations for permanent pacemaker implantation in pediatric patients with clinically significant pause-dependent ventricular tachycardia associated with CAVB⁶.

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**PO.52 | CARDIONEUROABLATION FOR COEXISTING INSULAR AUTONOMIC SEIZURES AND SEVERE CARDIOINHIBITORY SYNCOPE IN A CHILD: A PEDIATRIC BRAIN-HEART INTERPLAY, CASE REPORT****Aycan Çelebi¹, İlker Ertuğrul¹, Ülkühan Öztoprak², Tevfik Karagöz¹**¹ Department of Pediatric Cardiology, Hacettepe University Faculty of Medicine, Ankara, Turkey² Department of Pediatric Neurology, Hacettepe University Faculty of Medicine, Ankara, Turkey**Background and Aim**

Reflex syncope, characterized by transient cerebral hypoperfusion via inappropriate autonomic activation, often mimics epileptic seizures, complicating pediatric differential diagnosis. We report a pediatric patient with coexisting epileptic seizures and cardioinhibitory reflex syncope (marked by vagally mediated severe bradyarrhythmia) who was successfully treated with cardioneuroablation.

Case Report

A 9-year-old girl presented with recurrent syncopal episodes characterized by pallor, brief extremity twitching, and transient loss of consciousness, typically triggered by emotional stress or nausea/vomiting, and resolving spontaneously within 1–2 minutes. Initial cardiological evaluation, including electrocardiography, echocardiography, and tilt-table testing, was unremarkable. Due to suspicious findings on routine electroencephalography (EEG), the patient was admitted for video-EEG monitoring. Simultaneous cardiac monitoring and ictal recordings captured one episode of reflex syncope during blood sampling and two focal autonomic seizures (**Figure 1 and 2**). During syncope, prodromal autonomic symptoms were followed by progressive bradyarrhythmia, evolving from a 2:1 to a complete AV block lasting 20–30 seconds, causing loss of consciousness. Concurrent EEG recordings demonstrated cerebral hypoperfusion, with consciousness immediately recovering after the restoration of sinus rhythm. Levetiracetam was initiated because the autonomic manifestations raised suspicion of insular-origin seizures. However, stress-induced bradyarrhythmic episodes persisted despite the therapy. The findings were consistent with cardioinhibitory reflex syncope secondary to hypervagotonia, and the patient underwent cardioneuroablation. Ablation was performed using an irrigated catheter targeting fractionated electrograms in the left atrial posteroseptal region, followed by additional lesions targeting the ganglionated plexi in the left posteromedial and right atrium–superior vena cava junction (**Figure 3**).



After the procedure, the heart rate increased from 60 to 100 beats/min. Follow-up Holter monitoring demonstrated reduced heart rate variability, repeat video-EEG was normal, and no further syncopal episodes were observed.

Discussion

In patients with a preliminary diagnosis of epilepsy, reflex syncope should always be considered in the differential diagnosis, particularly when episodes of loss of consciousness are triggered by specific factors. In recent years, cardioneuroablation has emerged as a promising alternative to pacemaker implantation in selected patients with predominant vagal overactivity. However, current evidence is mainly derived from adult case series, highlighting the need for multicenter studies evaluating long-term outcomes in pediatric patients.

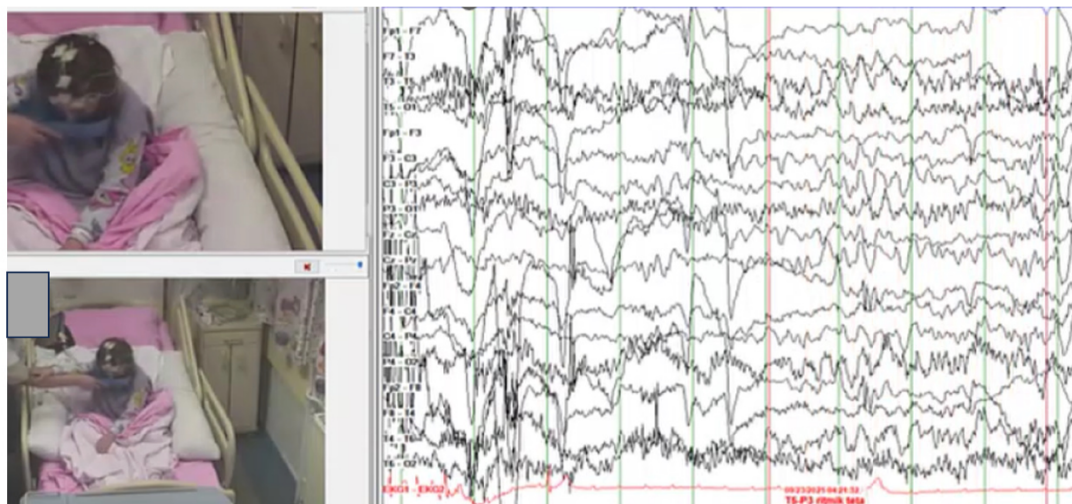


Figure 1. During an autonomic seizure characterized by awakening from sleep, nausea, vomiting, and retching with preserved consciousness, progressive bradyarrhythmia, complete atrioventricular block (lower ECG tracing), and simultaneous slowing of theta-wave activity were observed.

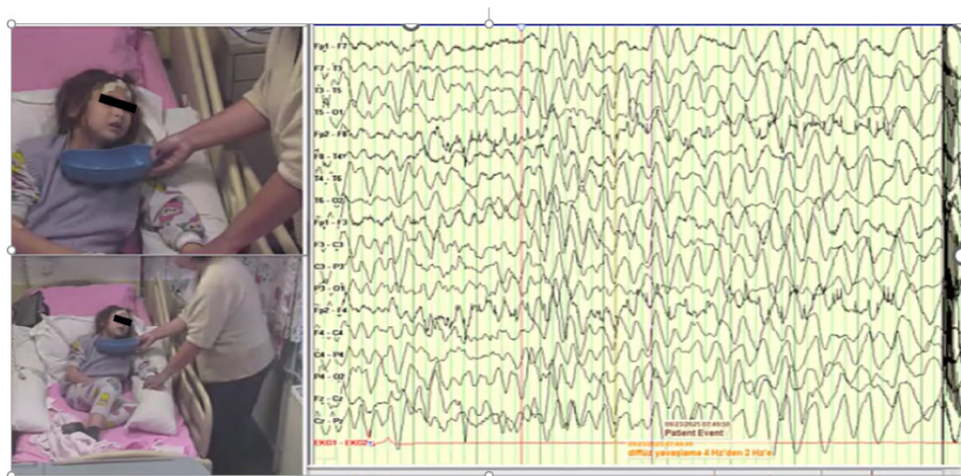


Figure 2. During a reflex anoxic seizure triggered by venipuncture, a complete atrioventricular block was observed on ECG, accompanied by diffuse delta-wave activity on EEG.

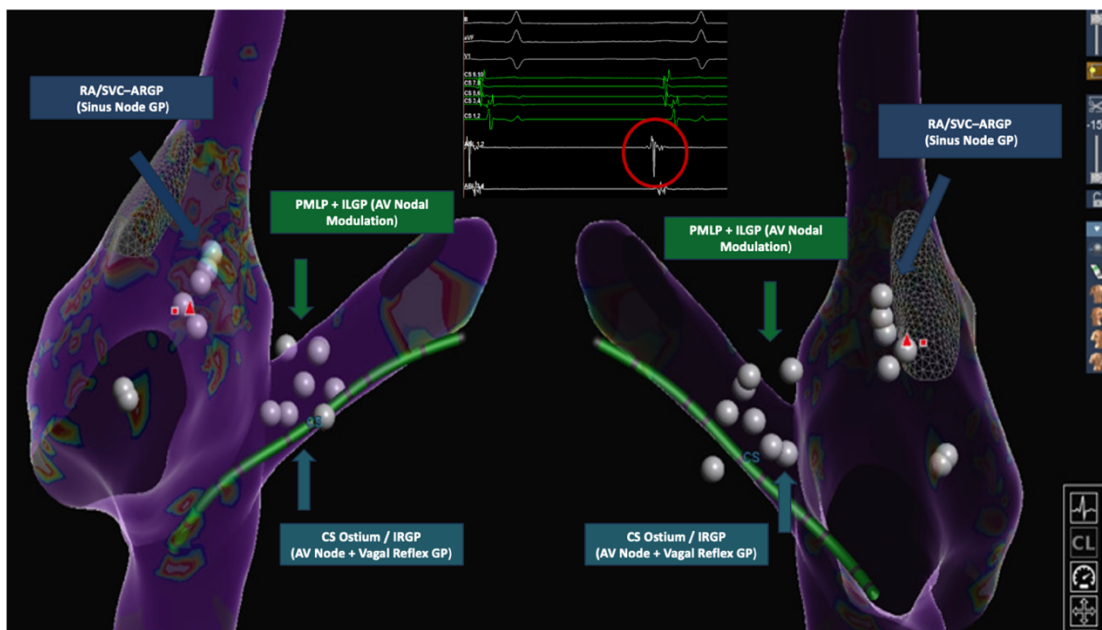


Figure 3. The left atrium was accessed via a transseptal approach using an Agilis sheath. Radiofrequency ablation was performed using an irrigated-tip catheter (30 W, 45°C). Because of the underlying atrioventricular block, ablation was limited to the ganglionated plexi involved in AV nodal and vagal modulation. The target sites included fractionated electrograms in the left atrial posteroseptal region, coronary sinus ostium (right posteroseptal region), and dominant RA-SVC ganglionated plexus.

**PO.53 | PRECLINICAL AND BENCHTOP EVALUATION OF A DEDICATED TOOL FOR INSERTABLE CARDIAC MONITOR EXPLANTATION**

Ryan O'Hara¹, Bryan Gonzalez¹, Devon Guerrelli¹, Mischa Cho¹, Vanessa Ogueri¹, Charles Berul¹

¹ Cardiology, Children's National Hospital

Introduction: Insertable cardiac monitors (ICMs) are increasingly used for long-term rhythm monitoring in adults and children, including those with congenital or inherited arrhythmia syndromes. However, device removal can be challenging in pediatric patients due to smaller anatomical dimensions, variable tissue characteristics, and a propensity for fibrotic encapsulation and device migration over time. These factors often necessitate sharp or blind dissection, increasing risk of tissue trauma, procedural difficulty, and pain. We developed the first dedicated ICM removal tool designed to safely dissect fibrotic encapsulation and securely engage the device, with the goal of improving the safety and efficiency of explantation.

Methods: A novel ICM-specific removal tool compatible with several available ICMs was designed and manufactured (Figure 1).

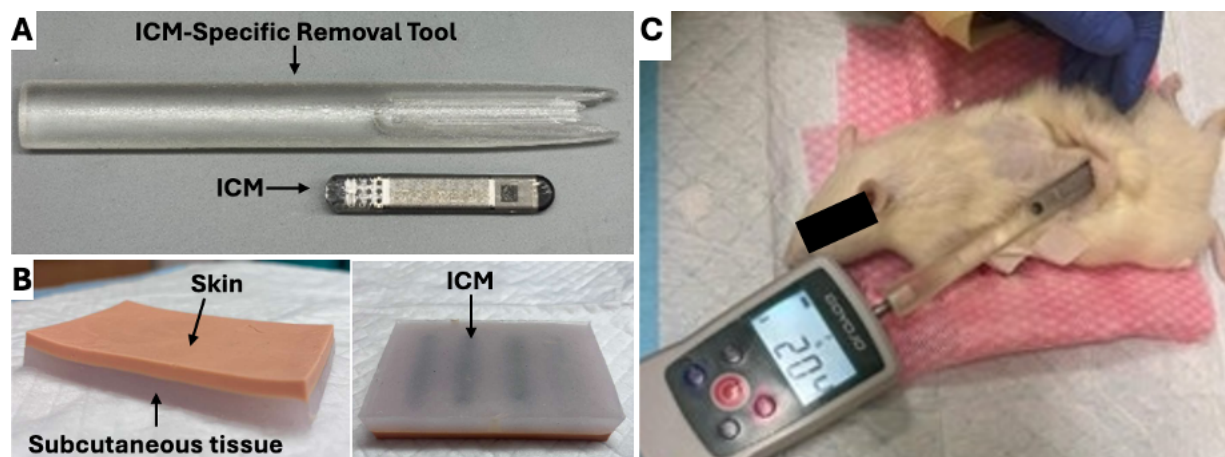
Benchtop Study: Tool performance was tested in a fibrotic encapsulation analogue exceeding the durometer of healed fascia and subcutaneous tissue. Eighty trials were performed (40 tool, 20 forceps, 20 direct pullout). Removal times and insertion and removal forces were measured.

Preclinical Study: Performance was evaluated in a chronic rat model. In each of 9 animals, 2 ICMs were implanted bilaterally. After 3 months, devices were explanted using either standard forceps or the novel tool in a randomized order (Figure 1). Time to removal and peak insertion force were recorded.

Results: The tool improved dissection in both biological and synthetic fibrotic models. In the animal model, median removal time was shorter with the tool (82 s [IQR 45.5 - 143.5]) compared with forceps (126 s [77.5 - 159]), a 35% reduction. Encapsulation severity varied between animals but was similar within each paired implant. Insertion forces were higher in the synthetic model than the rat model (36.9 N [34.3 - 38.1] vs 23.6 N [16.8 - 29.5], $p < 0.01$), supporting a conservative benchtop model. In benchtop testing, peak removal force with the tool was markedly lower than direct pullout (10.0 N [9.6 - 11.7] vs 32.3 N [26.0 - 38.9], $p < 0.001$). Benchtop removal time did not differ between the tool and forceps.



Conclusion: We developed the first dedicated tool for ICM explantation. As ICM use expands, particularly in pediatrics where smaller anatomy and longer dwell times may increase explant complexity, procedural demand will continue to grow. A purpose-built removal tool may reduce technical difficulty, tissue trauma, and operator variability during ICM explantation. These findings support continued refinement and larger-scale evaluation, with the potential to enable safer, more controlled ICM removal across diverse patient populations.





PO.54 | CRYOABLATION OF PARA-HISIAN PREMATURE VENTRICULAR CONTRACTIONS IN CHILDREN: A SINGLE-CENTER EXPERIENCE

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¹ Hacettepe University Pediatric Cardiology, Hacettepe University Faculty of Medicine, Ankara, Türkiye

Background and Aim: Catheter ablation of para-Hisian premature ventricular contractions (PVCs) is challenging because of the close proximity to the atrioventricular conduction system. Data regarding the safety and efficacy of cryoablation in pediatric patients are limited.

Methods: We retrospectively reviewed seven consecutive pediatric patients who underwent cryoablation for para-Hisian ventricular arrhythmias at a tertiary referral center. Clinical characteristics, surface electrocardiographic findings, procedural data, complications, and follow-up outcomes were analyzed.

Results: Seven patients (median age 14 years, range 5–15 years) underwent cryoablation for para-Hisian PVCs. PVC burden ranged from 16% to 78% (median 25%). Among patients with available electrocardiograms, PVC morphology was characterized by an inferior axis, left bundle branch block–type pattern, dominant negativity in lead V1, and early precordial transition, consistent with a para-Hisian or high septal origin. Acute procedural success was achieved in all patients. No atrioventricular conduction disturbances occurred. Two patients developed right bundle branch block following ablation. During a median follow-up of 41 months (range 9–54 months), recurrence was observed in one patient, resulting in a long-term success rate of 85.7%.

Conclusions: Cryoablation appears to be a safe and effective treatment option for pediatric para-Hisian ventricular arrhythmias. High acute success, low recurrence rates, and the absence of permanent atrioventricular conduction injury support the use of cryoenergy in this anatomically challenging region.

**PO.55 | SUCCESSFUL CONDUCTION SYSTEM PACING IN A PATIENT WITH
HYPERTROPHIC CARDIOMYOPATHY POST MYECTOMY****Mathan Munusamy¹, Hasri Samion¹**¹ Paediatric and Congenital Heart Center, National Heart Institute Kuala Lumpur

Background and Aim: Complete atrioventricular block and left bundle branch block are common complications following surgical myectomy to relieve left ventricular outflow tract (LVOT) obstruction in hypertrophic cardiomyopathy. This could lead to deleterious decline in ventricular function. Conduction system pacing can be a reasonable approach to overcome the conduction block and improve ventricular synchrony.

Methods:

A 26-year-old girl with Noonan Syndrome and hypertrophic cardiomyopathy presents with poor ventricular function during follow up. She had LVOT myectomy at the age of 14 complicated by complete atrioventricular block requiring placement of endovascular dual chamber pacemaker. She soon regained 1:1 AV conduction, though remained in 1st degree AV block with PR interval 194 msec with left bundle branch block (LBBB) resulting in QRS duration of 145 msec. Echocardiogram revealed LV ejection fraction of 53% with elevated proBNP.

Following careful evaluation, we decided to perform a left bundle branch area pacing (LBBAP) to shorten the PR interval and overcome the LBBB. A third lead using the UltiPace Pacing Lead from Abbott targeting the left bundle area was performed resulting in QRS duration of 120 msec without LBBB. Post procedure echocardiogram revealed immediate improvement of the LVEF to 60% and the patient was discharged well.

Discussion and Conclusion:

This case illustrates the therapeutic utility of conduction system pacing to improve LV function in the setting of LBBB post surgical relief of LVOT obstruction in the setting of hypertrophic cardiomyopathy. In resource limited setting, this may alleviate the need for ICD placement by improving ventricular function.

**PO.56 | MODELING SINUS NODE DYSFUNCTION WITH PATIENT-DERIVED HUMAN STEM CELLS HARBORING HCN4 MUTATION**

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Background:

Mutations in HCN4, an ion channel gene that drives pacemaker activity in the sinoatrial node (SAN), are a cause of familial sinus node dysfunction (SND). SND manifests as sinus bradycardia and chronotropic incompetence, an inadequate heart rate response to autonomic stimulation. Human induced pluripotent stem cell (hiPSC)-derived cardiomyocyte monolayers (CMs) recapitulate SAN-like automaticity driven by HCN4-mediated current, I_f . Current management remains limited to pacemaker implantation, underscoring the need to understand HCN4 genotype-phenotype relationships.

