



PJRT Spring 2020 Edition

(Pediatric Journal Review by Trainees)

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Section # 1: Ablation

Tom Carberry, MD

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Arrhythmia Mechanisms and Outcomes of Ablation in Pediatric Patients With Congenital Heart Disease

Houck CA, Chandler SF, Bogers AJJC, et al. Circ A&E 2019. [PubMed Link](#)

Background:

- Arrhythmia recurrence rates after ablation for adults with CHD are high (~40%)
- Ablation in pediatric CHD is complicated by complex anatomy and patient size
- Acute success and recurrence rates for ablation in pediatric CHD are not well studied

Objective:

- Determine arrhythmia mechanisms, procedural/long-term outcomes, and complication rates of ablation in children with CHD

Methods:

- Retrospective study of 232 pediatric patients with CHD who underwent ablation at 2 centers
- Clinical Arrhythmia Severity Score:

Documented arrhythmia		Cardioversion	
None	0	None	0
Nonsustained	1	One cardioversion	1
Sustained	2	AAIT cardioversion	1
Incessant	3	≥2 cardioversions	3
Arrhythmia severity		Antiarrhythmic medications	
Asymptomatic	0	None or digoxin only	0
Palpitations*	1	Class II or Class IV	1
Syncope/CHF/thrombosis	2	Class I or Class III	2
Cardiac arrest	3	Amiodarone toxicity	3

Results:

Arrhythmia mechanisms:

- 17% undetermined mechanism of SVT

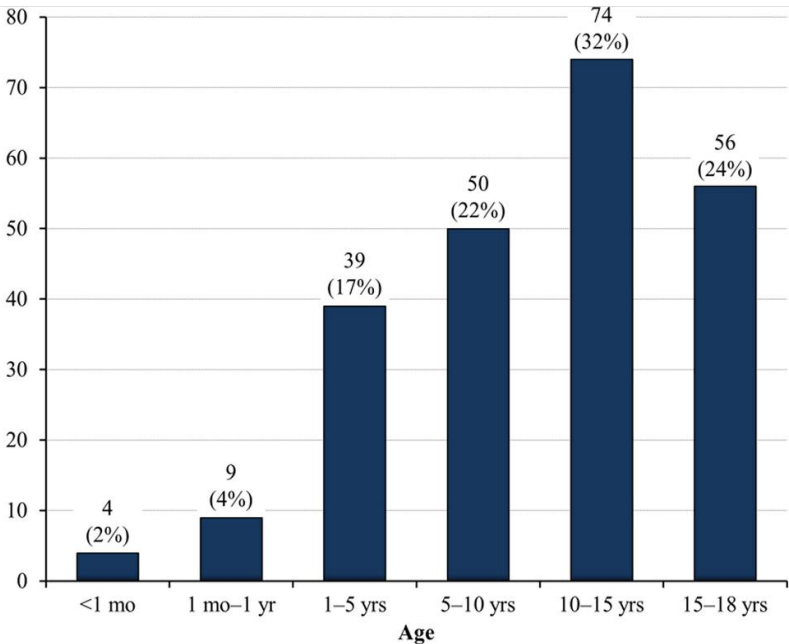
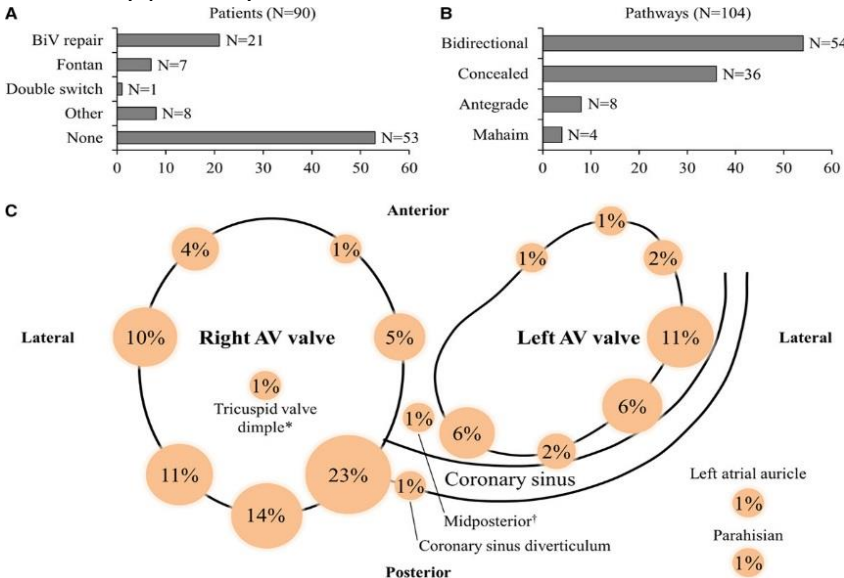
Congenital:

