PJRT
(Pediatric Journal Review by Trainees)

December 2018

Fellows Participants:

Louis Rigos, MD – Cohen Children's Hospital

Henry Chubb, MD, Phd – Stanford University – Lucile Packard Children's Hospital

Chalese Richardson, MD – Columbia University - CHONY

Ellis Rochelson, MD – Albert Einstein College of Medicine – The Children's Hospital at Montefiore

Education Committee Advisors:

Andrew Blaufox, MD and Scott Ceresnak, MD

Section # 1: Devices
Henry Chubb, MD

Dual-Site Ventricular Pacing in Patients With Fontan Physiology and Heart Block

Does it Mitigate the Detrimental Effects of Single-Site Ventricular Pacing?

Edward T. O'Leary, Kimberlee Gauvreau, Mark E. Alexander, Puja Banka, Vassilios J. Bezzerides, Francis Fynn-Thompson, John K. Triedman, Edward P. Walsh, Douglas Y. Mah

J Am Coll Cardiol EP 2018;4:1289–97)

Background

Ventricular pacing in the single ventricle (SV) patient is increasingly recognized to be detrimental to long-term outcomes (Bulic et al- see top table on right). Clearly there are many confounding factors, and SV patients who develop AV block are most likely to have co-morbidities (such as heterotaxy) or to have undergone additional surgical interventions (such as tricuspid annuloplasty in HLHS). None-the-less, some of the detrimental effect may be related to pacing induced dyssynchrony, and therefore there may be a role for multisite pacing.

Prior to this publication, there was a limited amount of data in the largest pediatric/CHD CRT studies that related to SV patients (see bottom table on right). However, there was a suggestion that some did respond to multisite pacing, particularly from the Boston group (Cecchin et al) who are the authors of this new study.

This study aimed to compare the impact of multisite ventricular pacing to single-site ventricular pacing in the SV population

Table 3 Univariable factors associated with heart transplantation or death

	OR (95% CI)	*P* value
Moderate-to-severe systolic dysfunction	25 (4.7–131)	<.0001
Moderate-to-severe AVVR	9.45 (1.6–54)	.01
Ventricular pacing	4.9 (1.1–22.7)	.04
Systemic ventricle (LV)	0.13 (0.02–1.1)	.06

AVVR = atrioventricular valvar regurgitation; CI = confidence interval; OR = odds ratio.

Bulic et al,"Ventricular pacing in single ventricles- A bad combination" HRJ 2017;14:853-7

Table 4. Studies That Reported Response to CRT in Patients With Single Ventricles

	Dubin et al,[44] 2005	Cecchin et al,[42] 2009	Janousek et al,[45] 2009
Total patients single ventricles, n	7	13 but only 11 with >3 mo of follow-up	4
Median age (range)	3.1 y (5 mo–23.7 y)	17.3 y (0.5–42.5 y)	10.3 y (3.7–30.3 y)
Conventional pacing before CRT, n (%)	…	8 (61.5)	3 (75)
Median pre-CRT QRSd, ms	…	129	…
Median pre-CRT EF, %	…	37	…
Outcomes after CRT			
Change in QRSd, ms	↓ 44.8±26.2 (mean)	↓ 13 (median)	…
Change in EF units	No change	↑ 11 (median)	…
Clinical improvement, n (%)	2 (28.6)	10 (90.9)	3 (75)
Nonresponders, n (%)	5 (71.4)	1 (9.1)	1 (25)

CRT indicates cardiac resynchronization therapy; EF, ejection fraction; and QRSd, QRS duration.

Motonaga and Dubin, Circ 2014;18:1879-91

Summary

Retrospective study: 62 ventricularly paced SV patients (19 CRT and 43 single site, 1990-2016).

Primary endpoint: death or transplant. Secondary endpoint: development of abnormal vent function or significant AVVR.

Patient groups: As with all retrospective studies, there are differences in the characteristics of the CRT vs single-site pacing groups- most notably, the CRT group had poorer function at baseline and shorter follow-up (CRT implants were presumably generally more recent).

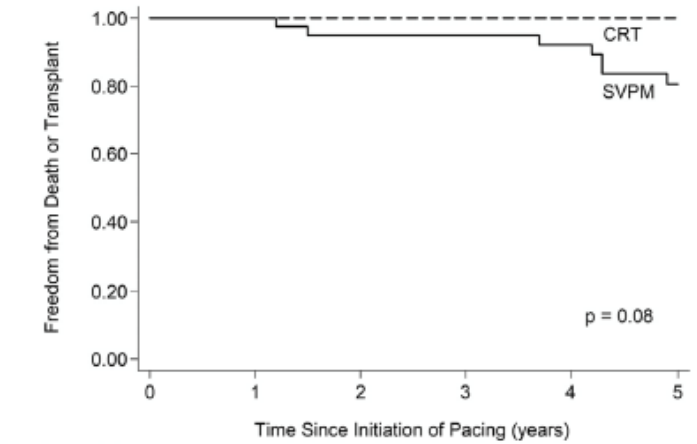
Primary endpoint: this was reached by 1 CRT and 11 single-site patients- not a significant difference (p=0.08)

Secondary endpoints: generally analyses were non-significant. However, there was a significant difference in the change in ventricular function: those with single site pacing tended to deteriorate (p=0.009). There is no comment on the pacing-related complications in the two groups (eg cardiac strangulation, lead failure)

Significance

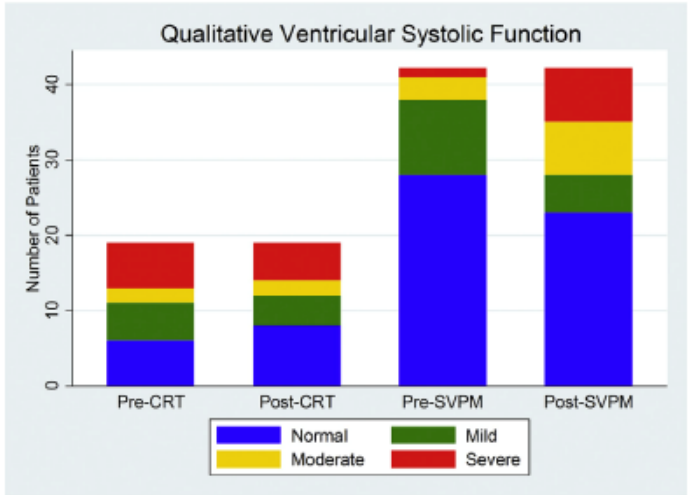
This is the largest published study of CRT for the single ventricle and is largely negative in its results. However, the cohort size is small and confounded by different baseline characteristics- it is clearly underpowered, and the authors suggest that a prospective multi-center study is warranted. There is the tantalizing possibility that CRT may mitigate some of the dire consequences of pacing the SV, and the question remains as to who should receive multisite pacing. However, at present both the risks and benefits of CRT remain unquantified.

FIGURE 2 Kaplan-Meier Curve Comparing Freedom From Death or Transplant in SVPM and CRT Patients



Kaplan-Meier curve comparing freedom from death or transplant in single-site ventricular pacemaker (SVPM) and cardiac resynchronization therapy (CRT) patients limited to 5 years of follow-up.