Methods:

hiPSC lines harboring homozygous and heterozygous HCN4 p.Thr789Ile variants and a wild-type (WT) line were differentiated into CMs. Spontaneous beat rates were recorded by microelectrode array over 5-minute intervals at baseline and during dose-response experiments (0.03–30 μ M) with isoproterenol, carbachol, and ivabradine. Beat rate variability (BRV) was quantified using SDNN, RMSSD, and Poincaré indices (SD1, SD2).

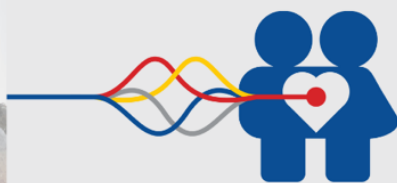
Results:

Basal beat rates were lower in homozygous CMs (72.1 ± 24.5 bpm) and heterozygous CMs (95.7 ± 39.5 bpm) than WT CMs (126.9 ± 35.4 bpm), consistent with impaired I_f -driven automaticity. Isoproterenol produced a peak chronotropic response of +40% in WT CMs (132 ± 25 to 185 ± 36 bpm), versus a blunted peak of +27% in homozygous CMs (70 ± 26 to 88 ± 27 bpm). Heterozygous CMs showed an intermediate response (peak isoproterenol response +21%), consistent with partial I_f impairment. Carbachol produced dose-dependent rate reductions in both homozygous (up to –37%) and WT CMs (up to –23%). Ivabradine reduced beat rate in WT CMs (up to –32%), but paradoxically increased beat rate in homozygous CMs (up to +28%). BRV was markedly elevated in homozygous CMs (RMSSD 22.8–396 ms, SD1/SD2 0.20–0.64) compared to WT CMs (RMSSD 1.7–2.2 ms, SD1/SD2 0.15–0.37), indicating greater intrinsic rhythm instability consistent with increased heart rate variability in patients with HCN4 point mutations.



Conclusions:

HCN4 mutant hiPSC-CMs recapitulate the hallmarks of SND including baseline bradycardia, chronotropic incompetence, and elevated BRV with a genotype-dependent severity gradient. These findings validate hiPSC-CMs as a human cellular model of HCN4-related SND and provide a platform for mechanistic and therapeutic discovery for SND.

**PO.57 | SAME-DAY DISCHARGE AFTER ELECTROPHYSIOLOGY PROCEDURES: A SAFE, FEASIBLE OPTION FOR PEDIATRIC AND ADULT CHD PATIENTS****Rayan Halabi¹, Khalid Dagreri¹, Lamia Al Shengeiti², Saleh Al Ghamdi¹, Ahmed Al Fagih²**¹ Pediatric Cardiac Electrophysiology,² Adult cardiac electrophysiology,**Objectives:**

This study aimed to evaluate the safety of same-day discharge for day-case electrophysiology (EP) procedures in pediatric and adult patients with congenital heart disease. Additionally, it sought to identify factors contributing to longer post-procedure observation durations and to assess differences in observation times between patients undergoing general anesthesia (GA) and those receiving conscious sedation.

Design:

A retrospective observational study was conducted.

Setting:

The study was carried out at a single tertiary specialized cardiac center.

Results:

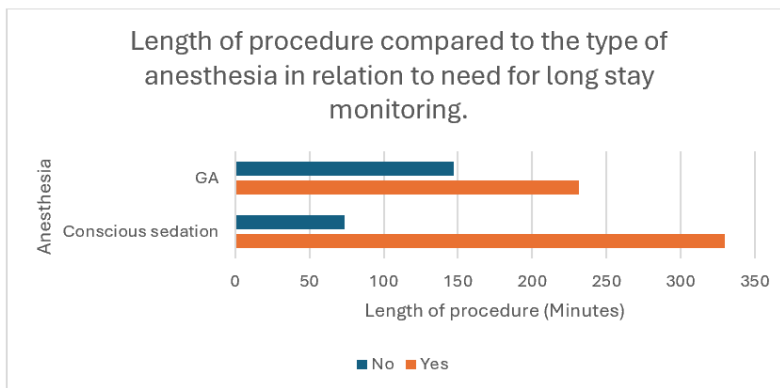
A total of 96 cases were identified, of which 77 met the inclusion criteria and were included in the analysis. Among these, 65 patients underwent electrophysiology studies, while 12 received device implantations. The majority of patients were discharged on the same day, with 72.7% monitored for 4-6 hours post-procedure. Factors influencing longer observation included patient age and procedural duration, with older patients requiring extended monitoring. No significant adverse events were reported.

Conclusions:

Same-day discharge for day-case EP procedures is safe for both pediatric and adult patients with congenital heart disease. Further research is needed to refine guidelines for post-procedure care based on patient demographics and procedural complexity.



Average Procedural Duration (Minutes) in correlation with post-procedure observation for each age group and type of anesthesia			
Anesthesia	age group	Post-procedure observation	
		< 6 hours	6-12 hours
Conscious sedation	> 18 years	43	330
	10 to 18 years	109	-
	< 10 years	60	-
Procedure Duration average For Conscious sedation Cases (Minutes)		73	330
General Anesthesia	> 18 years	157	243
	10 to 18 years	147	177
	< 10 years	141	290
Procedure Duration average For General Anesthesia Cases (Minutes)		150	230
Grand average		140	227



**PO.58 | ECTOPIC JUNCTIONAL TACHYCARDIA IN TAB2-RELATED CARDIAC SYNDROME:
EXPANDING THE ARRHYTHMIC PHENOTYPE**

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Background and Aim

TAB2 loss-of-function variants are associated with congenital heart defects, valvular abnormalities, and progressive cardiomyopathy. Although arrhythmias have been reported in affected individuals, their clinical spectrum remains incompletely defined. We describe a patient with TAB2-related syndrome who developed persistent ectopic junctional tachycardia (EJT) associated with structural and myocardial abnormalities.

Methods

A male patient with secundum atrial septal defect repaired percutaneously during childhood underwent genetic testing after identification of a pathogenic TAB2 variant within the family. Serial clinical, echocardiographic, cardiac magnetic resonance (CMR), electrocardiographic, Holter, and implantable loop recorder evaluations were reviewed.

Results

Genetic analysis identified the heterozygous pathogenic splice-site variant c.1604-1G>A in TAB2. Extra-cardiac findings included short stature, facial dysmorphism, and bilateral mixed hearing impairment.

Despite complete defect closure and absence of residual shunt, serial echocardiography demonstrated progressive right ventricular enlargement, moderate tricuspid regurgitation, and tricuspid valve apparatus dysplasia. Left ventricular systolic function remained preserved, although mild basal septal dyskinesia was consistently observed.



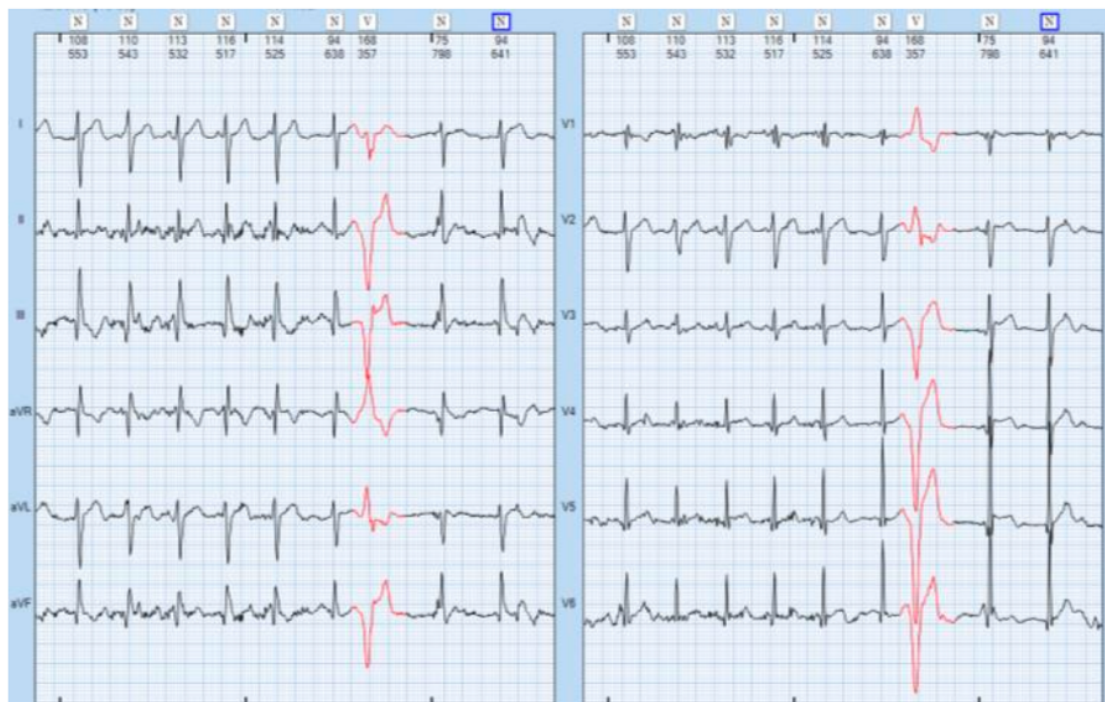
CMR showed right ventricular dilatation (RV end-diastolic volume indexed 133 mL/m²), mildly reduced right ventricular ejection fraction (48%), preserved left ventricular function (57%), and focal non-ischaemic late gadolinium enhancement involving the basal anteroseptal region and posterior insertion point.



Baseline ECGs demonstrated sinus rhythm with incomplete right bundle branch block. Twenty- four-hour Holter monitoring revealed frequent episodes of EJT and rare isolated premature ventricular contractions. Due to the unexpected arrhythmic burden, an implantable loop recorder was implanted and documented recurrent junctional tachycardia episodes reaching 230 bpm. Treatment with bisoprolol and ivabradine resulted in a marked reduction of clinically significant arrhythmic events during follow-up. Ventricular function remained stable and the patient remained largely asymptomatic.



Conclusions



This case expands the arrhythmic spectrum associated with TAB2-related cardiac disease by documenting sustained EJT requiring long-term rhythm surveillance and pharmacological treatment. The coexistence of right ventricular remodelling, focal myocardial fibrosis on CMR, and junctional tachyarrhythmia suggests a potential myocardial-conduction phenotype within TAB2 related syndrome. Multimodality imaging and prolonged rhythm monitoring may be valuable for early detection and management of arrhythmic manifestations in affected patients.



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PO.59 | THE EFFICIENCY OF LEFT CARDIAC SYMPATHETIC DENERVATION IN PAEDIATRIC PATIENTS WITH CPVT – A SINGLE CENTRE EXPERIENCE

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Background

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare inherited cardiac channelopathy characterized by adrenergically mediated polymorphic ventricular arrhythmias occurring in the absence of structural heart disease. Currently there are limited data on effectiveness of cardiac sympathetic denervation (LCSD) among paediatric population.

Aim

Analysis of the occurrence of complex ventricular arrhythmias among patients diagnosed with CPVT who underwent LCSD.

Methods

This is a retrospective analysis of ventricular arrhythmia among patients diagnosed with CPVT before and after LCSD. Authors made a comparison of symptoms and occurrence of ventricular arrhythmia before and after LCSD recorded in Holter ECG and exercise stress test (Bruce Protocol).

Results

9 patients (7 males and 2 females) diagnosed with CPVT underwent LCSD. 7 patients had the RYR2 gene mutation, and 1 patient had CALM 1 mutation. Before LCSD 6 patients experienced syncope and 7 patients experienced sudden cardiac arrest. 2 patients had polymorphic ventricular tachycardia and 2 patients had bidirectional ventricular couplets in ECG Holter. In exercise stress test prior to the LCSD 4 patients had polymorphic tachycardia. In two cases with ICD implanted prior to the LCSD patients received appropriate therapy. In all of cases the pharmacological treatment of beta blocker was implemented. In 6 patients with uncompleted antiarrhythmic effect of beta blocker, the flecainide was added. Both beta-blocker and flecainide were given in maximum tolerated doses. In 2 cases ICD was implanted at the same time of LCSD.

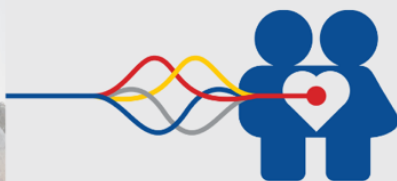


Documented polymorphic ventricular tachycardia despite pharmacological treatment was an indicator for LCSD. The mean age of LCSD was 10.7 years old.

In early post-operative period there were no major complications. The mean years of observation was 4.4years. None of the patients reported syncope. In ECG Holter after LCSD premature ventricular complexes were detected. Exercise stress after LCSD was performed in 7 cases in which both single premature ventricular complexes and ventricular bigeminies occurred. In cases with implanted ICD no interventions were recorded.

Conclusions

In presented group of patients diagnosed with CPTV, LCSD resulted in complete polymorphic ventricular tachycardia reduction and this method is safe and highly efficient sudden cardiac death prevention.

**PO.60 | THE ROLE OF ADENOSINE TESTING AFTER PEDIATRIC ACCESSORY PATHWAY ABLATION: DETECTION OF LATENT CONDUCTION AND PREDICTION OF RECURRENCE**

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Background and Aim: Radiofrequency ablation of atrioventricular accessory pathways (APs) is highly effective in pediatric patients. However, recurrence may occur despite apparent acute procedural success because of latent AP conduction. Through its hyperpolarizing effect on AP tissue, adenosine may unmask residual conduction.

This study aimed to evaluate the ability of adenosine testing to detect latent conduction in manifest and concealed APs, its potential to replace the conventional 30-minute waiting period, and its association with late recurrence.

Methods: This prospective cohort study included 114 children who underwent successful AP ablation. Adenosine was administered 5–10 minutes after ablation, following electrophysiological confirmation of AP block. If adenosine induced transient recovery of AP conduction, additional ablation was performed until the test became negative. Recurrence analysis was limited to patients with manifest APs. Early recurrence was assessed 30 minutes after ablation and late recurrence at the 6-week follow-up, based on ECG findings and clinical presentation. Logistic regression analysis was used to investigate whether an initially positive adenosine test or AP location predicted early or late recurrence.

Results: Latent AP conduction was detected by adenosine testing in 12 of 114 patients (10.5%). The frequency of a positive adenosine test did not differ significantly between manifest and concealed APs ($p=0.724$). Only 1 of 89 patients with manifest APs experienced recurrence within 30 minutes (1.1%). The 6-week recurrence rate was higher among patients with an initially positive adenosine test than among those with a negative test (44.4% vs. 15.0%; $p=0.051$). An initially positive adenosine test was associated with increased odds of late recurrence, although the association did not reach statistical significance (OR 3.67, 95% CI 0.96–14.05; $p=0.058$). AP location was significantly associated with 6-week recurrence ($p=0.001$); no recurrences occurred in left lateral APs, whereas recurrence rates were 24.1% for right lateral and 21.4% for septal APs.



Conclusions: Adenosine testing effectively identifies latent AP conduction after pediatric ablation and may replace the conventional 30-minute waiting period. The trend toward higher 6-week recurrence among patients with an initially positive adenosine test and the significant association between AP location and recurrence underscore the importance of careful follow-up.

**PO.61 | DELAYED HEART BLOCK AFTER PERIMEMBRANOUS VSD DEVICE CLOSURE:
RECOVERY OF CONDUCTION AFTER SURGICAL DEVICE REMOVAL IN TWO PATIENTS**

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Mohamed Al Nassef³, Ahmed Al Fagih²**

¹ Pediatric Cardiac Electrophysiology,

² Adult cardiac electrophysiology,

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Background

Delayed atrioventricular block (AV block) is an uncommon but serious complication of transcatheter closure of perimembranous ventricular septal defects (pmVSD). Management remains challenging, particularly when conduction abnormalities develop months after an apparently successful procedure. Whether late surgical removal of the device can restore atrioventricular conduction is not well established.

Case Summary

We describe two patients who developed delayed heart block after transcatheter pmVSD closure and subsequently recovered sinus rhythm following surgical device removal.

The first patient was a 15-year-old boy with Down syndrome and a pmVSD with muscular extension. One week after device closure, he presented with recurrent syncopal episodes and complete heart block [fig1] requiring temporary pacing. Surgical exploration revealed a large device compressing the tricuspid septal leaflet and entrapped within the septal chordae. The device was removed and the defect was closed surgically. The patient gradually recovered normal atrioventricular conduction and remained in stable sinus rhythm during follow-up [Fig2].

The second patient was a 2-year-old boy who underwent pmVSD closure using an ADO-I device. Three and a half months later, routine follow-up revealed advanced second-degree AV block with intermittent episodes of complete heart block [Fig3], despite the absence of symptoms. Holter monitoring demonstrated significant bradycardia with a minimum heart rate of 42 bpm. Surgical exploration showed the device impinging on the tricuspid valve apparatus and adherent to surrounding tissues. Device removal, VSD patch closure, and repair of the tricuspid and aortic valves were performed. Sinus rhythm returned within five days of surgery and remained stable thereafter [Fig4].



Conclusion

These cases highlight that delayed heart block can occur days to months after pmVSD device closure, even after an initially uneventful procedure. Mechanical compression and progressive tissue reaction around the conduction system may play an important role. Importantly, recovery of atrioventricular conduction is possible following surgical device explantation, even when performed several months after the onset of heart

block. In carefully selected patients, surgical removal of the device should be considered before committing to permanent pacemaker implantation.