- 39% accessory pathway
- 12% AVNRT

Acquired:

- 24% macroreentrant atrial tachycardia
- 11% focal atrial tachycardia
- 9% ventricular tachyarrhythmia

Accessory pathway characteristics:



	Biventricular Repair, n=92	Fontan, n=39*	Double switch, n=8	Other, n=15	None, n=78
Ebstein's anomaly (n=44)	6 (7)	1 (3)	...	2 (13)	35 (45)
Septal defect (n=39)	24 (26)	15 (19)
Single ventricle (n=36)	1 (1)	28 (72)	...	7 (47)	...
TOF+variants (n=27)	21 (23)	5 (13)	...	1 (7)	...
VHD (n=21)	4 (4)	1 (3)	...	1 (7)	15 (19)
ccTGA+variants (n=21)	2 (2)	1 (3)	8 (100)	3 (20)	7 (9)
TGA+variants (n=14)	11 (12)	2 (5)	1 (1)
AVSD (n=10)	7 (8)	1 (3)	...	1 (7)	1 (1)
CoA (n=6)	6 (7)
Other (n=14)	10 (11)	4 (5)

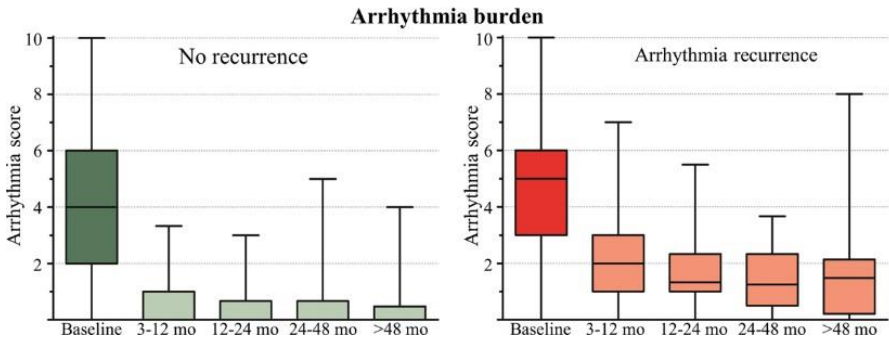
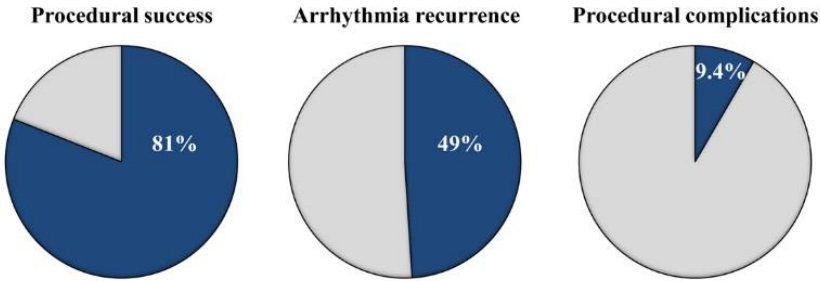
Arrhythmia Mechanisms and Outcomes of Ablation in Pediatric Patients With Congenital Heart Disease

Houck CA, Chandler SF, Bogers AJJC, et al. Circ A&E 2019. [PubMed Link](#)

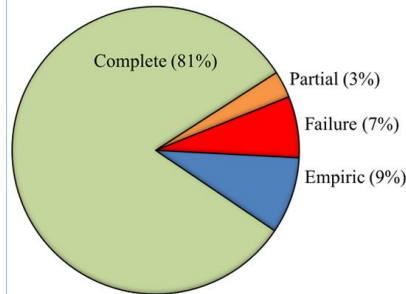
Results:

- 81% complete procedural success
- 49% arrhythmia recurrence
 - 20/51 patients with repeat procedure had new arrhythmia substrate
 - Recurrence more likely in acquired substrate
- Median arrhythmia score decreased significantly, even in recurrence group
- 9.4% complication rate
 - 1.6% (4) “major” complications
 - 1 transient CHB
 - 2 thromboembolic events
 - 1 mitral valve injury in 2.7kg infant

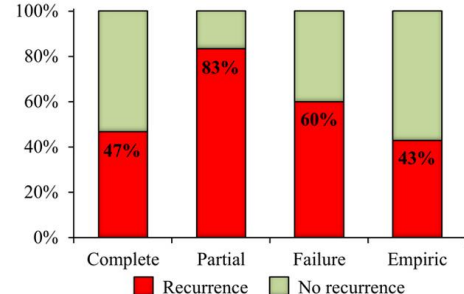
	Weight <15 kg*	Weight >15 kg	P Value
Complexity of CHD			0.053
Simple	6 (14)	25 (13)	
Moderate	14 (32)	95 (51)	
Severe	24 (55)	67 (36)	
Procedural time, min	227 (157–534)	264 (120–608)	0.018
Fluoroscopy time, min	27 (0.6–70)	22 (0.2–115)	0.053
Congenital substrate	22 (50)	97 (52)	0.823
Acquired substrate	19 (43)	79 (42)	0.910
>1 arrhythmia mechanism	7 (16)	54 (29)	0.079
Complete procedural success	38 (86)	151 (81)	0.385
Procedural complications	9 (22)	31 (19)	0.610
Arrhythmia recurrence	20 (59)	65 (46)	0.195



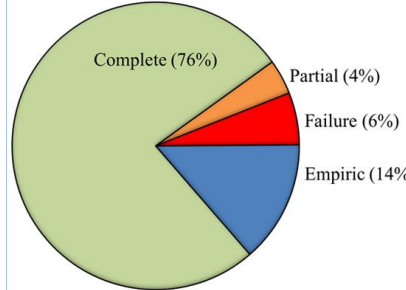
A Outcome 1st procedure (N=232)



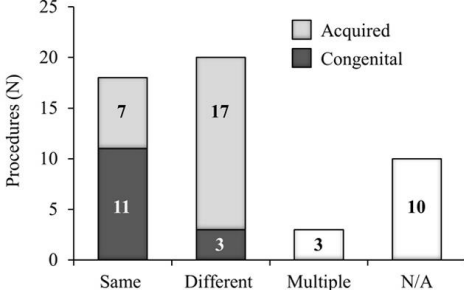
B Recurrence 1st procedure (N=174)



C Outcome repeat procedure (N=51)



D Substrate repeat procedure (N=51)



Significance:

- In pediatric CHD, **accessory pathway** is the most common substrate for arrhythmia requiring ablation
- Similar to ACHD, arrhythmia recurrence is common after ablation in pediatric CHD, but **arrhythmia burden improves despite recurrence**
- Recurrences often have different mechanism of arrhythmia



Section # 2: Clinical EP

Tom Carberry, MD

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Difficulties with invasive risk stratification performed under anesthesia in pediatric Wolff-Parkinson-White Syndrome

Schwayder MH, Escudero CA, Etheridge SP, et al. Heart Rhythm 2020. [PubMed Link](#)

Background:

- Children with WPW are at risk for SCD
- The gold standard to risk stratification in children with WPW is shortest pre-excited RR interval during a-fib (SPERRI). SPERRI ≤ 250 ms is considered high risk for SCD
- Anesthesia can affect the conduction properties of an accessory pathway

Objective:

- Compare SPERRI measured clinically to surrogate measurements made in EP lab

Methods:

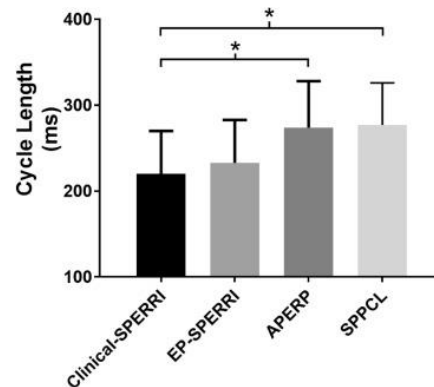
- Multicenter retrospective review of 49 children with SPERRI measured clinically and at least 1 other surrogate measurement in EP study – EP-SPERRI, APERP, SPPCL

Significance:

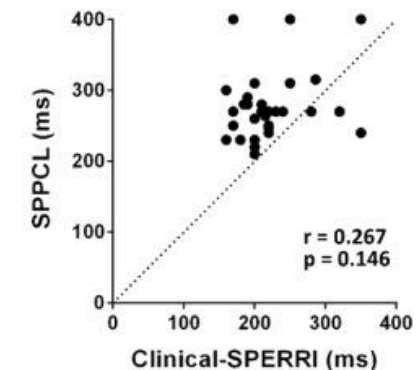
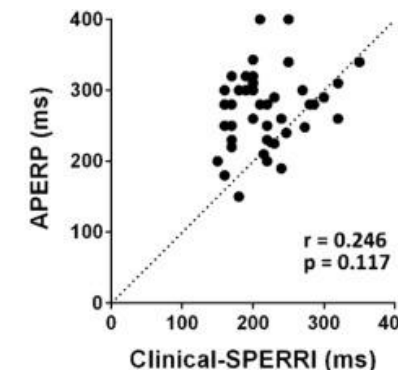
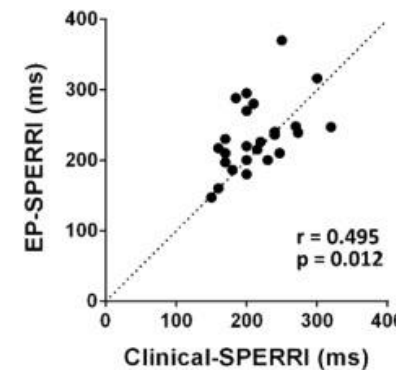
- Surrogate measurements for WPW risk stratification in the EP lab under anesthesia do not correlate well with SPERRI measured clinically
- EP study measurements may falsely classify some WPW patients as low risk

Results

- Moderate correlation between clinical SPERRI and EP-SPERRI ($r=0.495$, $p=0.012$)
- 24% of patients with high risk clinical SPERRI (≤ 250 ms) would have been classified as low risk based on EP-SPERRI > 250 ms
- No correlation between clinical SPERRI and APERP or SPPCL ($r < 0.3$, $p > 0.1$)
- Mean EP-SPERRI, APERP, and SPPCL all were greater than clinical SPERRI



Mean Clinical-SPERRI and risk stratification values. * $P < .05$.



Correlation of Clinical-SPERRI with risk stratification values

Limitatons:

- Retrospective study
- Insufficient data to perform additional analyses of type of anesthesia/sedation or EP study measurements on isoproterenol

Simple electrocardiographic criteria for rapid identification of wide QRS complex tachycardia: The new limb lead algorithm

Chen Q, Xu J, Gianni C, et al. Heart Rhythm 2020.

[PubMed Link](#)

Background:

- In wide complex tachycardia (WCT), it is often difficult to distinguish VT from SVT
- Several algorithms exist to accurately diagnosis WCT using a 12-lead ECG with varying sensitivity and specificity

Objective:

- Develop a simple algorithm to accurately diagnose WCT from a 12-lead ECG and compare this to existing algorithms

Methods:

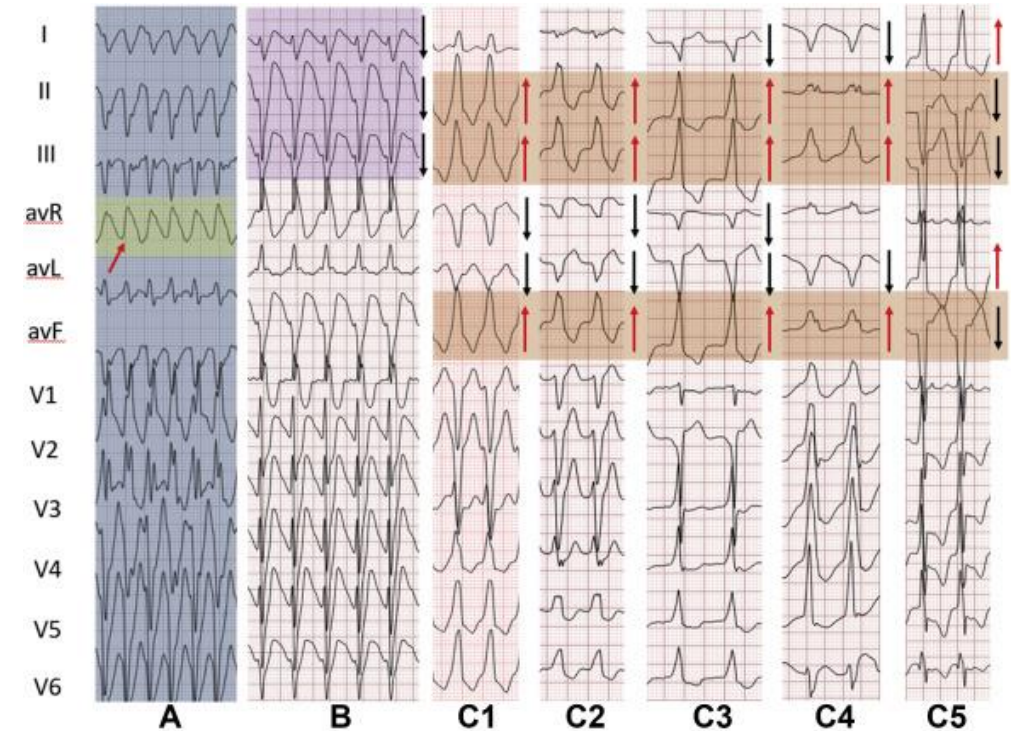
- The new limb lead algorithm (LLA) was evaluated by analyzing 528 monomorphic WCTs with diagnoses confirmed by EP study

Limb Lead Algorithm - VT if one or more of the following is present:

1. Monophasic R wave in lead aVR
2. Predominantly negative QRS in leads I, II, and III
3. Opposing QRS complex in the limb leads: concordant monophasic QRS in all 3 inferior leads and concordant monophasic QRS in 2-3 of the remaining limb leads with a polarity opposite to that of the inferior leads

Examples:

- A. Monophasic R wave in aVR
- B. Predominantly negative QRS in leads I, II, and III
- C. Opposing QRS complexes in the limb leads



Results:

Variable	LLA	Brugada	Vereckei	RWPT
Accuracy (%)	88.1 (85.0–90.7)	85.4 (82.1–88.3)	88.5 (85.4–91.15)	70.8 (66.8–74.7)
Sensitivity (%)	87.2 (83.5–90.3)	93.95 (91.1–96.1)	92.4 (89.4–94.8)	67.8 (62.9–72.3)
Specificity (%)	90.8 (84.6–95.2)	59.5 (50.6–68.0)	76.3 (68.1–83.3)	80.2 (72.3–86.6)
PPV (%)	96.7 (94.4–98.0)	87.6 (85.1–89.7)	92.2 (89.7–94.2)	91.2 (87.9–93.6)
NPV (%)	70.0 (64.2–75.2)	76.5 (68.3–83.1)	76.9 (70.0–82.7)	45.1 (41.0–49.2)

Significance: New LLA algorithm provides a more specific tool to identify VT using a 12-lead ECG that can complement existing algorithms



Section # 3: Devices

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RISK FACTORS FOR LETHAL ARRHYTHMIC EVENTS IN CHILDREN AND ADOLESCENTS WITH HYPERTROPHIC CARDIOMYOPATHY AND AN IMPLANTABLE DEFIBRILLATOR

Balaji S, DiLorenzo MP, Fish FA, Etheridge SP, Aziz PF, Russell MW, Tisma S, Pflaumer A, Sreeram N, Kubus P, Law IH. Heart rhythm. 2019.