FIGURE 3 Qualitative Ventricular Systolic Function Before Device Implantation and at Study Follow-Up



CRT = cardiac resynchronization therapy; SVPM = single-site ventricular pacemaker

Coronary artery compression from epicardial leads: More common than we think

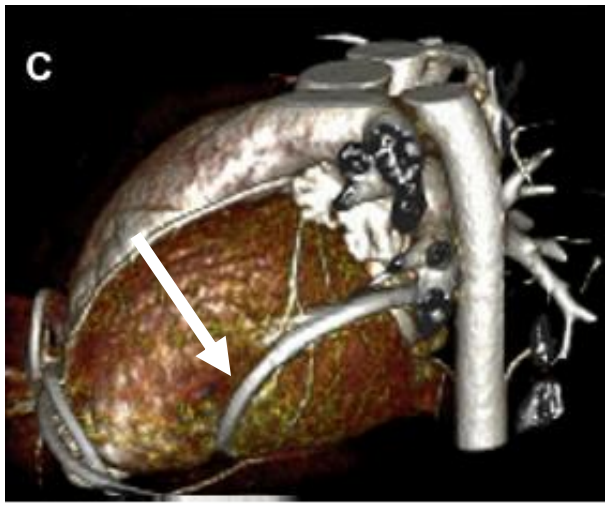
Douglas Y. Mah, Ashwin Prakash, Diego Porras, Francis Fynn-Thompson, Elizabeth S. DeWitt, Puja Banka

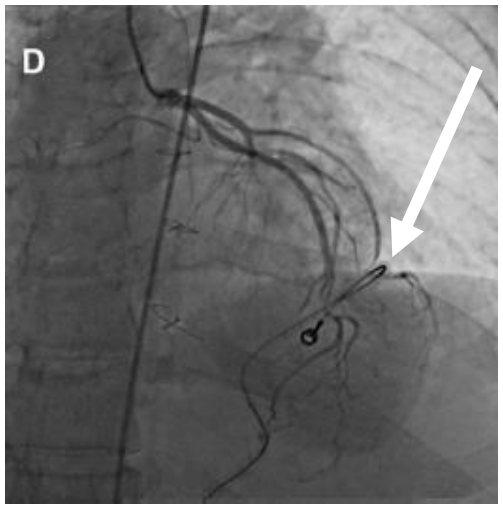
Heart Rhythm 2018;15:1439-47

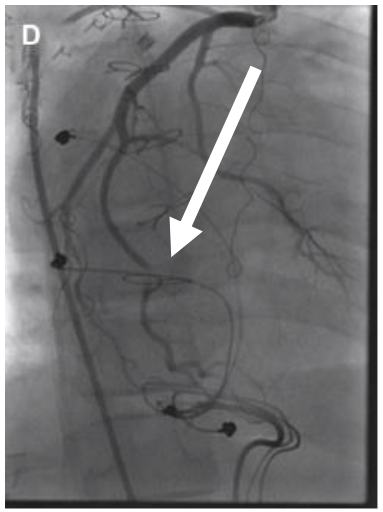
Background

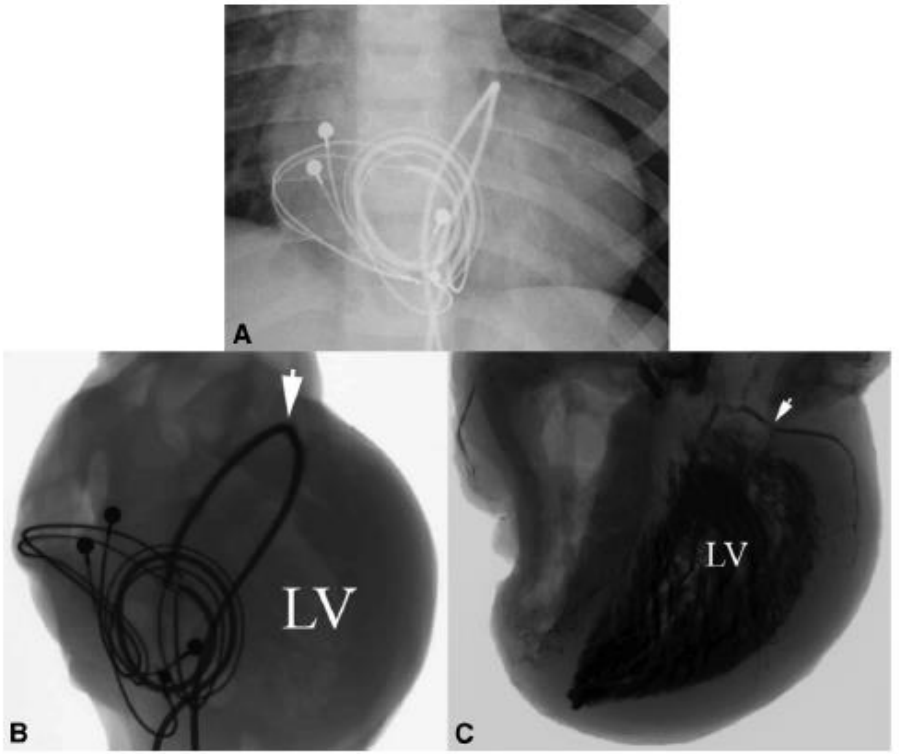
Leading on from the other study reviewed in this edition (CRT in the single ventricle), this study discusses a concern for patients with epicardial pacing leads. There have been several reports of 'cardiac strangulation' following epicardial pacing (see figure on right), but the prevalence of coronary artery compression is less well described.

With somatic growth, there is the potential for epicardial leads to wrap around the heart when additional loops are placed, or may be drawn into, the pericardial space ('cardiac strangulation'). However, what has been less well appreciated is that adherence of the leads to the epicardium may pull on the myocardium in their own right, without any wrapping around the heart. This in turn may result in coronary compression (see below).









A, Last antemortem posteroanterior chest radiograph of patient 2. B, Postmortem cardiac radiograph of patient 2. The solid arrow indicates the common biventricular epicardial lead situated on the atrioventricular groove. C, Postmortem cardiac angiography, with barium, of patient 2, depicting the compression of the circumflex coronary artery by the common biventricular EP lead. The solid arrow shows the area of compression in the circumflex coronary artery that was injected with barium after postmortem removal of the EP and all associated leads.

Carreras et al, J Thorac Cardiovasc Surg 2015; 149:522-7

Summary

Patient group: 145 patients with epicardial leads (age 1day-43yr) who underwent coronary imaging at any stage (angiography (n=126) or CT (n=37)).

Results: the method of diagnosis was complex, with many having multiple (and conflicting) imaging study results (CXR/CT/angio). In total, 8 (6%) were diagnosed with coronary artery compression, 1 post-mortem.

On CXR screening, the most accessible imaging modality, the authors categorized 119 as 'negative'. Amongst these, there were 2 coronary compressions, meaning a sensitivity of 67%, falling to 57% if the 13 'equivocal' CXRs are assumed to be negative. Sensitivity increases to 83% if only those with 'negative' CXR and no symptoms are considered to be negative on screening.