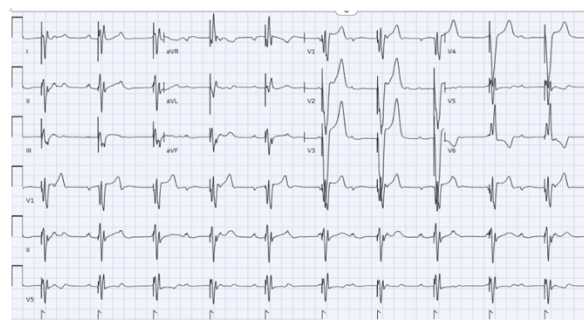


Fig 1: 12 leads ECG for the first patient showing complete heart block one week post VSD device closure

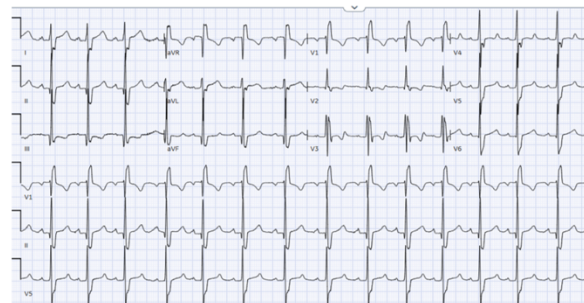


Fig 2: 12 leads ECG for the first patient showing normal sinus rhythm which was taken on the follow up

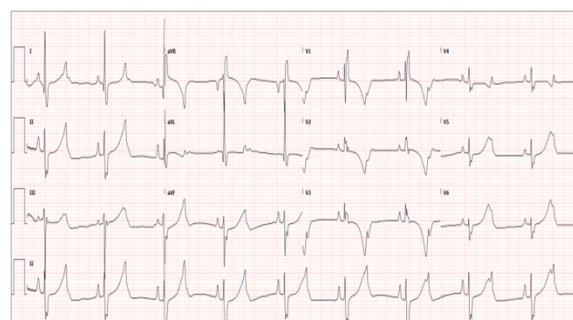


Fig 3: 12 leads ECG for the second patient showing advanced 2:1 2nd degree heart block post device

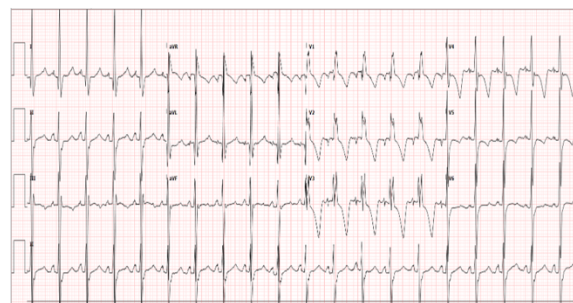


Fig 4: 12 leads ECG for the second patient showing normal sinus rhythm 5 days post device extraction



**PO.62 | CATECHOLAMINERGIC POLYMORPHIC VENTRICULAR TACHYCARDIA
ASSOCIATED WITH SINUS NODE DYSFUNCTION IN AN 11-YEAR-OLD BOY
MISDIAGNOSED WITH EPILEPSY FOR FIVE YEARS**

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⁷ Diagnostic Department, Ternopil Perinatal Center "Mother and Child"

Background. Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) is the most common phenotype of cardiac ryanodinopathies. Sinus node dysfunction (SND) is increasingly recognized as part of the CPVT phenotype, particularly in patients carrying pathogenic *RYR2* variants.

Aim. To present a case of CPVT associated with severe sinus node dysfunction that was initially misdiagnosed as epilepsy

Methods. Clinical evaluation, electrocardiography, transthoracic echocardiography, 24-hour Holter monitoring, genetic testing, and follow-up assessment were performed.

Results. An 11-year-old boy was referred to a pediatric cardiologist by a pediatric neurologist because of recurrent presyncope and syncope. Symptoms were triggered by sudden auditory stimuli, such as a school bell or air-raid alarm, and were accompanied by weakness and exercise intolerance. The symptoms had been present for five years, during which he had been treated for epilepsy with sodium valproate (450 mg twice daily).

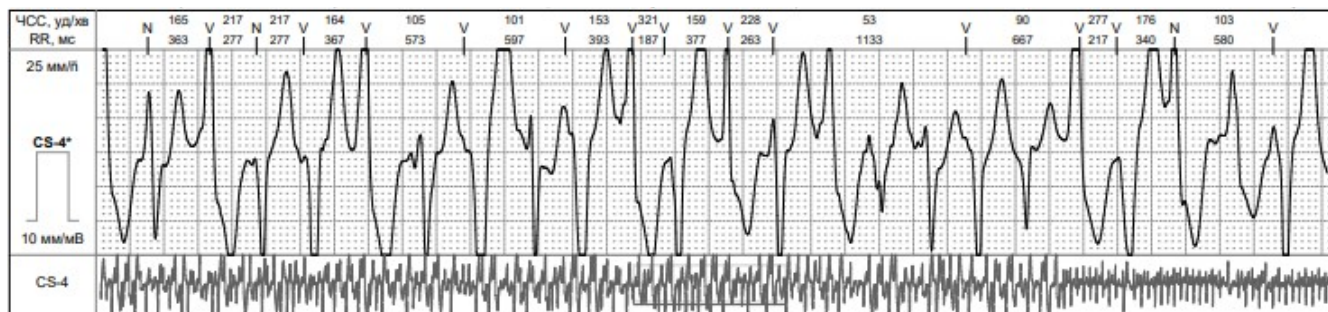


Resting electrocardiography demonstrated marked sinus bradycardia (38–40 bpm) and borderline QT prolongation (QTc 485 ms by Hodges, 454 ms by Fridericia, and 462 ms by Rautaharju formula). Echocardiography revealed no structural heart disease. Family history was unremarkable. 24 h ECG showed a minimum heart rate of 32 bpm and a maximum heart rate of 210 bpm during exercise. Episodes of narrow QRS tachycardia with gradual onset and termination transformed into wide QRS tachycardia. Sinoatrial block was also documented.

The initial diagnosis was symptomatic sinus node dysfunction. A dual-chamber permanent pacemaker (DDIR mode, lower rate limit 70 bpm) was implanted, followed by propranolol therapy (2 mg/kg/day). This resulted in marked clinical improvement, including resolution of dizziness during stair climbing and weight gain.

Comprehensive genetic testing identified three heterozygous variants: *MYBPC3* c.3697C>T (p.Gln1233*) and two *RYR2* variants, c.13812_13813del and c.13817_13820delinsCCCA. Six months later, recurrent palpitations occurred during bicycle riding. 24 h documented a two-minute episode of bidirectional ventricular tachycardia, confirming the diagnosis of CPVT. The propranolol dose was increased to 4.5 mg/kg/day.

Conclusion. CPVT may present with clinically significant sinus node dysfunction and be misdiagnosed as epilepsy or long QT syndrome, particularly when syncope is triggered by sudden auditory stimuli. Genetic testing and extended rhythm monitoring are crucial for establishing the diagnosis and guiding appropriate management.



**PO.63 | PATENT FORAMEN OVALE–MEDIATED ACCESS VERSUS TRANSSEPTAL PUNCTURE IN PEDIATRIC LEFT-SIDED CATHETER ABLATION PROCEDURES**

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Background:

Left-sided catheter ablation in children needs safe access to the left atrium or cardiac structures. Transseptal puncture is common, but a patent foramen ovale (PFO) may provide access without puncture. Data on these methods in pediatric ablation are limited. This study compares procedural features, fluoroscopy exposure, success, and complications between the two methods.

Methods:

We retrospectively analyzed 405 pediatric patients who underwent left-sided catheter ablation. Patients were divided into two groups based on the access route: patent foramen ovale–mediated access and transseptal puncture. We compared baseline characteristics, procedure and fluoroscopy times, number of lesions, total lesion duration, acute procedural success, and complications between groups.

Results:

A total of 405 pediatric patients were included. PFO access was used in 104 patients, whereas transseptal puncture was performed in 301 patients. The median age was similar between the PFO and transseptal puncture groups [13 years vs. 13 years, $p=0.299$], and body weight was also comparable [47 kg vs. 49 kg, $p=0.143$]. Procedure duration did not differ significantly between groups [90 min vs. 90 min, $p=0.220$].



Fluoroscopy time was significantly lower in the PFO group than in the transseptal puncture group [0 s vs. 15 s, $p<0.001$]. The number of lesions differed significantly between groups, although the median was 3 lesions in both groups [3 vs. 3, $p<0.001$]. Total lesion duration was longer in the patent foramen ovale group [135 s vs. 120 s, $p=0.009$].

Acute procedural success was achieved in 97 of 104 patients in the PFO group and in 291 of 301 patients in the transseptal puncture group [93.3% vs. 96.7%, $p=0.157$], with no statistically significant difference. No major complications occurred in either group. Minor complications were observed in 0 patients in the PFO group and in 1 patient in the transseptal puncture group.

Conclusion:

In pediatric left-sided catheter ablation, PFO-mediated access appears to be a safe and effective alternative to transseptal puncture when anatomically feasible. This approach was associated with significantly lower fluoroscopy exposure, comparable procedure duration, similar acute success, and no increase in complications. PFO-mediated access may help reduce radiation exposure and avoid septal puncture in selected pediatric patients undergoing left-sided ablation.

**PO.64 | CATHETER ABLATION OF PARAHISIAN ACCESSORY PATHWAYS IN CHILDREN**

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Background:

Parahisian accessory pathways are challenging ablation targets in children due to their proximity to the atrioventricular conduction system. Despite the safety advantages of cryoablation, recurrence remains a concern. This study evaluated outcomes and conduction-related complications of parahisian accessory pathway ablation in children.

Methods:

We retrospectively analyzed 55 children who underwent catheter ablation for parahisian accessory pathways. Patients were classified as having manifest or concealed pathways. Acute success, recurrence, and procedure-related complications were assessed.

Results:

A total of 55 pediatric patients were included. The mean age was 12.4 ± 4.5 years, and the mean body weight was 48.3 ± 19.8 kg. Thirty-three patients were male (60.0%). Manifest accessory pathways were present in 45 patients (81.8%), whereas concealed accessory pathways were present in 10 patients (18.2%).

Cryoablation was performed in 50 patients (90.9%), while radiofrequency ablation was used in 5 patients (9.1%). The mean procedure time was 113.1 ± 46.3 minutes, and the mean fluoroscopy time was 2.5 ± 16.2 seconds. The mean number of lesions was 4.8 ± 2.3 , and the mean total lesion duration was 1445.6 ± 869.5 seconds. Long sheath support was used in 2 patients (3.6%).



Acute procedural success was achieved in 44 patients (80.0%). Complications occurred in 3 patients (5.4%), including transient second-degree atrioventricular block in 2 patients and transient complete atrioventricular block, which occurred twice in 1 patient. No permanent atrioventricular block was observed.

Recurrence occurred in 7 patients (12.7%), all of whom had undergone cryoablation. Acute success was 75.6% in patients with manifest pathways and 100% in those with concealed pathways, although this difference did not reach statistical significance ($p=0.080$). Recurrence rates were 11.1% and 20.0% in the manifest and concealed pathway groups, respectively ($p=0.446$). Complication rates were also comparable between groups (4.4% vs. 10.0%, $p=0.863$).

Energy type, procedure time, number of lesions, and total lesion duration were not significantly associated with complications. However, total lesion duration showed a borderline association with complications ($p=0.054$).

Conclusion:

Catheter ablation of parahisian accessory pathways in children is feasible but remains challenging, with moderate acute success and a relevant recurrence rate. Cryoablation was the predominant energy source, and no permanent atrioventricular block was observed. However, all recurrences occurred after cryoablation, highlighting the delicate balance between safety and efficacy in this high-risk anatomical region. Careful lesion delivery, continuous monitoring of atrioventricular conduction, and individualized energy selection are essential in pediatric parahisian accessory pathway ablation.

**PO.65 | OFF-LABEL CARDIAC MAGNETIC RESONANCE IMAGING IN PEDIATRIC AND CONGENITAL HEART DISEASE PATIENTS WITH NON-CONDITIONAL CARDIAC IMPLANTABLE ELECTRONIC DEVICES: A SINGLE-CENTER SAFETY AND FEASIBILITY STUDY****Laleh Ghaeni¹, Boris Gorodetski², Oktay Tutarel³, Joachim Carsten Will⁴, Felix Berger**

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Background and Aim: Cardiac magnetic resonance imaging (CMR) is indispensable for structural and functional assessment in patients with congenital heart disease (CHD). However, CMR remains formally contraindicated in patients carrying non-conditional cardiac implantable electronic devices (CIEDs), including pacemakers, implantable cardioverter- defibrillators (ICDs), and cardiac resynchronization therapy (CRT) devices. In pediatric and CHD patients, non-conditional CIEDs are frequently implanted, yet evidence supporting off- label CMR in this population remains scarce. We aimed to evaluate the safety, feasibility, and diagnostic image quality of off-label CMR in pediatric and CHD patients with non- conditional CIEDs in a multidisciplinary setting.

Methods: We performed a retrospective single-center analysis of prospectively collected peri- and post-procedural data from 11 consecutive off-label CMR examinations in 10 pediatric and CHD patients with non-conditional CIEDs, performed between March 2025 and May 2026. Device types included pacemakers, ICDs, and CRT devices. Standardized device interrogation was performed immediately before and after each CMR examination at 1.5 Tesla in normal operating mode (specific absorption rate <2 W/kg). Parameters documented included pacing capture thresholds, sensing amplitudes (P- and R-wave), lead impedances, and battery status. A clinically significant change was predefined as a capture-threshold increase >0.5 V, a sensing-amplitude decrease >50%, or a lead-impedance change >50 Ω. Diagnostic image quality was assessed by two independent readers.



Results: Eleven CMR examinations were completed in 10 patients without any adverse events. Epicardial leads were present in 4 patients, abandoned leads in 7 patients (2 patients had both lead types), and 6 patients were pacemaker-dependent. No procedure-related symptoms, arrhythmias, or hemodynamic instability occurred. Device interrogation immediately post-CMR revealed no clinically significant changes in pacing capture thresholds, sensing amplitudes, or lead impedances compared to pre-CMR values. No power-on-reset or unintended mode switch was observed. Diagnostic image quality was good to excellent in all examinations, with full diagnostic yield in every case.

Conclusions: Off-label CMR appears safe and diagnostically valuable in pediatric and CHD patients with non-conditional CIEDs, when performed under a structured multidisciplinary protocol with standardized pre- and post-procedural device interrogation. Our findings support a cautious, individualized risk-benefit assessment and add to the limited evidence in this underrepresented population.

**PO.66 | CRYOABLATION ENERGY USAGE IN SMALL PEDIATRIC PATIENTS FOR THE ABLATION OF RE-ENTRY ARRHYTHMIAS – SINGLE CENTRE EXPERIENCE****Cristina Raimondo¹, Diāna Stoldere^{2,3}, Fabrizio Drago¹**

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2 - Children's university hospital of Latvia, Pediatric cardiology and cardiac surgery clinic, Riga, Latvia

3 – Riga Stradiņš University, Faculty of Medicine, Department of Pediatrics

Background and aim:

Cryoablation is a standard and preferred therapy for pediatric atrioventricular nodal and accessory pathway re-entry arrhythmias, primarily because of its favourable safety profile near the AV node. While its efficacy in adolescents is thoroughly documented, direct clinical evidence for children weighing 26 kg or less is scarce, relying heavily on data generalised from adult studies. For pediatric patients, reduced anatomical size and challenges in maintaining catheter stability can significantly impact lesion quality, overall procedural success, and the likelihood of complications

Methods:

This study was conducted at Bambino Gesù Children's Hospital in Rome and focused on pediatric patients weighing less than 26 kg who underwent cryoablation for re-entry arrhythmias between January 2023 and October 2025. Descriptive statistics were utilised to evaluate demographic and clinical profiles, as well as immediate procedural results and long-term outcomes up to one year post-ablation.

Results:

The study group consisted of 19 patients, male – 57.9 % (n=11) and female 42.1% (n=8), with a median age of 8 years (IQR 2.0) in the male group and 6.5 years (IQR 4.0) in the female group. Median weight in the study group was 24 kg (IQR 2.5). All of them presented with re-entry supraventricular arrhythmia: 57.9%, AVNRT and the rest with AVRT – 42.2%. All of the AVRT patients, selected for this study, presented with right-sided accessory pathways, most localised anteriorly, 75%, respectively. Centre policy is the use of cryoenergy for the ablation procedure of the right atrium.



All procedures included an electrophysiologic study, mapping of the right atrium as a pre-ablation procedure, and reducing the amount of received radiation to near zero. No complications were reported. During the 1-year follow-up period, overall 4 relapses were reported, 3 in the AVNRT group and 1 in the AVRT. Redo ablation was performed in 1 case, the rest continued antiarrhythmic drug therapy.

Conclusion:

Our findings indicate that cryoablation is a safe and effective initial treatment for pediatric patients weighing less than 26 kg, with a low long-term recurrence rate specifically in AVRT cases. Given the small sample size of this study, further large-scale research is warranted to validate these results

No conflict of interests to declare.



PO.67 | SUBCUTANEOUS ICD FOR SUDDEN CARDIAC DEATH PREVENTION IN ANDERSEN-TAWIL SYNDROME TYPE 1 – CHALLENGES DURING 15 YEARS FOLLOW UP OF TWO PATIENTS

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Background and Aim: Andersen-Tawil Syndrome Typ 1 (ATS1) is a rare condition caused by loss-of-function mutations in the KCNJ2 gene resulting in QTc/QUc prolongation with ventricular arrhythmias, periodic paralysis, and typical dysmorphic features. Recent studies revealed a high rate of life-threatening arrhythmic events (LAE).

Methods: We present two cases of a family. The boy presented first at the age of 3 years after recurrent syncope. Surface ECG showed a marked U wave and polymorphic premature ventricular complexes (PVCs) (figure 1). ATS1 was suspected and confirmed by genetic testing. The mother reported recurrent syncope and palpitations since teenage years. Genetic testing at the age of 38 years confirmed ATS1.