BACKGROUND:

Implantable cardioverter-defibrillators (ICDs) have changed the natural history for patients with HCM resuscitated from sudden cardiac death (SCD) or at high risk for lethal arrhythmic events (LAE). Unfortunately, younger patients are at increased risk of complications associated with ICDs and required more lead and device revisions. Currently, there are no clear risk factors identified in children less than 16 years with HCM for primary prevention of SCD.

Prior to this publication, there was a limited amount of data of clinical risk factors predicting SCD in childhood hypertrophic cardiomyopathy as reported in recent meta-analysis by Norrish G et al.

This study aimed to look for predictors of LAE and SCD in children with HCM and ICD.

SUMMARY:

Retrospective study: 446 children and adolescents 21 years and younger with “phenotypic” HCM and ICD.

Patients were grouped in those with secondary prevention ICDs or had a primary prevention ICDs with appropriate intervention (shocks or ATP) and those with primary prevention ICDs without appropriate therapies.

RESULTS: 34% children in group 1 and 66% in group 2. Risk factors by univariate analysis were Septal thickness, posterior left ventricular wall thickness, lower LV outflow gradient < 30 mmHg and Q wave > 3mm in inferior electrocardiographic leads.

Risk factors for SCD by multivariate analysis were age of ICD placement, LV posterior wall thickness and LV outflow gradient < 30 mmHg. LV posterior wall z score > 5 was associated with LAE.

SIGNIFICANCE:

This multicenter pediatric study identified difference in risk factors associated with LAE in children with HCM in comparison to conventionally accepted risk factors in adults. Limitations of this study include retrospective data collection, not all data available for statistical analysis, and potential differences in techniques and measurements by different institutions. Moreover, this study may not truly reflect risk of SCD as ICD intervention or LAE may not equate to SCD event.

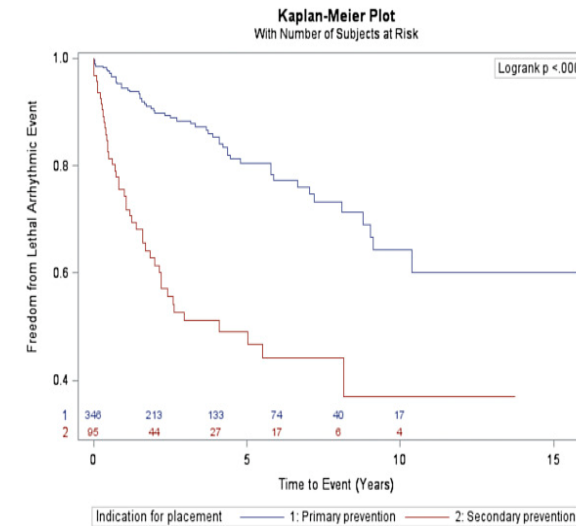


Figure 1

Time to first appropriate intervention based on indication for implantable cardioverter-defibrillator placement.

Table S4: Risk factors for Lethal Arrhythmic Events: Multivariate analysis.

Parameter	HR	95% CI	P value
Age at ICD (continuous)	0.940	0.903, 0.978	0.0025
Echo Gradient <30 mmHg	2.016	1.228, 3.308	0.0056
Posterior Wall Thick- ness z score	1.018	1.006, 1.031	0.0042

Permanent conduction system pacing for congenitally corrected transposition of the great arteries: A Pediatric and Congenital Electrophysiology Society (PACES)/International Society for Adult Congenital Heart Disease (ISACHD) Collaborative Study

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Background

- Patients with CC-TGA have a 2% annual risk for spontaneous AV block
- 25% with isolated CC-TGA and up to 67% with other structural heart disease develop RV failure by 45 years of age
- RV failure is often hastened by pacemaker implantation, but can be reversed with effective cardiac resynchronization therapy (CRT)
- 20% of patients have anatomic variation in CS, making CRT challenging
- Another strategy is to pace the His bundle or the left bundle branch
- Objective: Evaluate the feasibility and mid-term outcomes of cardiac conduction system pacing for patients with CC-TGA**