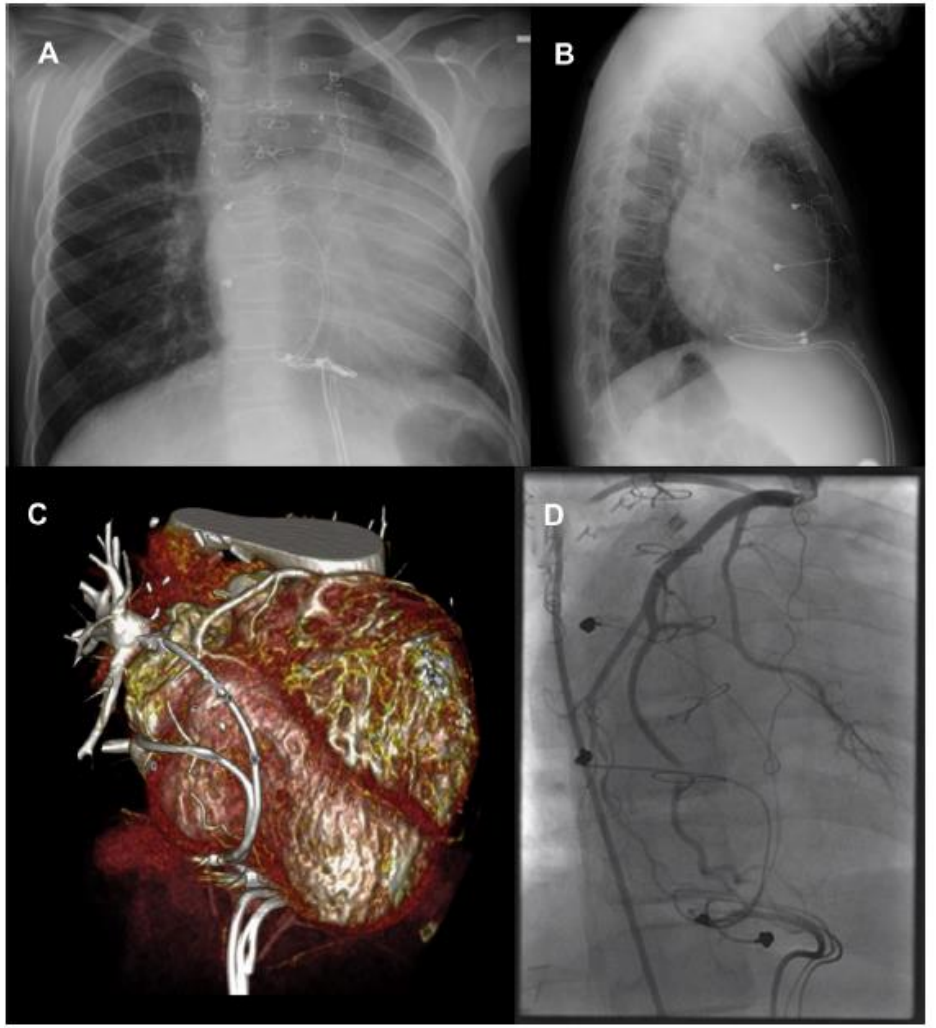
For those that had surgical correction of lead positions, there were no significant complications. Somewhat concerning, though, the one patient with repeat imaging post surgery continued to have (milder) coronary compression by fibrous sheath.

Significance

The prevalence of coronary compression in this patient cohort is much higher than previously reported, and the complication is important to consider in the planning of future lead implantations. However, two of main questions that this study provokes is how to find the patients with coronary compression, and then what to do once they are identified.

For identification, the CXR is the obvious initial screening tool but the signature findings of the 'suspicious' CXR are not entirely clear from the methods. The methods refer to the publication of Carreras et al (see image on previous page), but Carreras described only the 'classic' form, with posterior looping of the leads forming a "heart-shaped" appearance. In this study, only 2 had 'classic' pattern. The remaining 7 (or 15 including the equivocal) seem to have had some encroachment of the leads within the cardiac silhouette, but this is not stated categorically and further guidance is likely to be required. The need for and the ideal means to screening patients with epicardial devices remains to be determined.

Once identified, the natural history without reoperation for those with coronary compression is not clear. In this study many were symptomatic, but with increased screening it is likely that many more asymptomatic leads with varying degrees of coronary compression may be identified. The most prudent course for these patients also remains to be determined.



Chest X-ray films in the posteroanterior (A) and lateral (B) projections showing a nonclassic pattern of epicardial leads causing coronary artery compression. The atrial lead is seen wrapping around the right heart border on the lateral film. The "bend" of the lead in the posteroanterior film is within the cardiac silhouette. C: Computed tomography shows the lead constricting the right atrium and ventricle, along with the circumflex artery in a patient with corrected transposition of the great arteries. D: Catheter angiography shows loss of contrast within the circumflex and obtuse marginal branch as they course below the atrial lead.

Section #2: Ablation
Ellis Rochelson, MD

Safety and Efficacy of Radiofrequency Catheter Ablation for Tachyarrhythmia in Children Weighing Less Than 10 kg

Ozaki N, Nakamura Y, Suzuki T, et al. Pediatr Cardiol. 2018 Feb;39(2):384-389.
The Department of Pediatrics, Osaka Medical College, Japan (PDF link)

Background

Though ablation has become the standard of care for many types of tachycardia in school-age children, it is less commonly performed in small infants.

This may be due to concerns of high complication rates or poor results.

Research Question

What are the outcomes of RF ablation in small infants (<10 kg)?

Methods

Multi-center retrospective review of pediatric radiofrequency ablations, 2008 - 2016 (n = 307)

22 patients <10 kg were identified

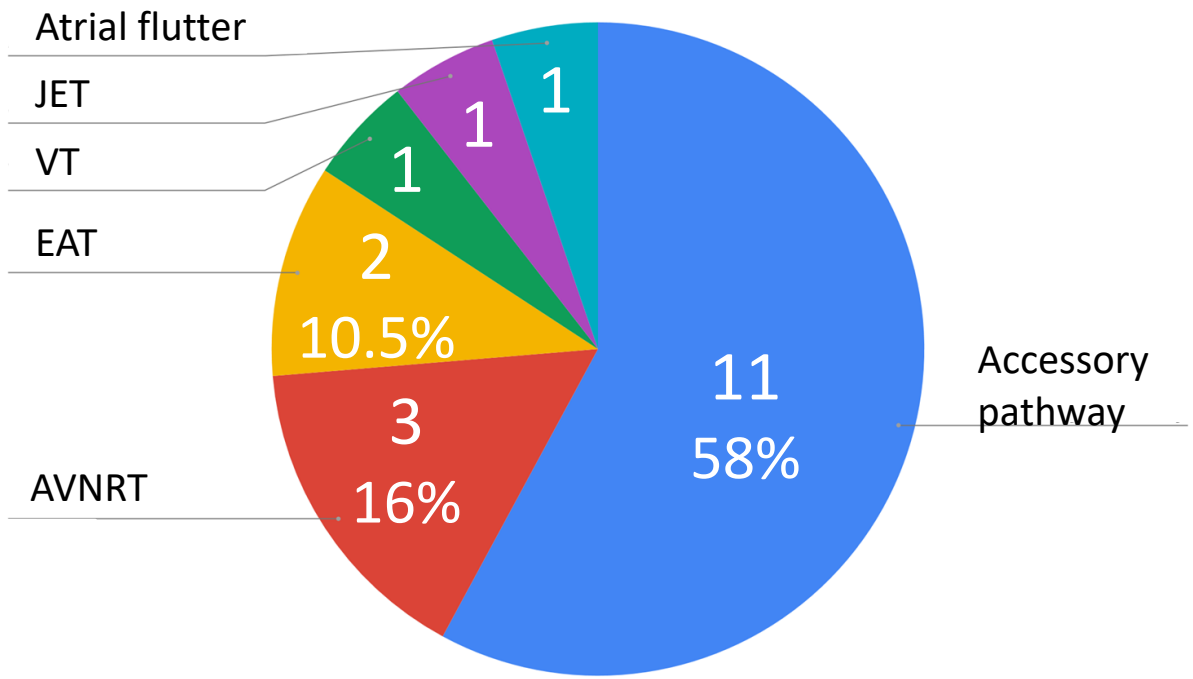
Safety and Efficacy of Radiofrequency Catheter Ablation for Tachyarrhythmia in Children Weighing Less Than 10 kg