Results: Both, mother and child started therapy with flecainide that was for years effective in reducing the burden of PVCs and polymorphic ventricular tachycardias (VT) in Holter recordings. Despite therapy, the mother had a LAE at the age of 42 and was successfully resuscitated. A subcutaneous ICD (sICD) was implanted for secondary prevention of SCD. Few months later she received an inappropriate shock due to double counting of a hemodynamically well tolerated VT (figure 2). One year later a TdP/VT was successfully terminated by 2 sICD shocks (figure 3). Up to date she suffered 3 more inappropriate sICD shocks. At the age of 15 years the boy presented at regular follow up with worsening ventricular arrhythmia burden and a sICD was implanted for primary SCD prevention (figure 4).



He received one inappropriate shock. At the age of 18 years the boy suffered a LAE resulting in cardiac arrest. The analysis of the sICD revealed that underdetection of TdP due to different vectors in the ECG recorded by the sICD might have resulted in a delay of shock delivery. While he received one appropriate shock, only, three inappropriate shocks were delivered as a result of double counting of VTs. Despite prompt resuscitation, he died two weeks later, following aspirational pneumonia, ARDS, and hypoxic brain damage.

Conclusions: These two cases illustrate the limitations of sICD therapy in ATS1, that may result in a substantial number of inappropriate shocks as well as underdetection of LAEs.

Keywords: Andersen-Tawil Syndrome Type 1, life-threatening arrhythmic events, subcutaneous ICD

Figure 1. Surface ECG with a marked U wave and polymorphic premature ventricular complexes (PVCs)

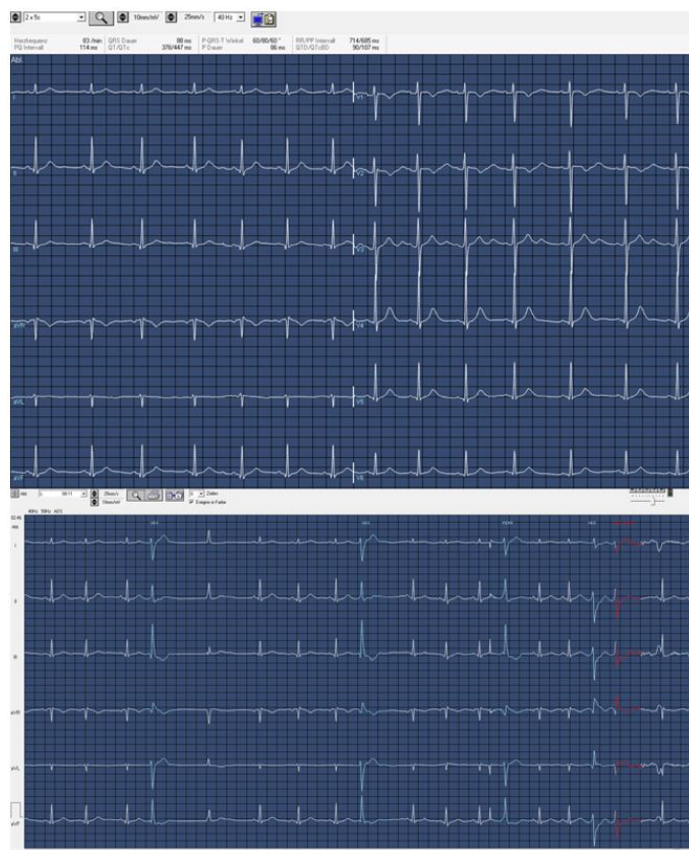




Figure 2. Inappropriate siCD shock due to double counting of a hemodynamically well tolerated VT

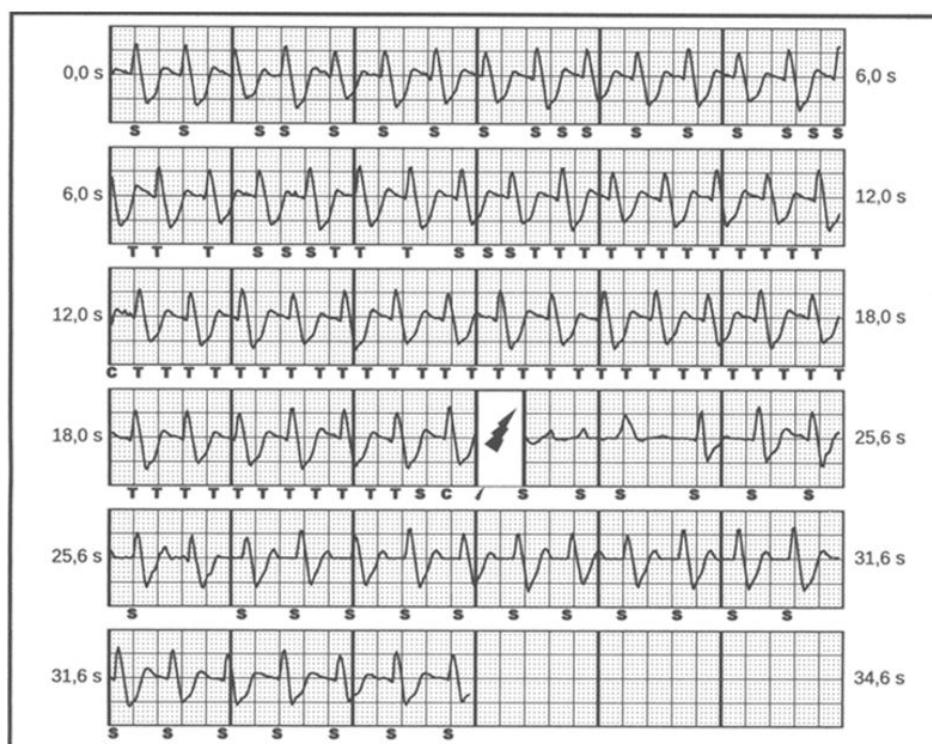




Figure 3. TdP/VT successfully terminated by two sICD shocks

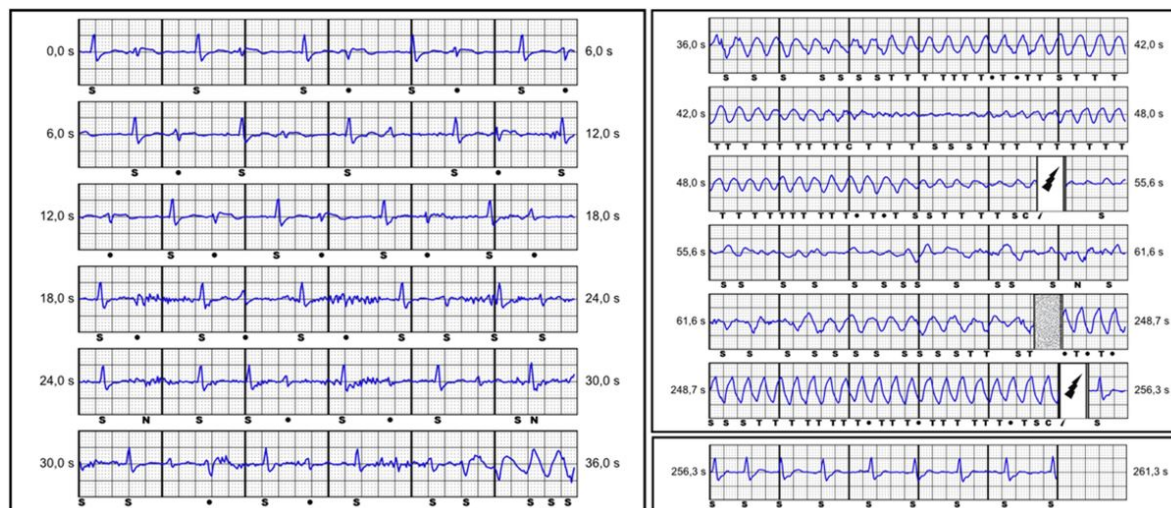
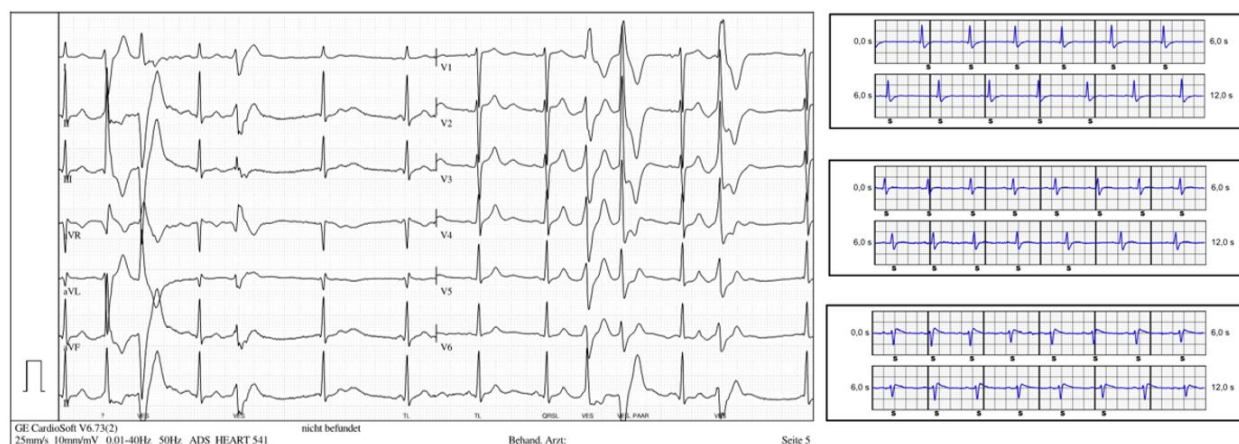
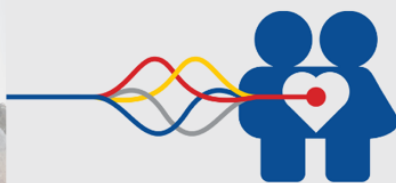


Figure 4. Worsening ventricular arrhythmia burden in surface ECG and well recognized QRS complexes in sICD recordings (primary, secondary and alternative configuration)



Disclosure:

Nothing to declare.

**PO.68 | MEXILETINE TREATMENT FOR HIGH RISK NEONATAL LONG QT SYNDROME TYPE 2 – CASE REPORT****Irena Odri Komazec¹, Miriam Michel¹, Tim Wegerhoff², Marion Pavlic³**

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Background and Aim: Long QT syndrome type 2 (LQT2) is the second most common type of long QT syndrome. It is a rare condition caused by mutations in the KCNH2 gene, resulting in QTc prolongation and a risk of life-threatening arrhythmic events (LAE). Recent research in a patient cohort (16+-14 years of age at diagnosis) showed that adding mexiletine to standard treatment with betablocker could shorten QTc and reduce arrhythmia risk. Evidence of treating newborns is scarce.

Methods: We report a case of a newborn girl that presented prenatally with intermittent bradycardia, growth restriction, and pleural effusions. Postnatal heart rate was normal, but surface ECGs showed a QTc prolongation of over 500 ms and premature ventricular complexes (Figure 1), thus indicating long QT syndrome with a high risk of malignant arrhythmia and sudden cardiac death.

Results: Genetic testing revealed a likely pathogenic mutation in KCNH2 gene. The standard therapy with propranolol was initiated and the dose gradually increased up to 2,4 mg/kg/day, with no relevant effect on the heart rate or QTc, but causing hypotension. Off-label mexiletine therapy was considered as add-on. The polymorphism testing of cytochrome enzyme CYP2D6 showed intermediate metabolizer. Oral drug challenge test with only 2,2 mg/kg of mexiletine was positive, causing at peak 40 ms QTc shortening (to 455 ms), 150 minutes after mexiletine administration (Figure 2). Mexiletine chronic therapy was indicated and the dose was carefully titrated up to 8,6 mg/kg/day, while monitoring for QTc (serial surface ECGs and Holter recordings), drug plasma concentration and oral suspension stability.



The baby girl is now 6 months old, thriving well, the dosage is being frequently adapted to weight gain and so far, there was no evidence of brake-through arrhythmia events or mexiletine side effects.

Conclusions: This case illustrates several challenges in clinical practice:

- recognizing long QT syndrome in a newborn with normal postnatal heart rate despite preceding subtle prenatal findings and in the absence of neonatal ECG screening programme,
- discussing a potentially life-threatening diagnosis, off-label therapy and life style recommendations with parents of an apparently healthy infant,
- quantifying therapy dose-effect balance,
- limited availability of oral mexiletine suspension.

Keywords: Long QT syndrome Type 2, mexiletine, off-label therapy



Figure 1. Surface ECG with a marked QTc prolongation (535 ms) and premature ventricular complexes.

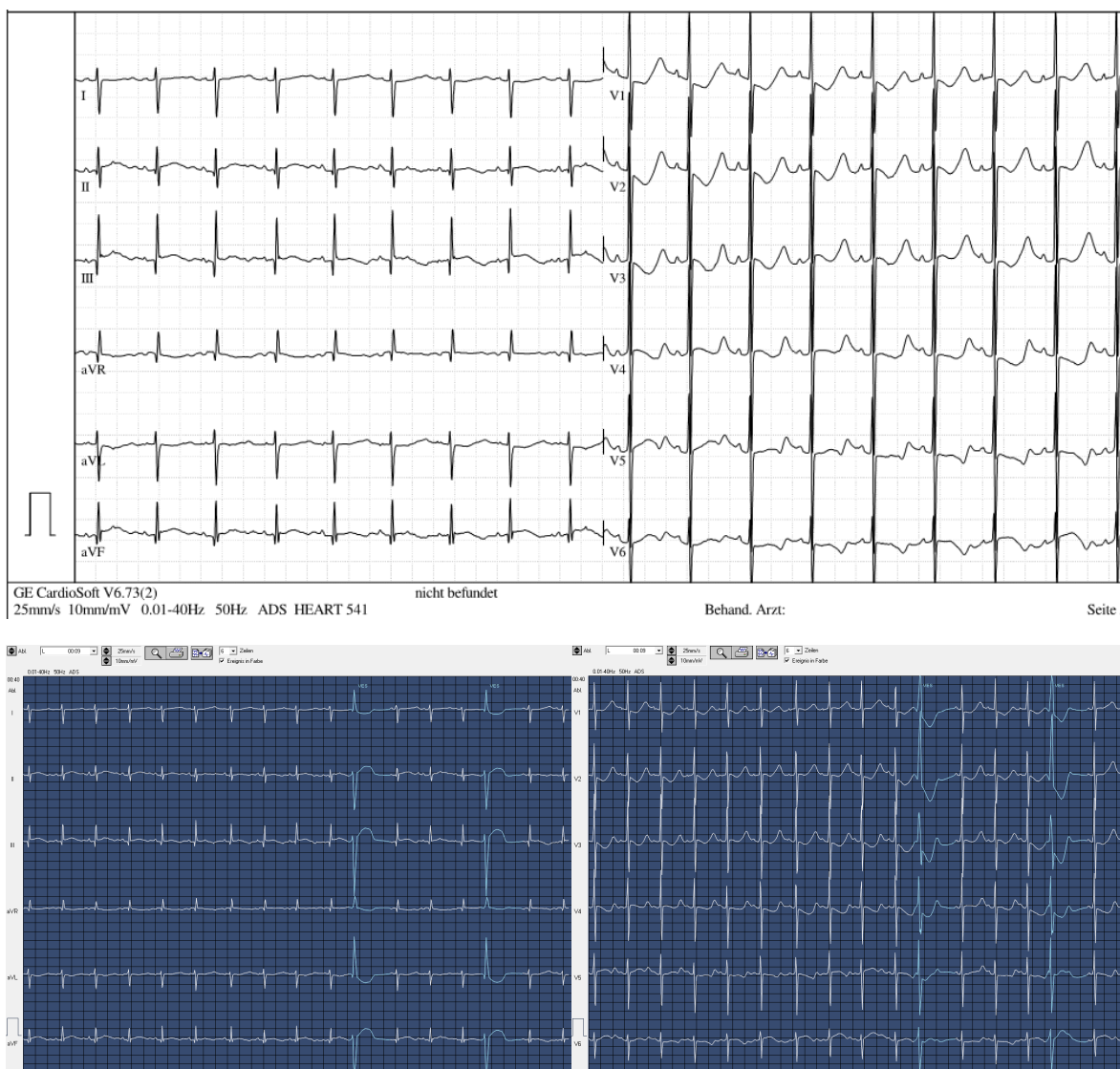




Figure 2. Surface ECG with a 40 ms QTc shortening 150 minutes after oral mexiletine challenge test (2,2 mg/kg)



Disclosure:

Nothing to declare.

**PO.69 | OUT OF HOSPITAL CARDIAC ARREST IN A PEDIATRIC PATIENT WITH WOLFF-PARKINSON-WHITE SYNDROME AND AN ANOMALOUS RIGHT ATRIAL APPENDAGE TO RIGHT VENTRICLE CONNECTION**

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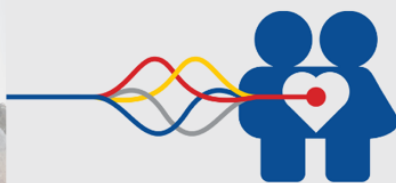
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Background and Aim:

Wolff-Parkinson-White (WPW) syndrome is well-characterized in the general population with occurrence of 1 to 3 people in 1000; however, its association with a right atrial appendage to right ventricle connection is exceedingly rare, posing unique diagnostic and electrophysiological challenges. We present the case of a 9-year-old patient who presented with this rare association and an out-of-hospital cardiac arrest.