Methods

- Multicenter retrospective review
- Inclusion criteria:
 - Underlying cardiac anatomy CC-TGA
 - Underwent attempted His-bundle pacing
- Exclusion criteria:
 - Patients status-post anatomic repair
 - Conventional CRT system had been previously implanted
- Implantation and mapping:
 - 3D electroanatomic mapping (EnSite or Carto)
 - Detailed annotation of His and left bundle potentials
 - Pre-pectoral pacemaker prep and subclavian or axillary access
 - Medtronic SelectSecure 3830 lead delivered via C315H or steerable C304 sheath into His or left bundle
 - If no mapping available, His potentials or pace-mapping was used

Table 1 Baseline characteristics of the 15 CCTGA patients

Characteristic	CCTGA cases (N = 15)
Male	13 (87)
Coexisting congenital anomaly	
Pulmonary stenosis	3 (20)
VSD	5 (33)
Tricuspid anomaly	6 (40)
Dextrocardia	4 (27)
Corrective surgery	8 (53)
Onset AV block (y)	15 (1–20)
AV block type	
Intermittent/high grade	4 (27)
Complete	10 (67)
Prior pacemaker/defibrillator	7 (47)
Medical therapy	
ACE/ARB	5 (33)
BB	5 (33)
Warfarin	3 (20)
NYHA functional class	1 (1–2)
RV systolic dysfunction	
Mild	10 (67)
Moderate	3 (20)
Severe	2 (13)

Values are given as n (%) or median (interquartile range).

Table 2 Acute and chronic pacing electrical parameters

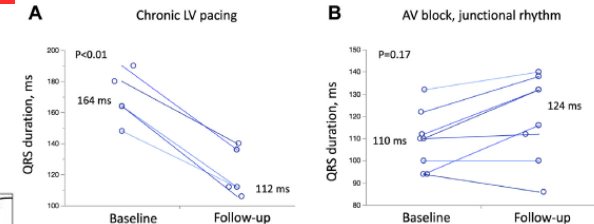
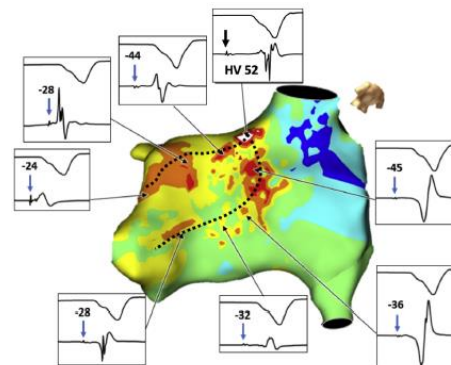
Pacing parameter	Implant	8 months	P value
Sensing threshold (mV)	6 (4–17)	11 (5–18)	.76
Pacing threshold			
@0.5 ms	0.5 (0.5–0.7)	1.0 (0.6–1.1)	.12
@1 ms	0.5 (0.4–1.5)	0.75*	.49
Paced QRS duration	112 (95–136)	116 (109–135)	.63

Nonselective pacing thresholds for all patients are shown. Values are given as median (interquartile range).

Results

- 15 patients, median 23 (IQR 15–36) years, 87% male
- 7 patients already had a cardiac device (2 ICD, 5 pacemaker)
- Electroanatomic mapping was used in 9 cases
- His-bundle pacing was successful in 13 cases (8 of 9 with 3D mapping)
- His bundle was located at superior aspect of ventricular septum below pulmonary valve, and separate from mitral annulus unlike normal cardiac anatomy where the His bundle is at the atrial aspect of the tricuspid valve
- After 8 months of follow-up, there were no complications

His-Purkinje network in CCTGA



Left: 3D mapping of conduction system in CC-TGA. Dashed black lines indicate the left bundle branches.

Top: Change in QRS duration from baseline to follow-up in patients with pre-existing LV pacing (middle) and patients with AVB and junctional escape (right)

Limitations

- Retrospective review
- Approach is not yet widespread, so this is a small sample size.

Conclusions

- Permanent conduction system pacing is feasible in patients with CC-TGA
- Superficial location of His bundle in the ventricular septum may favor this approach over conventional CRT.
- Narrow paced QRS and stable lead parameters were demonstrated at mid-term follow-up