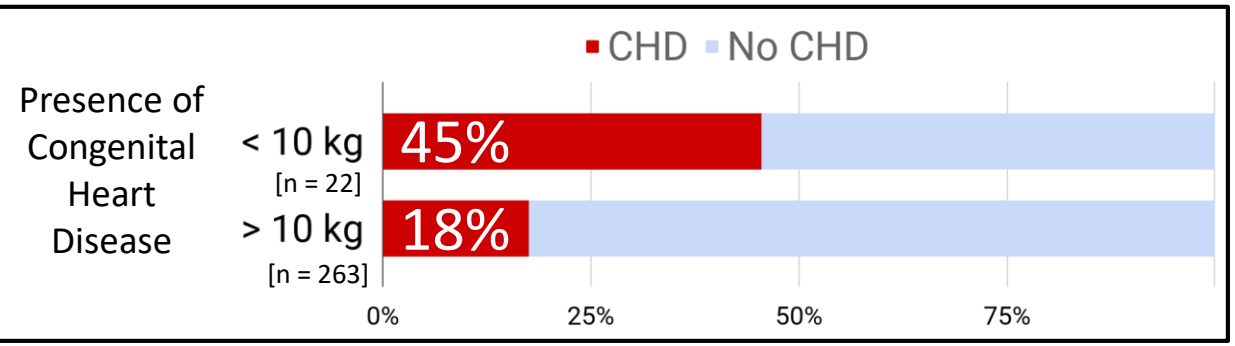
Ozaki N, Nakamura Y, Suzuki T, et al. Pediatr Cardiol. 2018.
The Department of Pediatrics, Osaka Medical College, Japan

Features of RF ablations in patients < 10 kg (n = 22)

Article PDF

Tachycardia type





Acute success rate:
91%
(20 of 22)

Medium-term success rate:
85%
(mean follow-up: 6.5 years)

Complications

- RBBB (n = 1)
- Transient AV block (n = 1)

Conclusion

Radiofrequency catheter ablation can be safely performed in small infants <10 kg with similar success and safety profiles as in older children.

Limitations

- Retrospective study
- Small number of patients

Section #3: CV Genetics
Louis Rigos, MD

Loss of p21-activated kinase 1 (Pak1) promotes atrial arrhythmic activity

Desantiago et al. - Heart Rhythm Volume *15, Issue 8,- 2018*

Background

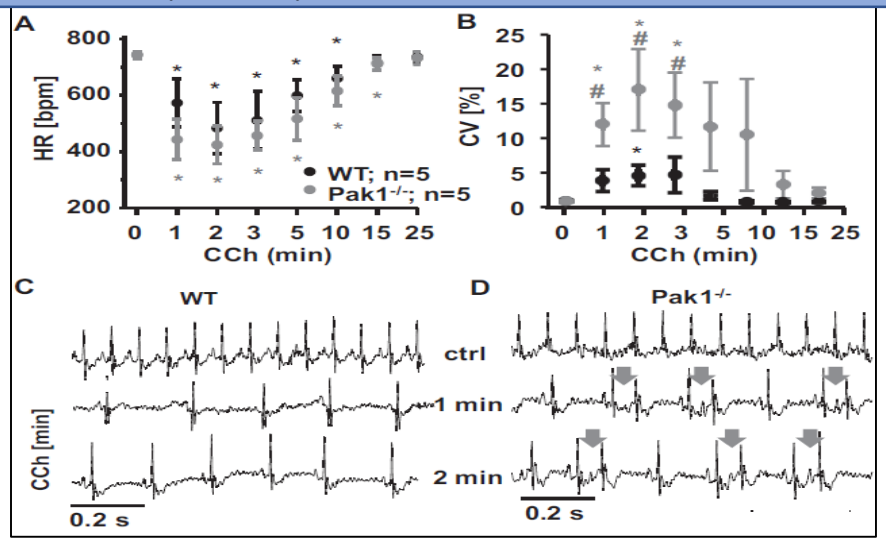
Atrial fibrillation (AF) is initiated through arrhythmic atrial excitation with Angiotensin II (AngII) implicated as an inciting factor through atrial remodeling, changes in CA^{+2} handling and production of reactive oxygen species (ROS) in myocytes. P21-activated kinase 1 (Pak 1) is a downstream target in Ang II signaling cascade whose deficiency exaggerates cardiac hypertrophic response and damage after ischemia-reperfusion injury in atrial tissue while regulating calcium release & T-tubular structure in the ventricle. In atrial tissue a connection between AngII- and NOX2-dependent ROS production has been proposed and NOX2 expression shown to be increased in humans with persistent and post-operative Atrial Fibrillation. The atria of Pak1 $^{-/-}$ mice included did not exhibit tissue fibrosis and normal baseline ECG.

NCX, whose activity depends on the membrane potential and the ion gradient for Na^{+1} and Ca^{+2} , can promote the occurrence of spontaneous afterdepolarization. During diastole, NCX removes Ca^{+2} from the cytoplasm, with resulting depolarization of the membrane potential which in turn can trigger action potentials.

Hypothesis: Modulation of PAK 1 activity may influence the occurrence of atrial arrhythmic events as well as attenuate ROS induced Ca^{+2} deregulation in atrial myocytes from a model of ventricular tachypacing–induced AF (VT-AMs).

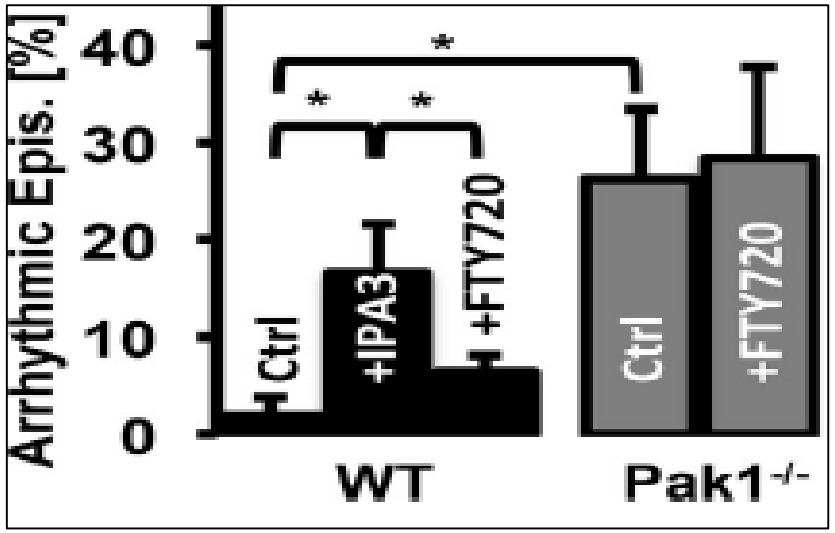
Methods

- Wild-type (control) and PAK 1 $^{-/-}$ mice atrial myocytes (AM) included
- Intraperitoneal carbachol (150 ng/g) was used for atrial arrhythmia induction
- To determine if arrhythogenecity of PAK 1 $^{-/-}$ hearts can be mimicked by acute Pak1 inhibition, WT hearts were perfused with the allosteric Pak1 inhibitor, (IPA-3) and attenuated by a PAK 1 stimulator (FTY720), and treated with NADPH inhibitor (APO), or NOX-2 deficient mice (gp91$^{phox-/-}$).