Case summary:

A 9-year-old female patient, previously asymptomatic, presented with palpitations and was diagnosed with WPW syndrome, for which she was prescribed flecainide and referred for an electrophysiology study (EPS). Six months following the initial presentation, the patient suffered an out-of-hospital ventricular fibrillation (VF) cardiac arrest, requiring defibrillation twice from an Automated External Defibrillator (AED). Following successful resuscitation, she underwent a comprehensive clinical evaluation, including an electrocardiogram (ECG), echocardiography, cardiac magnetic resonance (CMR) imaging, and an urgent EPS. Review of the AED data following the first shock, revealed rapid, pre-excited atrial fibrillation degenerating into ventricular fibrillation. Multimodal imaging, including CMR and a 3D echo identified two tricuspid valve orifices, effectively creating a two-chambered right ventricle with an anomalous accessory chamber connecting the right atrial appendage to the right ventricle.



An EPS performed under general anaesthesia demonstrated a high-risk accessory pathway with both antegrade and retrograde effective refractory periods of less than 200 milliseconds (without isoprenaline). Activation mapping localized the earliest signals to the lateral tricuspid annulus, with corresponding early signals identified within the accessory chamber on either side of the muscular septation. Radiofrequency ablation (RFA) delivered within the accessory chamber successfully abolished pathway conduction. A one-month follow-up ECG confirmed the absence of pre-excitation. Multidisciplinary team discussion regarding secondary prevention considered a subcutaneous implantable cardioverter-defibrillator (S-ICD). However, because the rapidly conducting pathway was identified as the primary aetiology of the arrest, the consensus opinion was for an implantable loop recorder (ILR) for continuous monitoring, pending further consideration by the family.

Discussion: This case highlights that complex structural anomalies, such as an accessory right-sided chamber or fistulous connection, can harbour extremely fast-conducting pathways capable of inducing sudden cardiac arrest. Careful multimodal imaging and precise endocardial mapping within these anomalous structures are essential for delivering successful, targeted ablation.

Disclosure

No disclosures.

Figure 1: AED Download

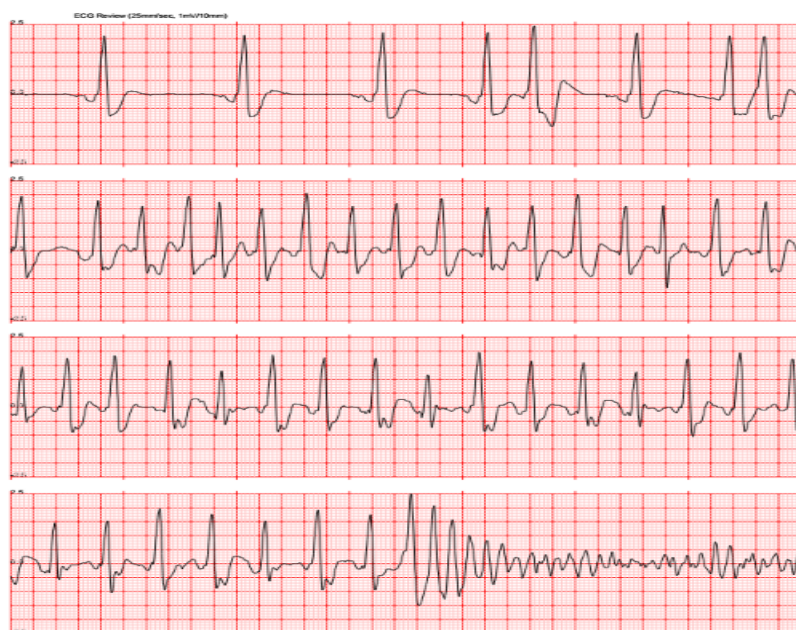
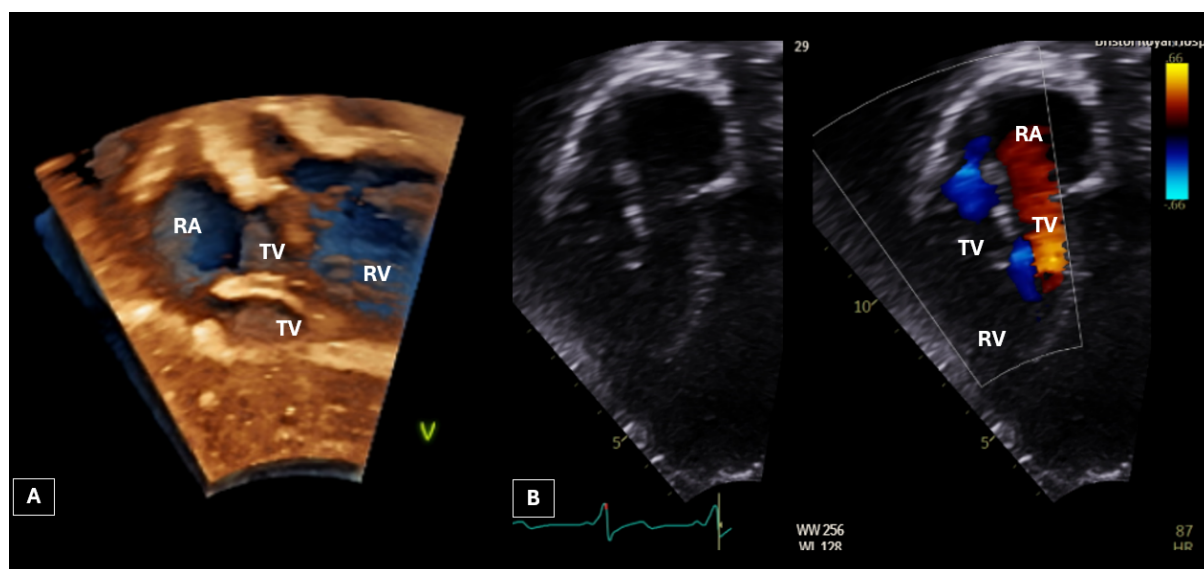
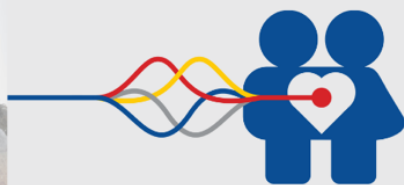




Figure 2A: 3D echo showing the duplication of the TV. Figure 2B. 2D image showing laminar flow across the larger TV on the left and some flow across the orifice connecting to the TV on the right. Both TV connect to the RV, although the flow across the right sided TV is minimal.





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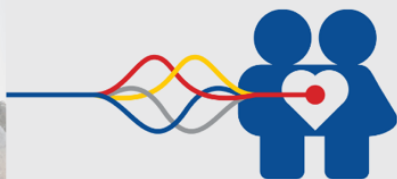
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**PO.70 | INITIAL CLINICAL EXPERIENCE AND FEASIBILITY OF THE AURORA
EXTRAVASCULAR IMPLANTABLE CARDIOVERTER-DEFIBRILLATOR (EV-ICD™ SureScan™)
IN A PEDIATRIC COHORT IN BRISTOL UNITED KINGDOM**

Rabeea Siddiqui¹, Palash Barman², Georgia Spentzou¹

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² Bristol Heart Institute,

Background

Implantable cardioverter-defibrillators (ICDs) in paediatric patients present unique anatomical and clinical challenges. While subcutaneous ICDs mitigate intravascular shock coil risks, the Medtronic extrinsic Aurora EV-ICD™ SureScan™ features a reduced generator profile and sub-sternal shock coil routing. This device also offers anti-tachycardia pacing (ATP) and pause prevention pacing in case of significant pauses where the device provides temporary bradycardic pacing support.

Methods

Five paediatric patients were evaluated for Aurora EV-ICD™ SureScan™ implantation. The clinical cohort comprised:

- A 16-year-old (45 kg, 161 cm) with refractory catecholaminergic polymorphic ventricular tachycardia despite optimal medical therapy and sympathectomy.
- A 12-year-old (39 kg, 161 cm) who survived a ventricular fibrillation (VF) arrest.
- A 6-year-old (20 kg, 121 cm) with long QT syndrome (LQTS), an out-of-hospital arrest, and a prior endocardial ICD.
- A 12-year-old (41 kg, 162 cm) with Marfan syndrome, mitral valve prolapse, and Holter-documented ventricular tachycardia.
- A 16-year-old (65 kg, 167 cm) diagnosed with LQTS following an out-of-hospital arrest.

Preoperative cross-sectional imaging was utilized for detailed anatomical assessment across the cohort. Furthermore, the novel nature of this device was thoroughly discussed during shared decision-making with the patients and their parents/guardians.



Results

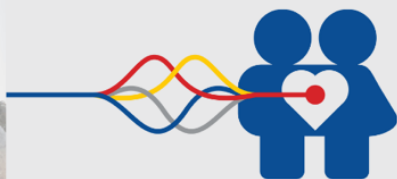
All procedures were conducted under general anaesthesia in the cardiac catheterization lab. The standardized Aurora EV-ICD™ SureScan™ implantations were performed successfully without any procedural complications. The Epsilon EV™ SureScan™ (EV2401) lead was deployed using a proprietary tunnelling tool and guided by fluoroscopy, achieving excellent sensing and pacing parameters. The Aurora EV-ICD™ SureScan™ (DVEA3E4) pulse generator was then positioned in the left lateral chest wall. Intraoperative defibrillation testing successfully terminated induced VF with a single shock with 30J shock except in one of the cases where VF was not inducible. Post-operative device parameters were good. No post procedural complications were observed, and patients were discharged within 24 hours.

Conclusions

This preliminary case series demonstrates that Aurora EV-ICD™ SureScan™ implantation can be performed uneventfully in paediatric patients, including those with a small body habitus. Due to its recent introduction and lack of official paediatric approval, longitudinal performance data remain inherently limited. Consequently, meticulous patient selection via advanced imaging and comprehensive shared decision-making regarding long-term uncertainties are imperative. As with most emerging technologies, we anticipate an expanding clinical interest in the integration of this device for paediatric arrhythmia management.

Disclosure

None



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**PO.71 | NAVIGATING BAFFLE COMPLICATIONS: A STRATEGY FOR ATRIAL PACING IN DEXTRO-TRANSPOSITION OF THE GREAT ARTERIES WITH ATRIAL SWITCH****Elliot Griffith¹, Hiroko Beck², Mohammad Al-Mousily¹**¹ Congenital Heart Center, University of Florida, United States² Department of Cardiology, University of Florida, United States

Background and Aim: Dextro-transposition of the great arteries is a congenital cardiac defect characterized by atrioventricular concordance and ventriculoarterial discordance. Following atrial switch, up to 65% of these patients have some degree of bradyarrhythmia or conduction system disease in adulthood and sinus node dysfunction is a common outcome. Leadless pacemakers offer a promising alternative for patients with venous access restrictions or elevated risk for thromboembolism and infection.

Methods: A 47-year-old male with d-TGA status post Mustard operation in infancy, longstanding history of sinus bradycardia with intermittent junctional rhythm, and known SVC baffle stenosis was admitted after cardioversion for atrial flutter with resultant sinus arrest. He was planned to undergo transvenous pacemaker implantation. Cath evaluation revealed an inferior baffle leak and superior baffle obstruction. He instead underwent successful leadless pacemaker implantation within the left atrium utilizing guidance from fluoroscopy and intracardiac echo.

Results: This case highlights the common barriers to standard transvenous pacemakers in patients with d-TGA and atrial switch procedure including superior baffle obstruction as well as baffle leaks. It also highlights the expanding role of leadless pacing technology in overcoming these complex anatomic challenges. Sinus node dysfunction is a frequent issue in this population, with many requiring pacemaker therapy in adulthood. In this case, the presence of a completely occluded SVC baffle and an inferior baffle leak presented significant limitations for traditional pacing options.

Conclusions: Atrial leadless pacemaker implantation offers a promising solution in adult patients with d-TGA post-Mustard repair who develop sinus node dysfunction and have unfavorable anatomy for transvenous pacing. This case highlights the common barriers to standard transvenous pacemakers in patients with d-TGA and atrial switch procedure including superior baffle obstruction and baffle leaks. It also highlights the expanding role of leadless pacing technology in overcoming complex anatomic challenges.

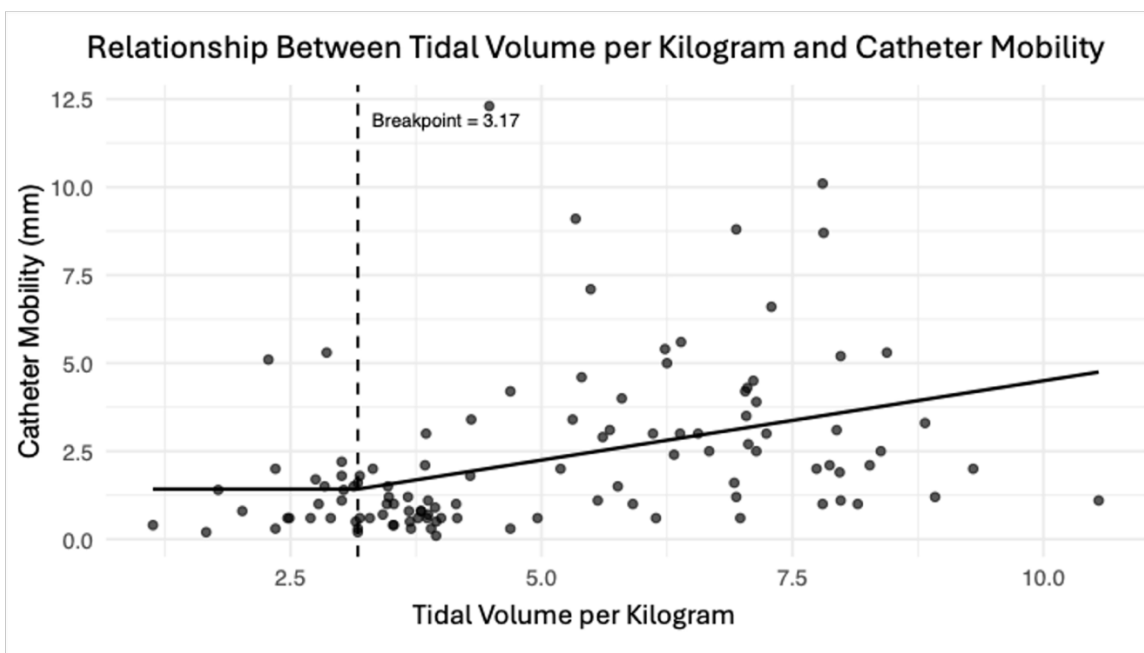
**PO.72 | FINDING THE SWEET SPOT: DEFINING AN OPTIMAL TIDAL VOLUME FOR CATHETER STABILITY IN PEDIATRIC EP ABLATION****Elliot Griffith¹, Nikolaus Gravenstein², Mohammad Al-Mousily³**¹ Congenital Heart Center, University of Florida, United States² Anesthesia, University of Florida, United States³ Congenital Heart Center, University of Florida. United States

Background and Aim: Catheter stability is crucial during pediatric ablations, and respiratory motion is a major source of displacement. High-frequency, low-tidal-volume ventilation (HF-LTVV) can reduce motion, but the optimal tidal volume per kilogram (TV/kg) is not well defined. This study aimed to compare catheter mobility during conventional ventilation versus HF-LTVV and determine whether a threshold TV/kg exists below which further reductions offer little added benefit.

Methods: In a prospective paired design, 58 pediatric patients undergoing EP ablation received conventional ventilation (mean TV/kg 6.7 ± 1.4) and HF-LTVV (3.3 ± 0.7). Catheter tip motion was measured with the Abbott EnSite mapping system. Mobility differences were evaluated using paired t-testing. Threshold regression was performed using a hinge (broken-stick) model to identify the non-linear relationship between ventilation and catheter displacement. The model sought where the derivative of the catheter mobility relative to tidal volume per kilogram approaches zero.

Results: HF-LTVV resulted in a 71.3% reduction in mean catheter mobility compared to conventional settings (1.03 ± 0.84 mm vs 3.59 ± 2.55 mm; $p = 0.03$). Threshold regression modeling identified a breakpoint at 3.17 mL/kg (95% CI 2.02–3.70 mL/kg), below which further reductions in tidal volume yielded minimal additional reductions in mobility.

Conclusions: Using the EnSite mapping system, HF-LTVV was shown to significantly reduce catheter mobility in pediatric ablations. A threshold of 3.17 mL/kg may optimize the trade-off between respiratory motion suppression and physiologic ventilation and may serve as a practical standard for procedural ventilation in this population.



**PO.73 | CARDIONEUROMODULATION FOR RECURRENT MALIGNANT NEUROCARDIOGENIC SYNCOPE IN PAEDIATRIC PATIENTS: RESULTS FROM A TERTIARY CENTRE.**

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Background and aim: Malignant neurocardiogenic syncope (MNS) is defined as recurrent syncope with minimal prodromes associated with very prolonged asystole during head-up tilt table test (HUTT). Although cardioneuromodulation (CNM) is an emerging therapy for cardioinhibitory reflex syncope and other functional bradyarrhythmias, and is increasingly considered an alternative to cardiac pacing in selected adult patients, evidence for its use in paediatric populations remains limited.

This study aims to describe the procedural characteristics and clinical outcomes of CNM in paediatric patients with MNS and to identify the clinical profile of those most likely to benefit from CNM.

Methods: This is a prospective observational single-arm cohort. All paediatric patients (from 11 to 16 years) with recurrent traumatic syncope and HUTT-documented asystole (≥ 15 s) undergoing CNM from May 2025 to May 2026 were enrolled. Baseline clinical features, HUTT findings, procedural data, acute endpoints, complications, and follow-up outcomes were reviewed. All patients underwent CNM using a right atrial-only approach, targeting right-sided ganglionated plexi using anatomical and electrogram guidance, in line with contemporary consensus guidance.

Results: Eight patients (60% female; mean age 14 ± 2 years) with a high burden of traumatic reflex syncope (≥ 1 episode/month; 3 ± 2 episodes/month) were enrolled in this study. All procedures were completed without major complications. After ablation, an implantable loop recorder (ILR) was implanted in all patients for a continuous rhythm surveillance. During a short-term follow-up (6 ± 2 months), all patients experienced marked reduction in syncope burden with no further traumatic recurrences; ILR monitoring showed no prolonged asystole.



Conclusions: In this small paediatric cohort with MNS, CNM was safe, feasible, and associated with favourable short-term clinical and rhythm outcomes. Larger paediatric studies with standardized selection criteria and longer follow-up are needed to define efficacy and durability.



PO.74 | CATHETER ABLATION IN CHILDREN WEIGHING LESS THAN 20 KG: PROCEDURAL OUTCOMES AND COMPARISON OF RADIOFREQUENCY VERSUS CRYOABLATION

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Background:

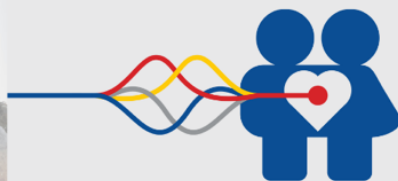
Catheter ablation in children weighing less than 20 kg remains challenging because of small cardiac structures, limited vascular access, and the risk of conduction system injury. Although both radiofrequency ablation and cryoablation are used in this vulnerable population, comparative data on procedural characteristics, efficacy, and safety remain limited. This study aimed to evaluate the safety and efficacy of catheter ablation in children weighing less than 20 kg and to compare procedural outcomes between radiofrequency ablation and cryoablation.

Methods:

We retrospectively analyzed 96 consecutive children weighing less than 20 kg who underwent catheter ablation for supraventricular tachyarrhythmias. Baseline demographics, arrhythmia mechanisms, ablation modality, procedural parameters, acute procedural success, recurrence, and complications were evaluated. Patients were divided into two groups based on energy source: radiofrequency ablation and cryoablation.

Results:

A total of 96 children were included. The median age was 60.5 months and the median body weight was 17.0 kg. The most common arrhythmia mechanism was manifest accessory pathway in 53 patients (55.2%), followed by typical atrioventricular nodal reentrant tachycardia in 16 patients (16.7%), concealed accessory pathway in 14 patients (14.6%), focal atrial tachycardia in 9 patients (9.4%), atypical atrioventricular nodal reentrant tachycardia in 3 patients (3.1%), and atrial flutter in 1 patient (1.0%). Arrhythmia substrates were right-sided in 67 patients (69.8%), left-sided in 25 patients (26.0%), and multiple in 4 patients (4.2%).



Radiofrequency ablation was performed in 74 patients (77.1%), and cryoablation in 22 (22.9%). Overall, acute procedural success was achieved in 93 patients (96.9%). Recurrence occurred in 6 patients (6.3%), and complications in 5 (5.2%).

Recurrences were observed after typical AVNRT slow pathway modification, right posteroseptal manifest accessory pathway ablation, right anterosseptal focal atrial tachycardia cryoablation, right anterosseptal manifest accessory pathway cryoablation, right posterior–posteroseptal manifest accessory pathway radiofrequency ablation, and right posteroseptal manifest accessory pathway cryoablation.

When radiofrequency ablation and cryoablation were compared, patients in the cryoablation group had significantly higher body weight and body surface area. Procedure time was significantly longer in the cryoablation group compared with the radiofrequency group [90 min vs. 60 min, $p < 0.001$]. The number of lesions was also higher in the cryoablation group [5 vs. 3, $p < 0.001$], and total lesion duration was longer [1380 s vs. 105 s, $p < 0.001$]. However, total lesion duration should be interpreted with caution due to inherent differences among energy delivery modalities.

Acute procedural success was 98.6% in the radiofrequency group and 90.9% in the cryoablation group, with no statistically significant difference between groups ($p = 0.130$). Recurrence occurred in 3 patients (4.1%) in the radiofrequency group and in 3 patients (13.6%) in the cryoablation group, which was also not statistically significant ($p = 0.131$). Complications were significantly more frequent in the cryoablation group compared with the radiofrequency group [18.2% vs. 1.4%, $p = 0.009$].

Overall, complications included right bundle branch block in 2 patients, transient complete atrioventricular block in 2 patients, and second-degree atrioventricular block in 1 patient. One patient with transient complete atrioventricular block required temporary pacing, and atrioventricular conduction recovered within 1 hour.

Conclusion:

Catheter ablation in children weighing less than 20 kg was associated with a high acute success rate and an acceptable recurrence rate. Radiofrequency ablation was associated with shorter procedure time, fewer lesions, and fewer complications compared with cryoablation, while acute success and recurrence rates were comparable between groups. Recurrences were mainly observed in septal and right-sided substrates, including posteroseptal and anterosseptal accessory pathways, AVNRT slow pathway modification, and anterosseptal focal atrial tachycardia. Although complications were infrequent overall, conduction disturbances including right bundle branch block and transient atrioventricular block were observed, emphasizing the need for careful energy selection, precise catheter positioning, and close intraprocedural monitoring in small children.

**PO.75 | NONO-associated disease with Long QTc****Maria Elena Gutierrez¹, Noboyuki Ikeda², Karine Guerrier¹**

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The NONO-associated disease is a rare, X-linked syndromic disorder. The presentation includes developmental delay, corpus callosum anomalies, left ventricular non-compaction cardiomyopathy (LVNC) and other congenital heart defects. There is no report of association with inherited long QT syndromes (LQTS) or a finding of a prolonged QTc in these patients. We present a case describing an infant with severe LVNC and prolonged QTc >500ms. Various genetic tests were performed, identifying a pathogenic NONO mutation. No genetic change known to be associated with LQTS was found.

This is a patient born at term readmitted to NICU at 3 days old with hypothermia. Evaluation at his local hospital included an echocardiogram showing severe LVNC with normal biventricular function. Other abnormal exam features included micrognathia, small ears, high arched palate and hypotonia. He had a negative chromosomal microarray and a newborn screen positive for sickle cell trait. At 8 weeks old his electrocardiogram (ECG) was reported as normal. At 3 months of age, he presented with severe LV dysfunction, at which time his ECG showed sinus rhythm and an automated QTc of 420ms. He was managed in a cardiac ICU with reported clinical improvement on milrinone. He was then referred to our institution for specialized cardiomyopathy management. On initial visit at our institution, his LV ejection fraction (LVEF) was normal and his ECG showed a QTc of 501ms. He was not on any known QTc prolonging medications, the list which included enalapril, aspirin, furosemide, spironolactone. A Holter was notable for prolonged QT throughout the study. A subsequent ECG again demonstrated a prolonged QTc of 483ms. At the first Electrophysiology clinic visit, QTc was 515ms. Medications were again reviewed, without significant additions. LVEF at this time was normal. Nadolol was initiated at 1mg/kg daily. Patient remained clinically asymptomatic. Known family history was unremarkable. There was no significant electrolyte derangement on outpatient testing. Genetic testing was sent for arrhythmia and cardiomyopathy panel and returned negative. Genetics consultation and whole genome sequencing was then performed, revealing a NONO pathogenic variant. To our knowledge, this is the first described association of the NONO-associated disease with prolonged QTc.

**PO.76 | CASE REPORT OF A NOVEL ASSOCIATION: CONGENITAL AV BLOCK AND PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM****Ezgi Mollaoglu¹, Ridvan Dryana², Chinara Asadova¹, Matthias Gorenflo¹**

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ABSTRACT**Background and Aim:**

Congenital complete atrioventricular (AV) block is a disorder that is mainly associated with maternal Anti-Ro and Anti-La antibodies or congenital heart defects, mainly congenitally corrected transposition of great arteries or endocardial cushion defects.

Pulmonary atresia with intact ventricular septum (PA/IVS) is a rare defect caused by complete obstruction of the right ventricular outflow tract in the setting of an intact interventricular septum.

While rhythm disturbances are relatively typical in association with complex congenital heart disorders, permanent third-degree AV block in conjunction with PA/IVS is extremely unusual.

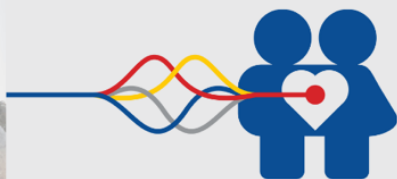
Primary congenital glaucoma is characterized by the enlargement of the neonatal globe due to abnormal development from increased intra-ocular pressure. The association of buphthalmos with PA/IVS and neonatal complete AV block has not been unequivocally demonstrated in the literature.

Methods:

We report a patient with all three congenital abnormalities. To our knowledge, this constellation of findings has not been previously reported and may represent a previously unrecognized developmental association.

Results:

We report a female neonate with PA-IVS and AVBIII°, born early term, weight and height appropriate for gestational age, with cyanosis and a transcutaneous saturation of 75% and heart rate of 50bpm. Alprostadil was initiated with 12,5 ng/kg/min in the delivery room.



Facial dysmorphism with low set ears and flat nasal bridge were noted on the postnatal physical examination. The medical histories of parents and the older siblings are unremarkable.

The postnatal echocardiography confirmed the diagnosis of PA-IVS with hypoplastic right ventricle, hypertrophic and dilated left ventricle.

Following Alprostadil infusion, as the bradycardia and AV Block persisted, the patient received a permanent pacemaker implantation at postnatal day one and a transcatheter palliation with atrial septostomy and pulmonary arterial balloon dilatation at second postnatal day. At the end of the first postnatal week the patient underwent a 3,5mm modified Blalock–Taussig shunt placement.

At the age of 3 months the patient showed signs of corneal clouding and ocular enlargement. Ophthalmological consultation revealed primary infantile glaucoma and the patient was operated with bilateral trabeculotomy.

Conclusions:

The coexistence of PA/IVS, congenital third-degree AV block, and congenital buphthalmos appears to represent a previously unreported combination of congenital anomalies



**PO.77 | PACE MAPPING-GUIDED CATHETER ABLATION FOR PREMATURE VENTRICULAR COMPLEXES IN CHILDREN: A SINGLE-CENTRE EXPERIENCE WITH MID-TERM FOLLOW-UP****Umut Berk Mercan¹, Ömer Nazım Gülçek¹, İlker Ertuğrul¹, Tevfik Karagöz¹**¹ Pediatric Cardiology, Hacettepe University Faculty of Medicine, Ankara, Türkiye

Background: Premature ventricular complexes (PVCs) are common in children; however, frequent PVCs may cause symptoms, ventricular dysfunction, high ectopic burden, or non-sustained ventricular tachycardia requiring intervention. Catheter ablation is established in selected children. Localization may be challenging under general anesthesia because ectopy may diminish, limiting activation mapping. Pace mapping may facilitate localization by matching paced QRS with clinical PVC morphology, particularly for reproducible RVOT ectopy.

Methods: We retrospectively reviewed 16 pediatric patients undergoing catheter ablation for clinically significant PVCs at our center. Patients with high PVC burden, non-sustained ventricular tachycardia, and/or significant symptoms were considered for ablation. Demographic, echocardiographic, procedural, and follow-up data were collected. Three-dimensional mapping was performed using the Abbott EnSite Precision™ system, and pace mapping was performed in all patients. Radiofrequency ablation was delivered with an irrigated contact force-sensing catheter. Acute success was defined as absence of PVC after recovery from general anesthesia. Pace-map concordance and post-ablation PVC burden were analyzed.

Results: Mean age was 12.9 ± 3.7 years and mean body weight was 48.8 ± 17.4 kg. Structural heart disease was present in 3/16 patients (18.8%), and pre-ablation non-sustained ventricular tachycardia in 8/16 (50.0%). Mean pre-ablation PVC burden was $24.0 \pm 14.2\%$. Ablation sites were RVOT in 12 patients (75.0%), moderator band in 3 (18.8%), and para-Hisian tricuspid annulus in 1 (6.3%). Acute success was achieved in 15/16 patients (93.8%), without complications. Among non-recurrent patients (n=13), mean pace-map concordance was $93.8 \pm 2.8\%$, median 94.0% (interquartile range, 92.0–95.0%). During 31.8 ± 24.0 months of follow-up, recurrence occurred in 3/16 patients (18.8%); all underwent repeat ablation.



On repeat mapping, recurrence arose from the same region in one patient and a different site in two; foci were RVOT, para-Hisian region, and aortic cusp. Repeat procedures were performed without general anesthesia to preserve ectopy. Post-procedural ventricular tachycardia occurred in 2/16 patients (12.5%), both with recurrence. Excluding recurrent cases, PVC burden decreased from $22.4 \pm 14.8\%$ to $1.3 \pm 3.2\%$ ($p < 0.001$).

Conclusion: Pace mapping-guided ablation was safe and effective in children. High pace-map concordance suggests that pace mapping can identify PVC origin and guide ablation when anesthesia suppresses ectopy.



PO.78 | EVALUATION OF PEDIATRIC PATIENTS WITH WOLFF-PARKINSON-WHITE (WPW) DIAGNOSIS WITH CARDIOPULMONARY EXERCISE TESTING (CPET) BEFORE AND AFTER ABLATION

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Background and Aim

Wolf-Parkinson-White (WPW) syndrome is a common type of arrhythmia in children. An accessory pathway (AP) may lead to dyssynchrony in ventricular contraction. Dyssynchrony may lead to evident or subclinical decrease in myocardial performance. The present study aims to investigate the effects of these APs on subclinical myocardial performance, particularly in the absence of overt clinical manifestations of heart failure.

Methods

This is a single-center observational study including pediatric patients aged between 8 and 18 years who underwent catheter ablation for WPW syndrome. Cardiopulmonary exercise test (CPET) was performed prior to the ablation procedure and repeated at least three months post-ablation, during routine follow-up.

Results

A total of 25 individuals with the median age of 11,2 (8-17) years, underwent ablation for WPW evaluated with CPET. Echocardiographic evaluation with m mode of these patients showed that left ventricular function and ejection fraction were within normal ranges. Seven patients were on antiarrhythmic medication prior to the procedure, but these medications were discontinued before the CPET evaluation. The mean QRS duration prior to ablation was 94.4 ms (range: 80–130 ms). Posteroseptal (32%) were the most prevalent accessory pathway localization. The VO₂max of the patients before ablation was measured at 24,3 ml/min/kg, and after ablation, it was 26,4 ml/min/kg (p:0,018). The maximum heart rate of the patients was 177 beats/min before ablation and 186 beats/min after ablation (p:0,002). The maximum METs before ablation was 6,4 and after ablation, it was 7,5 (p:0,001).



When comparing the pre- and post-ablation results, a statistically significant increase was observed in VO₂max, maximum heart rate, and METs values. The endocardial peak global longitudinal strain in the apical four-chamber view was 19,7% before ablation and remained 19,15% after the procedure. In the comparison of global longitudinal strain values, three patients in the Wolff–Parkinson–White group were excluded from the analysis due to missing data, and no significant difference was detected (p:0,935).

Conclusions

CPET will be a valuable indicator in the future for the diagnosis, treatment, and follow-up process of WPW due to its detailed assessment of exercise capacity through metabolic measurements.

Table 1: Comparison of CPET results before and after ablation of the WPW cohort

	Before (n=25)	After (n=25)	P
	Median (IQR)	Median (IQR)	
Weight (kg)	46 (23)	46 (22)	0,001
Time (s)	966 (139)	1011 (154)	0,005
AT VO ₂ (ml/min)	723 (508)	671 (744)	0,201
VO ₂ max (ml/min)	942 (708)	1160 (554)	0,003
AT VO ₂ /kg (ml/min/kg)	14,9 (8,4)	16,3 (6,7)	0,493
VO ₂ max (ml/kg/min)	24,3 (8,2)	26,4 (10,8)	0,018
METs max	6,4 (2,6)	7,5 (3)	0,001
RQ max	1,15 (0,13)	1,18 (0,17)	0,028
OUES (ml/min/l/min)	1391 (929)	1382 (981)	0,211
VE max (l/min)	36,5 (26,5)	41,5 (24,2)	<0,001
VT max (l)	0,918 (0,682)	1,108 (0,569)	0,001
HR Max (beat/min)	177 (17)	186 (9)	0,002
HR Max (%)	86 (8)	90 (5)	0,002
VO ₂ /HR max (ml/beat)	5,7 (3,3)	6,1 (3,2)	0,023

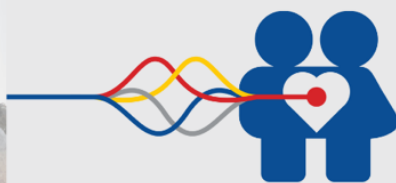
IQR- Interquartile range; VO₂ max- oxygen consumption at the peak of exercise; RQ-respiratory exchange ratio; VE- minute ventilation; VO₂-oxygen output; AT- anaerobic threshold; HR- heart rate; METs-Metabolic Equivalent of Task; OUES-oxygen uptake efficiency slope; s-second; kg-kilogram; l-liter; ml-milliliter; min-minute; max-maximum; n-count; p-value; Wilcoxon Signed Ranks Test, *result with p value < 0.05



Table 2. Comparison of echocardiography results of the WPW cohort

	Before (n=22)	After (n=22)	P
	Median (IQR)	Median (IQR)	
Gls_EP_A4C(%)	19,7 (6)	19,15 (5,93)	0,935
Gls_EP_A2C(%)	18,9 (7,12)	19,75 (5,8)	0,436
Gls_EP_A3C(%)	20,4 (5,98)	18,9 (4,32)	1,000
Gls_EP_Avg(%)	20,35 (5,92)	19,35 (4,98)	0,697
EF (%)	70,5 (9,75)	71 (7,75)	0,169

Gls_EP_A4C: Global longitudinal strain endocardial peak apical 4 chamber view; Gls_EP_A2C: : Global longitudinal strain endocardial peak apical 2 chamber view; Gls_EP_A3C Global longitudinal strain endocardial peak apical 3 chamber view; Gls_EP_Avg: Global longitudinal strain endocardial peak average; EF: ejection fraction IQR: Interquartile range, n — count; p — p value; Wilcoxon Signed Ranks Test, *results with p value < 0.05

**PO.79 | SCN5A VARIANTS: FROM DISEASE-CAUSING MUTATIONS TO SECONDARY FINDINGS IN A SINGLE-CENTER PEDIATRIC COHORT**

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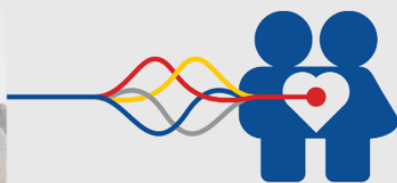
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Background: *SCN5A* encodes the sodium-channel α -subunit Nav1.5 and is associated with inherited arrhythmia and cardiomyopathy phenotypes. Interpretation is particularly challenging in children and in variants detected incidentally as secondary findings. This study aimed to describe the clinical and genetic spectrum of *SCN5A* variants in a pediatric cohort.