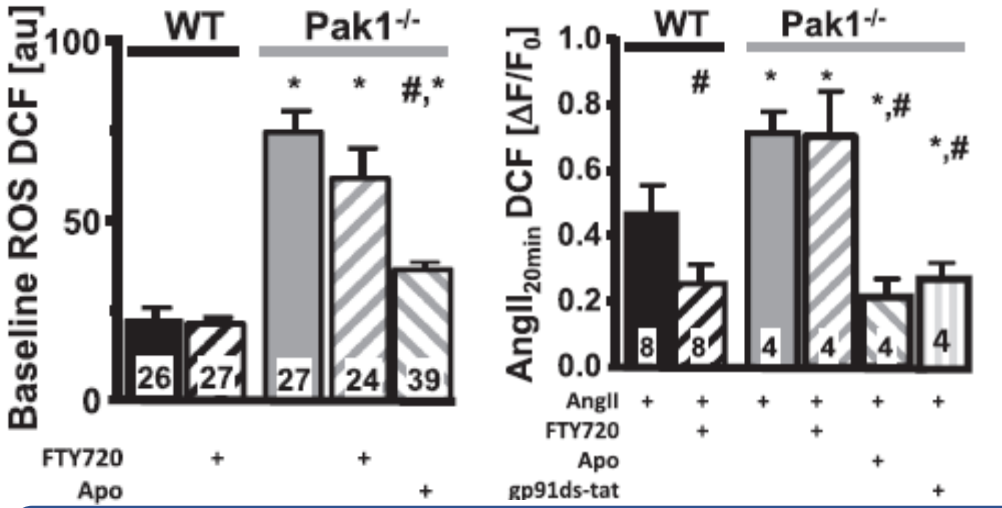
Heart rate/variability response after carbachol (cch)

(A): Comparable transient decrease in heart rate (B): Coefficient of variation of R-R interval increased in the PAK 1 $^{-/-}$ (C/D): Frequent spontaneous PACs in PAK 1 -/- compared to wild type (WT)

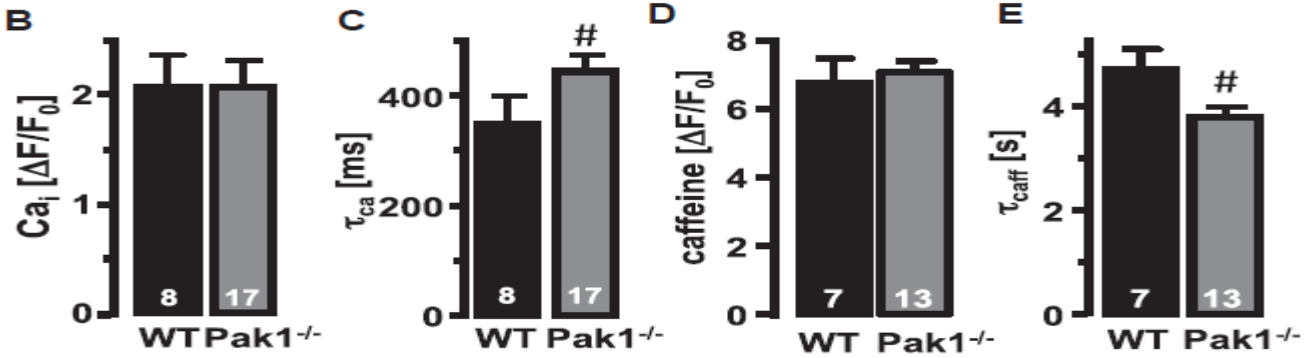


% of burst pacing episodes that induced arrhythmic events in Pak1 $^{-/-}$ and WT hearts under Ctrl conditions and during treatment with IPA-3 or FTY720

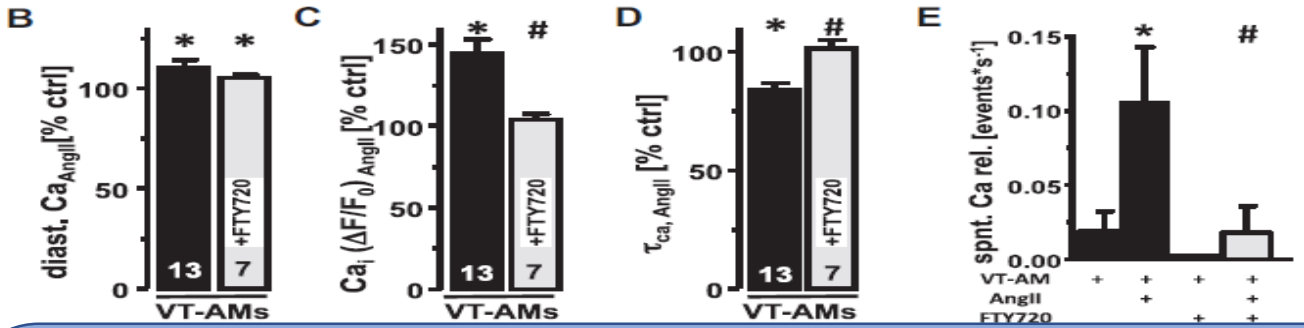
-Episodes were short in duration, with 80%, < 1 second.



Reactive O2 species were elevated in PAK 1 $^{-/-}$ compared to WT & exaggerated with Ang II and attenuated with APO & gp91$^{phox-/-}$



Despite higher ROS levels in Pak1$^{-/-}$, Ca^{+2} transient amplitudes were not different from those in WT (Figure 5B). However, the Ca^{+2} transient decay constant tCa was prolonged in Pak1$^{-/-}$ (Figure 5C). No difference with Ca^{+2} release from the intracellular stores with caffeine, but removal (which reflects the activity of NCX), was accelerated (Figures 5D and 5E).



(B): Ca^{+2} transient amplitude, (C): decay constant tCa (D) after AngII stimulation under Ctrl conditions and in the presence of FTY720 (E): Mean number of spontaneous Ca^{+2} transients in VT-AMs under indicated treatment conditions. (canine model)

Pak1 stimulation in atrial tissue could be a pharmacological tool to attenuate NOX2- dependent ROS production and prevent Ca^{+2} overload through the attenuation of NCX activity.

Discussion

- Regulation of Pak1 activity significantly influences the propensity for atrial arrhythmic triggers & can prevent cellular Ca^{+2} overload by suppressing the NOX2/ROS-dependent stimulation of NCX activity.
- The arrhythmias noted in Pak1 $^{-/-}$ mice are likely due to electrical changes, not unlike what is noted in patients with lone AF, specifically Pak1 $^{-/-}$ mice demonstrating frequent spontaneous PACs, consistent with the increased incidence of spontaneous Ca^{+2} release events.
- The study demonstrated on a cellular level that FTY720 may prevent and reverse AngII-induced NOX2/ ROS production in atrial myoctyes, and Pak1 stimulation may be a good target to reduce arrhythmic events and prevent remodeling.

Clinical Significance

Pak1 may serves as a pharmacological target to attenuate baseline and AngII-induced ROS production and arrhythmic activity.