Methods: We retrospectively reviewed 22 patients carrying *SCN5A* variants classified as pathogenic, likely pathogenic, or variants of uncertain significance according to ACMG criteria. Patients were grouped by testing indication: cardiac manifestations or secondary findings identified during exome sequencing for non-cardiac indications. Clinical, electrocardiographic, imaging, electrophysiological, inheritance, and genotype–phenotype data were analyzed.

Results: Fourteen distinct *SCN5A* variants were identified. Six patients carried pathogenic or likely pathogenic variants, including p.(Val411Met) in two patients and p.(Met1766Ile), p.(Arg893Cys), p.(Arg451GlyfsTer19), and p.(Gln1506_Pro1508del) in one patient each; the remaining 16 carried variants of uncertain significance. Most variants were missense, although frameshift, splice-region, and in-frame deletion variants were also observed. Patients with pathogenic or likely pathogenic variants had earlier symptom onset and diagnosis and a more homogeneous phenotype, predominantly long QT syndrome type 3 (n=4) and Brugada syndrome (n=2), with higher frequencies of syncope or cardiac arrest and implantable cardioverter-defibrillator implantation. The variant-of-uncertain-significance group was more heterogeneous. Five VUS carriers had cardiac manifestations. Two related patients carrying the same heterozygous c.2443G>T, p.(Val815Phe) variant had Brugada phenotype. Among VUS carriers who underwent implantable cardioverter-defibrillator implantation, indications were arrhythmogenic right ventricular dysplasia/cardiomyopathy-like phenotype in one patient and Brugada phenotype with ventricular tachycardia in the others.



The remaining symptomatic VUS carriers had ventricular arrhythmia-related findings. Long QT syndrome type 3 and Brugada syndrome were the most frequent phenotypes overall, each present in 18.2% of patients. Variants clustered in functionally important Nav1.5 regions. p.(Val411Met) was associated with marked QT prolongation and mexiletine responsiveness. All secondary-finding cases were VUS and largely asymptomatic at baseline.

Conclusion: Pediatric *SCN5A* variants show broad phenotypic variability, from severe arrhythmia syndromes to incidental secondary findings. Pathogenic or likely pathogenic variants were associated with earlier onset and greater severity, whereas secondary-finding variants and other VUS require cautious interpretation. Detailed phenotyping, segregation analysis, and structured follow-up remain essential.

**PO.80 | GENETIC DIAGNOSIS AS A TURNING POINT IN THE MANAGEMENT OF TECRL-RELATED ARRHYTHMOGENIC SYNDROME: A FIVE-PATIENT CASE SERIES**

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Background:

Pathogenic variants in the *TECRL* (trans-2,3-enoyl-CoA reductase-like) gene cause a rare inherited arrhythmogenic syndrome with overlapping features of Long QT Syndrome (LQTS) and Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT). The condition is associated with a high risk of sudden cardiac death (SCD), but diagnosis is often delayed due to phenotypic variability.

Case Series:

We identified five patients from three unrelated families carrying homozygous *TECRL* variants. Two siblings carried the *TECRL* c.730+1G>C variant and were initially diagnosed with LQTS following the sudden cardiac death of a sibling. These patients displayed QTc intervals ranging from 0.40 to 0.44 s. One experienced recurrent ICD shocks despite beta-blockers and multiple left cardiac sympathetic denervations; following genetic diagnosis, clinical stabilization was achieved with the addition of flecainide, while the other remains asymptomatic on propranolol. Two additional siblings carried a homozygous *TECRL* c.551+5G>C variant; they exhibited normal QTc intervals. One survived a ventricular fibrillation arrest and the other was identified through family screening after documenting exercise-induced ventricular arrhythmias. Treatment for these patients was optimized with nadolol, flecainide, and sympathetic denervation. The fifth patient, unrelated to the other families, presented with exercise-induced syncope and a family history of exertional sudden cardiac death. Whole-exome sequencing identified a homozygous *TECRL* c.730+1G>C variant, with a QTc of 0.49 s, leading to the initiation of genotype-directed therapy with nadolol, flecainide, and sympathetic denervation.

Conclusion:

TECRL-related arrhythmogenic syndrome carries a substantial risk of malignant ventricular arrhythmias and SCD and may be misclassified as isolated LQTS. In all surviving patients, genetic diagnosis led to clinically meaningful changes in management, including genotype-directed use of flecainide and optimization of antiarrhythmic therapy. These cases underscore the critical role of early genetic testing and cascade family screening after sentinel SCD events.

**PO.81 | PRE-EXCITATION SYNDROME IN AN INFANT WITH INVASIVE MENINGOCOCCAL DISEASE**

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Background And Aim: pre-excitation syndrome is characterized by an early activation of the ventricles due to impulses bypassing the AV node via an accessory pathway. It is usually present from birth, however in most patients the condition is discovered incidentally on a routine ECG, whilst other patients can present with supraventricular tachycardia (SVT), atrial fibrillation, ventricular fibrillation or even sudden cardiac death.

Methods: the case is about a healthy male infant, who presented with fever, diarrhea and hemorrhagic rash. He developed hemodynamic instability, with normal ECG pattern, together with impaired level of consciousness. He was admitted to the PICU due to septic shock by *Neisseria Meningitidis* type B and intubated under sedation and inotropic support initially with noradrenaline and adrenaline. Fluid resuscitation and antibiotic therapy started immediately.

Results: During his hospitalization, he was under regular pediatric cardiological assessment, with normal anatomy of the heart and the great vessels, small amount of pericardiac fluid, EF variation 50-60% and sinus tachycardia, without signs of septic cardiomyopathy or cardiac dysfunction. He was gradually stabilized hemodynamically with no need of inotropic support. However, during the weaning phase and under intravenous administration of clonidine, he presented ECG rhythm shifts between sinus tachycardia when he was calm and initially intermittent premature ventricular contractions and then a short PR interval and a delta wave when he was anxious or febrile. He was reassessed by the pediatric cardiologist and treated with propranolol, with gradual improvement of the clinical condition together with the de-escalation of medication after 3 months under regular pediatric cardiological assessment. No electrophysiology study was made.



Conclusions: pre-excitation syndrome presentation in children may vary. Whereas older children may develop palpitation, chest pain, throat throbbing, fainting or syncope, infants usually have nonspecific symptoms such as irritability, gastrointestinal and respiratory symptoms, anxiety, and even shock. Factors that can induce pre-excitation syndrome may be infections, physical exertion, puberty or other conditions that can cause stress. Taking these into consideration, it is important for pediatricians and especially for intensivists, to diagnostically exclude SVT and pre-excitation syndrome in every sick infant or asymptomatic young child with tachyarrhythmia.



PO.82 | SURGICAL IMPRINTS ON POST-FONTAN RHYTHM CHANGES: A LONG-TERM EVALUATION

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Objective: Rhythm disturbances are significant morbidities following Fontan palliation. The primary objective of this study was to evaluate the potential impact of different surgical techniques on the development of long-term rhythm changes in post-Fontan patients

Methods: Between January 2000 and June 2026, 105 patients who were followed up at our center and underwent Fontan palliation were retrospectively screened. Eighty-two patients who met the study criteria were included in the analysis. Clinical assessments included surface electrocardiography (ECG), 24-hour Holter monitoring, and exercise stress testing. Surgical types were also recorded

Results: We included 82 patients (53 males, 29 females; median age 18 years, range 5–55). Primary diagnoses included tricuspid atresia (n=32), unbalanced atrioventricular septal defect (n=12), double outlet right ventricle (n=9), and other complex congenital heart defects. Surgical techniques included intra-extracardiac (n=44), extracardiac (n=22), lateral tunnel (n=6), and classic Fontan (n=6). The median age at Fontan surgery was 7.9 years (1.2–28), with a median follow-up of 9.2 years (2–28). At evaluation, 65 patients were in normal sinus rhythm. Conduction abnormalities and rhythm variations included first-degree AV block (n=8), paced rhythm (n=6), atrial fibrillation (n=1), junctional rhythm (n=1), and low atrial rhythm (n=1). The median heart rate was 81 bpm, with 15 patients exhibiting age-inappropriate bradycardia (9 with intra-extracardiac, 4 with extracardiac, and 2 with classic Fontan). Exercise testing (n=25) identified chronotropic incompetence in 6 patients (4 with intra-extracardiac and 2 with lateral tunnel type). Atrial arrhythmias were documented in 13 patients, 9 of whom underwent catheter ablation. During the follow-up period, there were 5 deaths, and one patient required heart transplantation.

Conclusion: The analysis of surgical techniques suggests a potential association between specific Fontan procedures and rhythm disturbances. Notably, intra-extracardiac Fontan approaches were observed more frequently in patients with bradycardia and chronotropic incompetence. However, these findings are preliminary, and further large-scale, longitudinal studies are warranted to better elucidate the impact of surgical approach on long-term postoperative rhythm outcomes.

**PO.83 | AN ANIMAL MODEL OF ATRIAL FIBRILLATION****Gabriel-Laurentiu Cismaru¹, Catalin Bota¹, Andrei Mihordea¹**¹ Cardiology, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania**Background And Aim**

Small-animal models of atrial fibrillation (AF) are valuable tools for preclinical evaluation of antiarrhythmic therapies, including those applicable to pediatric and congenital arrhythmias. Transesophageal pacing offers a minimally invasive approach to AF induction, avoiding the need for open-chest surgery. We describe a reproducible protocol for AF induction in the Wistar rat using commercially available electrophysiology equipment.

Methods

Fifteen Wistar rats were assigned to two groups: stimulation (n=10) and sham (n=5). Sham animals underwent sedation and esophageal tube insertion only. In the stimulation group, a quadripolar EP catheter was advanced into the esophagus and connected to a Micropace III stimulator. Burst pacing was delivered at a cycle length of 30 ms for 5 minutes. Atrial capture was confirmed by continuous surface ECG monitoring. AF inducibility, episode duration, and adverse events were recorded.

Results

No arrhythmias occurred in the sham group. AF was induced in 6/10 stimulated animals (60%), with episodes lasting 2–10 minutes (mean 4.5 min). A key hazard was inadvertent diaphragmatic stimulation, causing respiratory arrest followed rapidly by cardiac arrest. An innovative veterinary resuscitation technique — tail-spinning — proved superior to standard CPR with cardiac massage and epinephrine.

Conclusion

Transesophageal burst pacing achieves reproducible, self-terminating AF in Wistar rats with a 60% inducibility rate. Diaphragmatic co-stimulation is a serious hazard requiring resuscitation readiness. This model provides a practical platform for testing antiarrhythmic agents such as vernakalant and interventional strategies applicable to pediatric and congenital arrhythmias.



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**PO.84 | RISK STRATIFICATION BY TRANSESOPHAGEAL ELECTROPHYSIOLOGICAL STUDY IN ASYMPTOMATIC WPW CHILDREN UNDER 10 YEARS****Gabriel-Laurentiu Cismaru¹, Andrei Mihordea¹, Simona Cainap², Cecilia Lazea²**¹ Cardiology, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania² Pediatric Cardiology, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania**Background**

Children with Wolff-Parkinson-White (WPW) syndrome carry a risk of sudden cardiac death related to rapid antegrade conduction over the accessory pathway. Electrophysiological study (EPS) enables risk stratification by characterizing accessory pathway properties. Transesophageal EPS offers a non-invasive alternative, but data in very young children remain limited. We aimed to evaluate the feasibility and informative value of transesophageal EPS in asymptomatic WPW children under 10 years of age.

Methods

46 asymptomatic children with WPW pattern underwent transesophageal EPS (mean age 13.5 ± 4.2 years, range 1–18). Sedation was used in all children under 10 years ($n=12$, mean age 7.2 ± 2.8 years). Three risk parameters were assessed: (1) accessory pathway effective refractory period (APERP), (2) shortest cycle length with 1:1 antegrade conduction over the accessory pathway, and (3) shortest pre-excited RR interval during induced atrial fibrillation. Arrhythmia inducibility and procedural safety were also recorded.

Results

Orthodromic reentrant tachycardia was induced in 6 children and atrial fibrillation in 14. Comparison between the under-10 and over-10 groups showed no significant differences in any risk parameter: mean APERP 294 ± 36 ms vs. 287 ± 48 ms ($p=0.46$), mean shortest 1:1 conduction cycle length 282 ± 41 ms vs. 283 ± 46 ms ($p=0.61$), and mean shortest pre-excited RR interval during AF 284 ms vs. 268 ms ($p=0.65$). No complications were recorded in either group.

Conclusion

Transesophageal EPS is safe and feasible in asymptomatic WPW children under 10 years of age. Accessory pathway risk parameters are comparable to those in older children, supporting the use of this non-invasive approach for early risk stratification regardless of age. These findings may inform decisions on monitoring and ablation timing in young asymptomatic WPW patients.



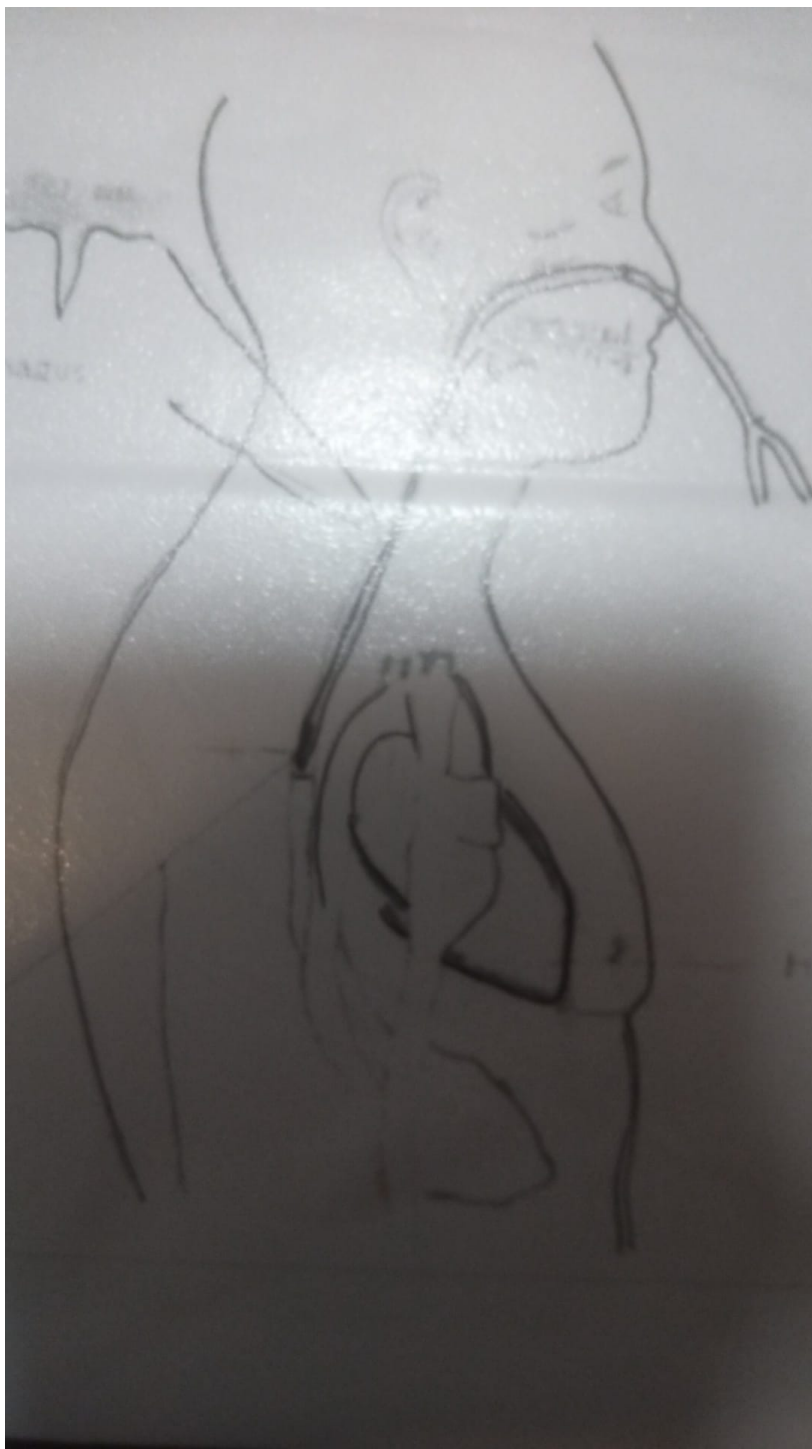
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**PO.85 | RADIOFREQUENCY ABLATION ALONG THE CORONARY OSTIUM-TRICUSPID LINE FOR AVNRT IN CHILDREN: A SINGLE-CENTRE EXPERIENCE****Gabriel-Laurentiu Cismaru¹, Andrei Mihordea¹, Cecilia Lazea¹, Simona Cainap²**¹ Cardiology, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania² Pediatric Cardiology, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania

Background: In atrioventricular nodal reentrant tachycardia (AVNRT), the slow pathway is most commonly located between the tricuspid annulus and the coronary sinus ostium. Radiofrequency (RF) applications along the coronary ostium–tricuspid (COT) line represent a targeted anatomical approach to slow pathway modification that may streamline the ablation procedure. We report our single-centre experience with this technique in a paediatric cohort.