Whether Pak1 signaling is able to limit AngII-induced fibrotic remodeling and AF substrate in humansformation remains to be seen.

Beyond the Length and Look of Repolarization: Defining the Non-QTc Electrocardiographic Profiles of Patients with Congenital Long QT Syndrome.

Lane, Conor M., et al. *Heart Rhythm*, vol. 15, no. 9, 2018,

Background

Although LQTS genetic testing has become a standard part of the evaluation of LQTS, a negative test result does not necessarily exclude disease with current tests accounting for approximately 80% of all LQTS

Furthermore, 25% of patients with genetically confirmed LQTS have a normal QT interval, which complicates the diagnostic process.

Aside from QT prolongation, the best described ECG features have all been associated with repolarization; such as T-wave alternans, T-wave morphology or rates of bradycardia and bradyarrhythmias.

Aim: To determine the complete ECG profile of patients with LQTS and to evaluate differences by age and genotype

Methods

Inclusion: Schwartz LQTS score ≥ 3.5, and/or had an unequivocally pathogenic mutation in 1 of the LQTS genes or had QTc ≥ 500 ms. (Fig 1)

Exclusion: Concomitant arrhythmic disorders, actively paced, multiple LQTS mutations, LQTS genotypes 4–15 & genotype-negative/phenotype-positive patients or those without genetic testing

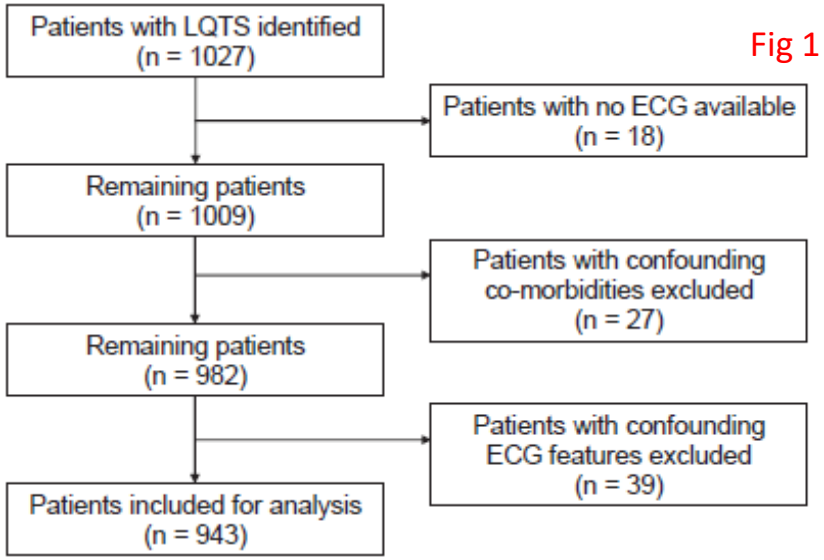
Abnormal QTc: QTc ≥ 460 ms if < 12 yrs old, QTc ≥ 470 ms in males > 12 yrs old, and QTc ≥ 480 ms in females > 12 yrs old.

Standard lead placement and recorded at 25 mm/s with a gain of 1 mV/ 10 mm

QT measurements were measured as the average of 3 consecutive beats in lead II or V5.

Patient selection flowchart, (1999-2017)

Fig 1



Baseline characteristics of the study cohort, n=934

Age (yrs)	25 (9–34)
Female	535 (57)
QTc (Bazett) (ms)	459 (439–483)
Symptomatic before diagnosis	205 (22)
Proband	332 (35)
Family history of LQTS	797 (85)
Family history of SCD	520 (55)
LQTS genetic subtypes	
LQT1	456 (48)
LQT2	303 (32)
LQT3	80 (8.5)
LQTS minor	48 (5.1)
LQTS multiple	36 (3.8)
LQTS G–P+	17 (1.8)
LQTS not tested	3 (0.3)
Treatment	
Beta-blocker	793 (84)
ICD	172 (18)
LCSD	124 (13)

Table 2 Selected ECG features of all LQTS patients and selected genotypes

	LQTS cohort (N = 943)	LQT1 (n = 456)	LQT2 (n = 303)	LQT3 (n = 80)	*P* value
Rhythm					
Sinus rhythm	679 (72)	336 (74)	223 (74)	56 (70)	NS
Sinus arrhythmia	246 (26)	115 (25)	75 (25)	17 (21)	NS
Ectopic atrial rhythm	14 (1.5)	3 (0.7)*	4 (1.3)	7 (8.8)*	<.0001
Junctional escape	3 (0.3)	1 (0.2)	1 (0.3)	0	NS
Atrial fibrillation	1 (0.1)	1 (0.2)	0	0	NS
Rate					
Bradycardia	320 (34)	161 (35)	90 (30)	26 (33)	NS
Tachycardia	5 (0.5)	2 (0.4)	2 (0.7)	1 (1.3)	NS
Abnormal QRS duration					
Prolonged	35 (3.7)	19 (4.2)	8 (2.6)	4 (5)	NS
QRS abnormalities					
Nonspecific IVCD	21 (2.2)	12 (2.6)	5 (1.7)	1 (1.3)	NS
Partial LBBB	7 (0.7)	5 (1.1)	1 (0.3)	1 (1.3)	NS
Partial RBBB	4 (0.4)	1 (0.2)	2 (0.7)	0	NS
RBBB	3 (0.3)	2 (0.4)	0	1 (1.3)	NS
LBBB	2 (0.2)	1 (0.2)	1 (0.3)	0	NS
LAFB	2 (0.2)	0	1 (0.3)	1 (1.3)	NS
LPFB	1 (0.1)	0	0	0	NS
T-wave inversion					
I	10 (1.1)	5 (1.1)	2 (0.7)	1 (1.3)	NS
II	30 (3.2)	7 (1.6)*	8 (2.7)	9 (11)*	<.0001
III	349 (37)	138 (31)*	123 (41)	48 (61)*	<.0001
aVR	922 (98)	448 (99)	297 (98)	75 (95)*	.04
aVL	94 (10)	52 (12)*	24 (8)	3 (3.8)	NS
aVF	107 (11)	35 (7.7)*	32 (11)	23 (29)*	<.0001
V_1	655 (70)	274 (61)*	249 (83)*	57 (72)	<.0001
V_2	373 (40)	143 (32)*	158 (52)*	30 (38)	<.0001
V_3	194 (21)	65 (14)*	88 (29)*	16 (20)	<.0001
V_4	76 (8.1)	27 (6)	30 (9.9)	6 (7.6)	NS
V_5	30 (3.2)	9 (2)*	12 (4)	5 (6.3)	NS
V_6	16 (1.7)	5 (1.1)	4 (1.3)	4 (5.1)*	.03
Notched T waves	257 (27)	70 (16)*	140 (46)*	20 (25)	<.0001
T-wave alternans	9 (1)	4 (0.9)	1 (0.3)	0	NS

Table 3 Significant differences in T-wave morphology by QTc value

	Normal QTc (n = 598)	Abnormal QTc (n = 340)	*P* value
TWI in lead I	2 (0.3)	8 (2.4)	.006
TWI in lead II	12 (2)	18 (5.3)	.01
TWI in lead III	205 (34)	144 (42)	.02
TWI in lead aVF	52 (8.7)	55 (16)	.0009
TWI in lead V_2	207 (35)	166 (49)	<.0001
TWI in lead V_3	91 (15)	103 (30)	<.0001
TWI in lead V_4	28 (4.7)	48 (14)	<.0001
TWI in lead V_5	12 (2)	18 (5.3)	.01
TWI in lead V_6	4 (0.7)	12 (3.5)	.001
T-wave alternans	1 (0.2)	8 (2.3)	.001
Notched T waves	139 (23)	120 (35)	.0002