Methods: We retrospectively analyzed all AVNRT ablations performed using the COT approach in children at our centre between January 2023 and May 2026. All procedures were performed with conventional RF current and CARTO electroanatomical mapping. Acute success rate, number of RF applications, procedural time, complications, and recurrence at 6-month follow-up were assessed. The distance between the successful ablation site and the His bundle was measured in all cases.

Results: 16 children were included (median age 9.4 years, 44% female). The mean number of RF applications was 4 ± 2 per patient. Total procedural time was 114 ± 42 minutes. Acute success rate was 100%. One complication occurred: a second-degree AV block persisting for 2 months post-ablation. At 6-month follow-up, the same patient presented with AVNRT recurrence. The mean distance between the His bundle and the successful slow pathway ablation site was 14.8 ± 6.4 mm.

Conclusion: RF ablation along the COT line is an effective and safe approach for slow pathway modification in paediatric AVNRT, achieving a 100% acute success rate with a limited number of applications and acceptable procedural times. The low recurrence rate and the safety profile support its use as a primary ablation strategy in children. Proximity to the His bundle warrants careful mapping to minimise conduction risk.



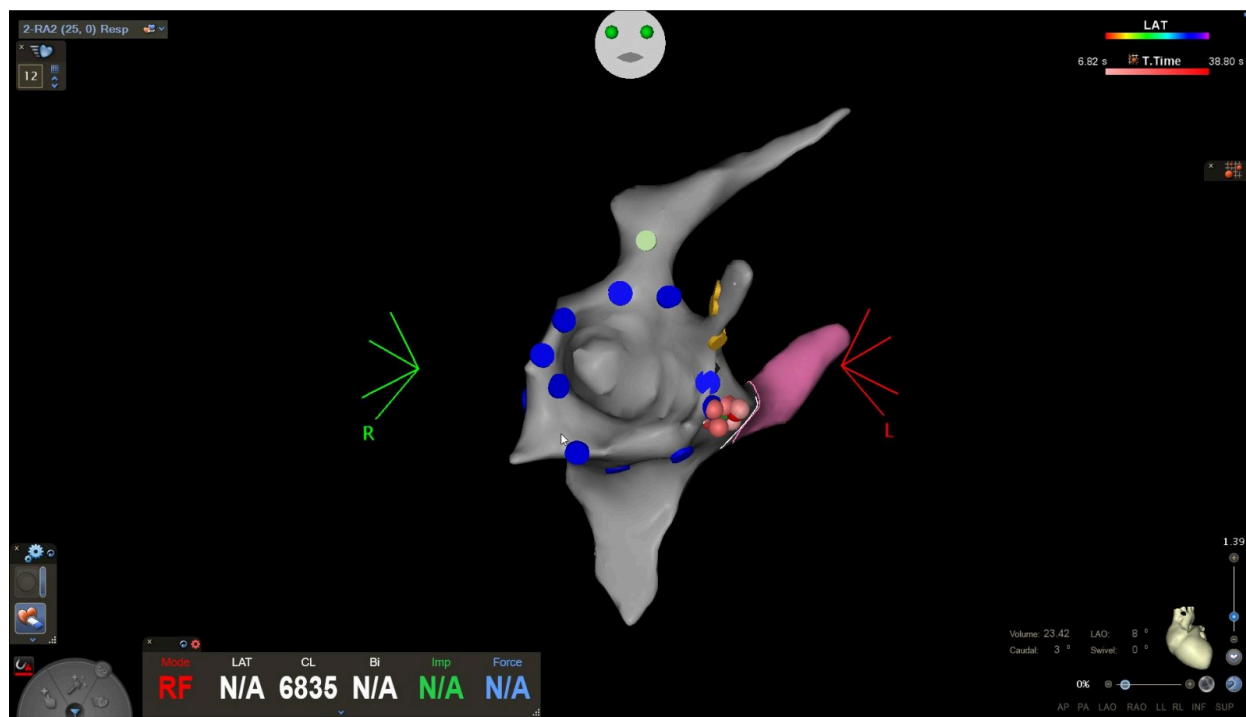
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**PO.86 | EXPANDING THE PHENOTYPIC SPECTRUM OF CACNA1C LEU762PHE: FAMILIAL VENTRICULAR FIBRILLATION WITHOUT QT PROLONGATION****Mariana Amorim Ribeiro¹, João Antunes Sarmento¹, Ricardo Alves Pinto², Joana Pimenta¹**¹ Department of Paediatric Cardiology, Local Health Unit of São João, Porto, Portugal² Department of Cardiology, Local Health Unit of São João, Porto, Portugal

Mutations in the CACNA1C gene, encoding the $\alpha 1C$ subunit of the cardiac L-type calcium channel, are associated with a broad spectrum of arrhythmia syndromes. Manifestations range from syndromic and non-syndromic long QT syndrome (LQTS) to short QT and Brugada syndromes, with marked phenotypic variability.

A 13-year-old boy suffered cardiac arrest due to ventricular fibrillation (VF) and was successfully resuscitated. He had a structurally normal heart, and baseline electrocardiogram (ECG) showed relative sinus bradycardia and normal QTc interval.

At the time he was awaiting the result of cascade genetic screening due to significant family history of arrhythmia and sudden death on the maternal side. His 17-year-old brother, with autism spectrum disorder, had survived a VF cardiac arrest the prior year and carried a heterozygous CACNA1C (NM_000719.7) c.2284C>T(p.(Leu762Phe)) variant.

Following confirmation that our patient carried the same mutation, he started nadolol and had an ICD implanted. Two years later, he presented with recurrent syncope and appropriate ICD shocks. Device interrogation revealed bradycardia followed by frequent premature ventricular complexes with bigeminy, progressing to polymorphic ventricular tachycardia and VF. Electrolytes were normal, and QTc remained within normal limits. Despite nadolol optimization and temporary right ventricular pacing at 80 bpm, VF episodes persisted. Quinidine was therefore initiated, abolishing further episodes of arrhythmia.



His brother had a similar phenotype, with normal QTc and sinus bradycardia, and developed recurrent VF triggering ICD shocks, despite beta-blocker therapy, achieving arrhythmia control only after introducing quinidine.

This report describes a novel phenotype associated with CACNA1C (NM_000719.7) c.2284C>T(p.(Leu762Phe)), characterized by normal QTc interval, sinus bradycardia, recurrent VF, and limited response to beta-blockers. To our knowledge, this variant has been reported in only one family, presenting with LQTS type 8. Our findings expand the phenotypic spectrum, suggesting that life-threatening ventricular arrhythmias may occur despite normal QTc. Complete suppression of arrhythmias with quinidine in both siblings highlights a potential therapeutic option, possibly related to its multichannel electrophysiological effects. A deeper understanding of CACNA1C mutations and their phenotypic variability is fundamental for risk stratification and targeted therapeutic strategies.

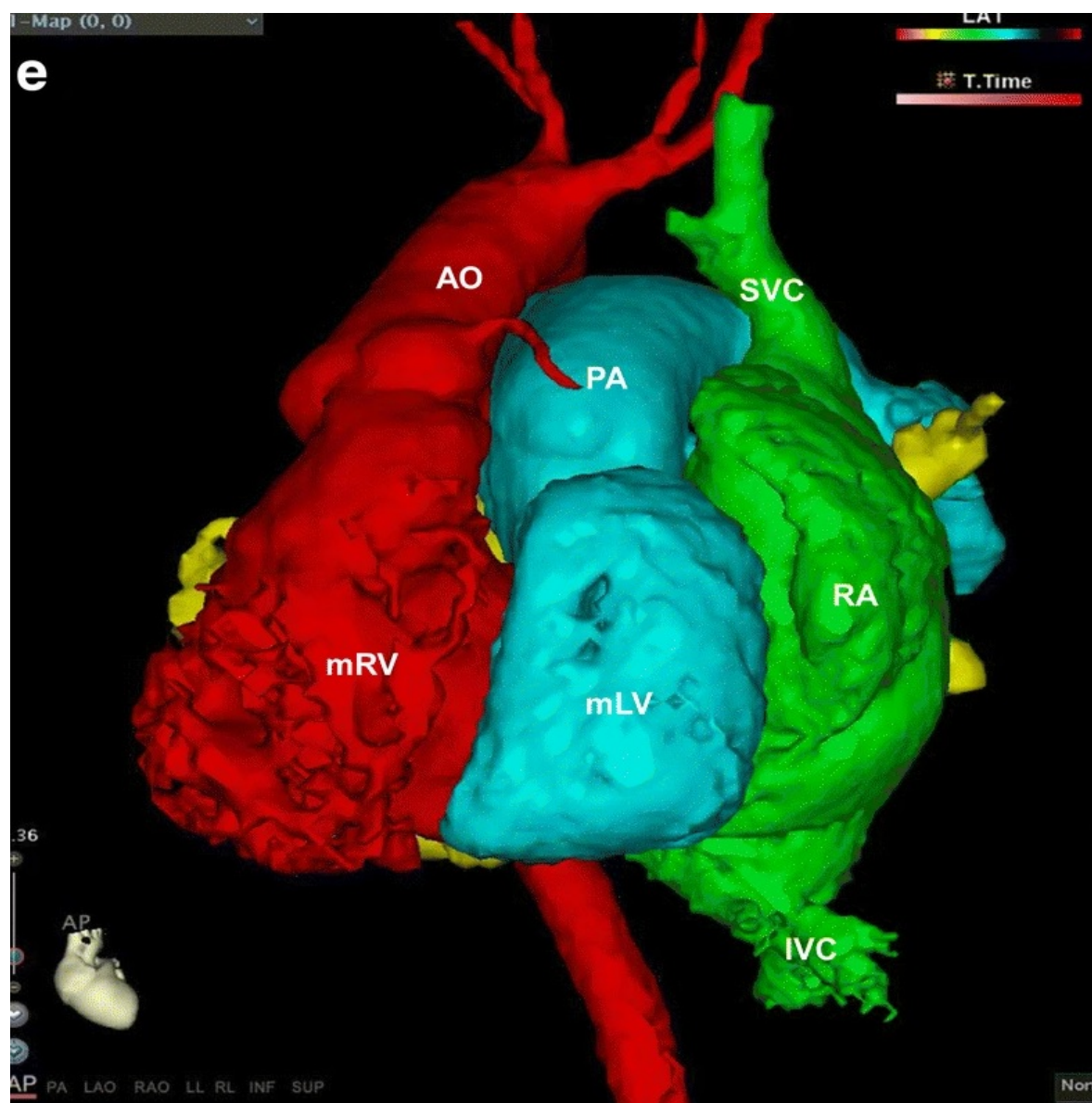
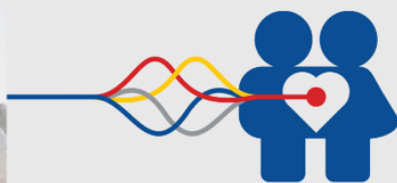
**PO.87 | SPONTANEOUS INVOLUTION OF AN ACCESSORY PATHWAY IN A PATIENT WITH CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES: A CASE REPORT WITH 2.5-YEAR FOLLOW-UP****Gabriel-Laurentiu Cismaru¹, Andrei Mihordea¹**¹ Cardiology, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania

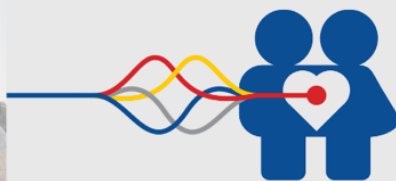
Background: Congenitally corrected transposition of the great arteries (CCTGA) is associated with progressive atrioventricular node (AVN) degeneration. Accessory pathways may coexist and pose a risk of sudden cardiac death via antegrade conduction during atrial fibrillation. We report an unusual case of spontaneous accessory pathway involution in a young non-competitive athlete with CCTGA and a WPW pattern, in whom catheter ablation carried a prohibitive risk of complete AV block.

Case Report: A 12-lead ECG revealed a right posteroseptal accessory pathway. Transesophageal EPS (TEEPS) demonstrated an accessory pathway effective refractory period (APERP) of 200 ms and a shortest pre-excited RR interval (SPERRI) of 210 ms, indicating high-risk conduction properties. Catheter ablation was recommended per current guidelines. However, intracardiac EPS revealed absent antegrade AVN conduction, making ablation inadvisable due to the risk of complete AV block. The patient declined ablation, pacemaker implantation, and antiarrhythmic therapy, and continued non-competitive sport activity after being fully informed of the risk of ventricular fibrillation.

Results: At 18-month follow-up, repeat TEEPS showed marked prolongation of the APERP to 340 ms and SPERRI to 350 ms, suggesting spontaneous accessory pathway degeneration. At 2.5 years, APERP was 310 ms and SPERRI 404 ms, consistent with ongoing involution. The patient remained asymptomatic throughout, continued his sporting activities, and declined any therapeutic intervention.

Conclusion: This case illustrates that in CCTGA, both the AVN and accessory pathway may undergo spontaneous degeneration over time, with progressive loss of high-risk conduction properties. Serial electrophysiological assessment is essential in such patients. As AVN degeneration advances, pacemaker implantation will eventually be required, underscoring the importance of long-term follow-up in this complex population.



**PO.88 | SEVERE ARRHYTHMIC PHENOTYPE IN COMBINED KCNH2 AND KCNJ2 GENETIC VARIANTS SUGGESTING DETRIMENTAL POTASSIUM CHANNEL EFFECTS****Julia Paul¹, Christine Stratmann¹**¹ Inherited Cardiovascular Disease, Great Ormond Street Hospital, London**Background and Aim:**

Anderson Tawil Syndrome (ATS1) is a rare genetic condition that predisposes to complex ventricular arrhythmia. It has been connected to loss-of function variants in the KCNJ2 gene, destabilizing the resting membrane potential. Pathogenic loss-of function variants in KCNH2 prolong cardiac repolarisation and result in Long QT syndrome (LQTS2), which is associated with cardiac arrhythmias and sudden death.

We describe a clinical case with two heterozygous missense variants affecting the cardiac potassium channels: R218Q in KCNJ2 and R537Q in KCNH2. Our patient presented with a high burden of ventricular ectopy, including complex bidirectional couplets, as well as polymorphic VT. Our objective is to describe the clinical and electrocardiographic manifestations, diagnostic pathway and experience in management of this rare constellation of genetic findings.

Case:

Our patient was referred following auscultation of an irregular heartbeat on anaesthetic pre-assessment. Resting ECG showed a normal QTc interval with prominent U-waves in lateral leads. There were no cardiac symptoms and no description of episodic muscular weakness, though characteristic facial features of ATS1 were noted. Holter monitoring revealed a 40% burden of polymorphic ventricular ectopy, with recurrent runs of asymptomatic non-sustained VT (NSVT). Exercise testing showed bidirectional couplets at rest, progressing to multi-focal triplets and a short run of polymorphic VT at peak exercise. Frequent ventricular ectopy continued into recovery, terminating by the third minute. Nadolol and Flecainide were commenced, which significantly reduced the ectopic burden and complexity of ventricular arrhythmia. Subsequent genetic testing revealed a likely pathogenic variant in KCNH2 and a pathogenic variant in KCNJ2, suggesting that this phenotype is a combination of LQTS2 and ATS1 features.



The dual antiarrhythmic therapy showed overall good protective effect, though led to quick breakthrough arrhythmia after a brief episode of medication non-compliance. This prompted the decision for left cardiac sympathetic denervation (LCSD). Loop recorder monitoring has since revealed no further complex arrhythmia with a much reduced ectopic burden.

Conclusion:

We describe an unusual case of two concomitant genetic variants affecting cardiac potassium channel function, its clinical presentation and course.

**PO.89 | SUBCUTANEOUS IMPLANTABLE DEFIBRILLATOR: A REASONABLE OPTION FOR OLDER CHILDREN AND YOUNG ADULTS WITH CONGENITAL HEART DISEASE AND INHERITED ARRHYTHMIAS****Avramidis D., Koussi T., Karafotia A., Sofianidou A., Papagiannis J.**

Background: Subcutaneous Implantable Cardiac Defibrillators (SICDs) have been increasingly used for multiple indications in adults. Experience in pediatric and adult congenital heart disease (CHD) patients is still limited. We present our single-center series of young patients with SICDs.

Methods: Review of clinical information, procedure reports and follow-up records of all patients with CHD and inherited arrhythmias as well as pediatric patients with unexplained cardiac arrest, who underwent SICD implantation or replacement in our center. Pre-implantation testing for suitability was performed in all and patients were chosen for implantation if they passed the test in at least 2/3 configurations. All procedures were performed by pediatric electrophysiologists under general anesthesia.

Results: From 8/2021 to 12/2025, 18 patients were treated by our department (ages 13-46, median 15,6 years, 83,3% males). The main diagnoses included: Cardiomyopathies in 9 (50%), CHD in 6 (33,3%), channelopathies in 2 (11%) and unexplained cardiac arrest in 1 (5%). Indication was primary prevention in 78,8% and secondary prevention in 22,2%. A replacement of a previously existing SICD was performed in one patient. In all patients an intermuscular position of the device with a 2-incision technique for the defibrillating coil was used. Two patients with CHD had a preexisting pacemaker, with no interference noted with the SICD. Two patients had SICD implanted because of inducible ventricular tachycardia during an electrophysiologic study before percutaneous pulmonary valve replacement. Defibrillation testing (DFT) was performed in 17 patients and was successful in all except for one who required repositioning of the coil. DFT was not performed in one patient with dilated cardiomyopathy and very low ejection fraction. Defibrillation impedance was 50-115 65,8±13). During 3-50 (14,5±12,9) months of follow-up, 2 patients had a shock including 1 appropriate (5%) and 1 inappropriate (5%) shocks. No infections occurred. There was one partial wound dehiscence which was repaired without sequelae.

Conclusions: SICD implantation is a reasonable option in young patients with inherited arrhythmias, cardiomyopathies, unexplained cardiac arrest and CHD, without a need for pacing and can be performed with minimal procedural risk and long-term complications.