Table 4 Significant differences in T-wave morphology by genotype

	LQT1 (n = 453)	Other genotypes (n = 381)	*P* value
Lead II	7 (1.6)	17 (4.5)	.02
Lead III	138 (31)	171 (45)	<.0001
Lead aVF	35 (7.7)	55 (14)	.002
Lead V_1	274 (61)	306 (80)	<.0001
Lead V_2	143 (32)	188 (49)	<.0001
Lead V_3	65 (14)	104 (27)	<.0001
Notched T waves	70 (16)	160 (42)	<.0001
	LQT2 (n = 302)	**Other genotypes (n = 532)**	***P* value**
Lead V_1	249 (83)	331 (62)	<.0001
Lead V_2	158 (52)	173 (33)	<.0001
Lead V_3	88 (29)	81 (15)	<.0001
Notched T waves	140 (46)	90 (17)	<.0001
	LQT3 (n = 79)	**Other genotypes (n = 755)**	***P* value**
Abnormal T-wave axis	15 (19)	42 (5.6)	.0001
Borderline abnormal QRS-T angle	10 (13)	37 (4.9)	.009
Lead II	9 (11)	15 (2)	.0002
Lead III	48 (61)	261 (35)	<.0001
Lead aVF	23 (29)	67 (8.9)	<.0001
Lead V_6	4 (5.1)	9 (1.2)	.03

Table 5 ECG features of LQTS patients by age (N = 943)

	<1 yr (n = 60)	1–4 yrs (n = 48)	4–10 yrs (n = 141)	10–16 yrs (n = 224)	16–25 yrs (n = 136)	25–40 yrs (n = 170)	>40 yrs (n = 164)	*P* value
Rhythm								
Sinus rhythm	58 (97)*	37 (77)	79 (56)*	128 (57)*	83 (61)	140 (82)*	154 (94)*	<.0001
Sinus arrhythmia	2 (3.3)*	10 (21)	61 (43)*	93 (42)*	46 (34)*	26 (15)*	8 (4.9)*	<.0001
Bradycardia	7 (12)*	13 (27)	19 (14)*	101 (45)*	65 (48)*	60 (35)	55 (34)	<.0001
PR interval								
Prolonged	4 (6.7)*	1 (2.1)	4 (2.8)	1 (0.4)	2 (1.5)	1 (0.6)	6 (3.7)	.03
P-wave abnormalities								
RAE	1 (1.7)	0	1 (0.7)	1 (0.5)	0	1 (0.6)	5 (3.1)*	.09
LAE	0	0	0	0	1 (0.8)	3 (1.8)	7 (4.3)*	.003
Early repolarization pattern	5 (8.5)	5 (10)	18 (13)	38 (17)	38 (28)*	22 (13)	13 (8.1)*	<.0001
T-wave inversion								
V_1	56 (95)*	46 (96)*	129 (92)*	166 (74)	82 (60)*	97 (57)*	79 (49)*	<.0001
V_2	54 (92)*	45 (94)*	111 (79)*	115 (51)*	19 (14)*	15 (8.9)*	14 (8.7)*	<.0001
V_3	36 (61)*	30 (63)*	56 (40)*	40 (18)	13 (9.6)*	10 (5.9)*	9 (5.6)*	<.0001
Notched T waves	15 (25)	16 (33)	67 (48)*	76 (34)*	34 (25)	30 (18)*	19 (12)*	<.0001

Discussion & Clinical Significance

Outside the QT interval, T-wave notching and sinus bradycardia, ECG abnormalities are uncommon in patients with LQTS; especially bundle branch block.

The genotype-specific features identified may help guide and interpret genetic testing, which can help initiate tailored therapies. The age-related ECG findings should help to provide better phenotype correlation based on age.

Patients with LQT1 have significantly less distortion of T-wave morphology & less TWI than other genotypes. TWI and abnormal T-wave axis were significant features of LQT3.

LQT2 patients showed more TWI in leads V1–V3 & t-wave notching, similar to ARVC, which could incur a potential for misdiagnosis and diagnostic delay.

Limitations

Single-center, retrospective study with all ECG analysis by a single reviewer

Selection bias has caused a lower incidence of AV block due to exclusion of paced patients

The inclusion of patients on beta-blockade limits the analysis of bradycardia

Section #4: Invasive EP
Chalese Richardson, MD

Implantation techniques and outcomes after cardiac resynchronization therapy for congenitally corrected transposition of the great arteries

Heart Rhythm Aug 2018; 1-8 https://doi.org/10.1016/j.hrthm.2018.08.017

Moore JP, Cho D, Lin JP, Lluri G, Reardon LC, Aboulhosn JA, Hageman A, Shannon KM

Background: While literature supports favorable outcomes for acute cardiac resynchronization therapy (CRT) in patients with congenital heart disease and a failing systemic right ventricles, there is limited data on CRT in patients with congenitally corrected transposition of the great arteries. CCTGA

Objective: (1) To show that patients with CCTGA are amenable to a CRT via a transvenous approach despite variant coronary sinus (CS anatomy) and (2) to determine clinical parameters that could predict CRT outcomes in these patients

Methods: Single institution, retrospective chart review of all CCTGA patients who underwent attempted CRT lead placement from March 2002 to March 2018. Indications for CRT included congestive heart failure (CHF) with intact AV conduction and QRS prolongation, cardiomyopathy 2nd to chronic LV pacing or complete AV block with anticipation of > 40% ventricular pacing. Baseline demographic, clinical (NYHA functional class, arrhythmia), echocardiographic, procedural (Final RV lead position) and preoperative ECG parameters were evaluated. For objective #1, coronary venous drainage was categorized and procedural success reported. For objective #2 the primary outcome measure was CRT failure defined as lack of acute response or return of NYHA class < 1 point from baseline. Secondary outcome measures included changes in QRS duration and rate of change, echo indices after CRT, final RV lead position and death/or transplant during follow up.

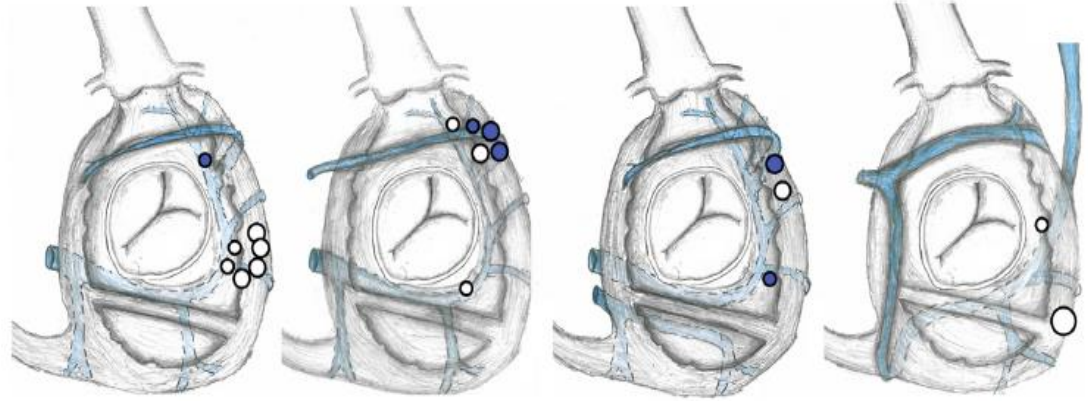


Figure 1 Variants of coronary venous anatomy in congenitally corrected transposition of the great arteries. The systemic right ventricle is shown in a modified left anterior oblique view, with the coronary sinus shaded in *blue*. For ease of comparison, both situs inversus and dextrocardic hearts are shown in their corresponding *l*-looped and levocardic positions. *Circles* represent the final position of the pacing cathode (*small*, basal position; *medium*, mid-ventricular position; *large*, apical position) for the 18 patients undergoing a successful transvenous approach. *Blue circles* represent patients who failed to respond to cardiac resynchronization therapy (n = 4) or who developed recurrent symptoms during follow-up (n = 2).

Results: 19/20 patients with CCTGA underwent transvenous lead placement for CRT. 18/19 were successfully placed (95%) with 12/18 placed via a posteroseptal CS ostium. Mean age 40 ±15. Indications for CRT was chronic LV pacing with ventricular dysfunction in 12, complete AV block with anticipation of >40% LV pacing in 5 and CHF with intact AV conduction and QRS prolongation in 3.

1º outcome: 12 patients had NYHA class ≥ 2 with 8 (67%) acute responders to CRT and 7/8 (88%) with minimal symptoms and median NYHA class 1.5 at median follow up of 4.6 years.

Implantation techniques and outcomes after cardiac resynchronization therapy for congenitally corrected transposition of the great arteries

Heart Rhythm Aug 2018; 1-8 https://doi.org/10.1016/j.hrthm.2018.08.017

Moore JP, Cho D, Lin JP, Lluri G, Reardon LC, Aboulhosn JA, Hageman A, Shannon KM

Results (continued): 2º outcomes: Median QRS duration decreased by 18 ms in all patients except those with complete AV block. Higher rate of change in QRS duration was associated with death or heart transplant (slope difference 1.9 ms/year; $P < 0.01$). When excluding patients with complete AV block, they noted a significant improvement in RV fractional area change on echo (22 to 27; P 0.048). Lead location at the RV free wall associated with freedom from CRT failure (Hazard ration 0.14; 95% CI 0.01-0.97). 3/4 patients listed for heart transplant at time of CRT were delisted due to improvement. 2 patients who were acute responders lost response, 1 due to infections of transvenous system with conversion to epicardial and the other due to refractory multifocal atrial tachycardia requiring AV node ablation. 4 total patients required revision of CRT system with repositioning in 2 for phrenic nerve capture and conversion in 2 others due to infection.

Conclusion: Implantation of transvenous CRT systems in patients with CCTGA is feasible despite variants in coronary sinus venous drainage. There also seems to be an acutely favorable response in a majority of patients (67%) however some patients will experience return of heart failure. QRS duration was likely to increase gradually overtime however patients with a more rapid rate of change experienced poorer outcomes.

Comments: Retrospective design with a small sample size (n=20). Although not statistically significant it appeared that CCTGA patients undergoing CRT for complete AV block did slightly worse after CRT with increases in QRS duration and less improvement in fractional area change on echocardiogram.

Percutaneous Transhepatic Venous Access for Atrial Tachyarrhythmia Ablation in Patients with Single Ventricle and Interrupted Inferior Vena Cava

Heart Rhythm October 2018 doi: 10.1016/j.hrcr.2018.10.005

Soto R, Jimenez A, Garcia F, Saenz, LC, Rodriguez DA

Background: In patients with interrupted inferior vena cava, a superior or retrograde aortic arterial approach can be used to gain access cardiac for ablations, however may present challenges with catheter stability and manipulation. Percutaneous transhepatic venous access (described in 1995 and initially used for patients with congenital heart disease [CHD] and limited venous access) may provide an alternative approach.

Case report: 41 year old woman with complex single ventricle anatomy of DILV/DOLV, transposition of the great arteries, a left sided interrupted IVC w/ azygous continuation into the SVC and direct drainage of the suprahepatic veins to the right atrium. She underwent initial PA banding at 1 month of life and a Kawashima at 3 years old. She had a persistent LSVC to the CS which was closed with an Amplatz device at age 30. Presented with palpitations and diagnosed with atrial tachycardia (AT) at 39 years of age refractory to antiarrhythmic therapies.

Access: Bilateral femoral vein access was obtained and a 9 French (F) intracardiac ultrasound catheter and 6F quadripolar electrophysiology catheter was placed posterior to the right atrium (RA) to visualize intracardiac structures and obtain far field atrial electrograms. Percutaneous transhepatic venous access was obtained with the assistance of both ultrasound and fluoroscopy. A 5F micropuncture kit with an 18-cm/21-gauge needle was inserted along the right costochondral space along the anterior axillary line and accessed the middle hepatic vein lumen. Intravascular access confirmed with contrast within the suprahepatic vein and RA. A 5F long sheath dilator was placed allowing for wire exchange followed by upsizing to a 8.5F bidirectional deflectable sheath (Agilis TM Nxt, Abbott).

EPS and Ablation: Voltage mapping was performed in the RA using a multipolar circular mapping catheter and EnSite velocity mapping system (Abott). Low bipolar voltage (<0.1 mV) was found on the posterolateral RA. The mapping catheter was exchanged for a 4-mm irrigated tip ablation catheter and intra-atrial reentrant tachycardia (IART) with a tachycardia cycle length (TCL) was induced with burst rapid atrial pacing at 270 ms. A double line of ablation along the suprahepatic tricuspid isthmus was place and terminated the tachycardia. The ablation catheter was exchanged for two 4F diagnostic catheters which confirmed bidirectional block across the ablation line however a second IART was induced with a TCL of 290 ms near the posterolateral RA scar. Radiofrequency (RF) ablation was performed along the longitudinal aspect connecting to the suprahepatic vein. The tachycardia changed with a TCL of 600 ms. The double catheter technique again confirmed bidirectional block across the ablation line and the third tachycardia was mapped near the coronary sinus ostium. This area was ablated with RF and no further tachycardia were provoked on postablation pacing maneuvers. Unfractionated heparin administered throughout case with goal activated clotting time 300-350 seconds.

Access removal: Protamine administered. Ablation catheter removed and the 8.5F steerable sheath was exchanged over a 0.035-inch x 150-cm guidewire for a 8F, 11 cm long vascular introducer. The vascular introducer was then slowly withdrawn with contrast injections to reveal the residual tract left by the steerable sheath. Again using ultrasound and fluoroscopy the guidewire was removed and a -0.038-inch, 3cm x 5-mm vascular Cook embolization coil was deployed and achieved total occlusion of the tract with no compromise in hepatic venous flow. No capsular hematoma or abdominal bleeding found on ultrasound, No arrhythmia recurrence at 12 month follow up.

Conclusion: Percutaneous transhepatic venous access may be a safe alternative to superior venous access in patients with complex CHD and interrupted IVC requiring ablation.

Comments: Although this case was performed safely, it is undeniably a riskier approach given the complication profile, including but limited to subcapsular liver hematoma, hemoperitoneum, cholangitis, liver abscess, transaminitis and hepatic vein thrombosis. It also adds to the complexity of the case with the requirement of an embolization coil within the residual tract.