

**PJRT (Pediatric EP Journal Review by Trainees)**

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# Flecainide Versus Digoxin for Fetal Supraventricular Tachycardia: Comparison of Two Drug Treatment Protocols

Heart Rhythm, September 2016, Volume 13, Issue 9:1913-9

Sridharan S, Sullivan I, Tomek V, Wolfenden J, Škovránek J, Yates R, Janoušek J, Dominguez TE, Marek J

**Background:** Fetal supraventricular tachycardia (SVT) is associated with fetal hydrops and risk of fetal demise. There is little data to support the specific treatment protocol that is likely to be the most effective.

**Objective:** To compare the effectiveness of 2 transplacental treatment protocols used in 2 fetal cardiac centers.

**Methods:** Dual-center, retrospective longitudinal cohort of 84 fetal SVT ( heart rate >180 bpm with 1:1 atrioventricular relationship).

## **Results:**

- Flecainide (300mg daily) resulted in higher treatment success rate (conversion to sinus or rate control ) compared to digoxin (97.1% vs 51.7%, P <0.001). The overall rate of conversion in SVT with short ventriculoatrial (VA) interval was higher than long VA SVT.
- Flecainide was particularly more effective in hydropic fetuses than digoxin (success rate of 100% vs 38.1%).

**Conclusion:** flecainide is superior to digoxin for control of fetal SVT, particularly in the presence of hydrops. No adverse fetal outcomes were attributed to flecainide. Outpatient maternal flecainide exposure was well tolerated and avoided unnecessary hospital admission.

**Comments:** This study is a retrospective analysis of 2 different treatment protocols of a small number of patients in 2 centers. The digoxin cohort had a higher proportion of hydrops and cardiac compromise at presentation, as well as younger maternal age. Nevertheless, the results still suggested flecainide superiority after subgroup analysis of hydropic fetuses while controlling for both factors.

# Competitive Sports Participation in Patients With Catecholaminergic Polymorphic Ventricular Tachycardia

## A Single Center's Early Experience

Journal of the American College of Cardiology: Clinical Electrophysiology, June 2016, Volume 2, Issue 3: 253 – 62  
Ostby S, Bos JM, Owen H, Wackel P, Cannon B, Ackerman M

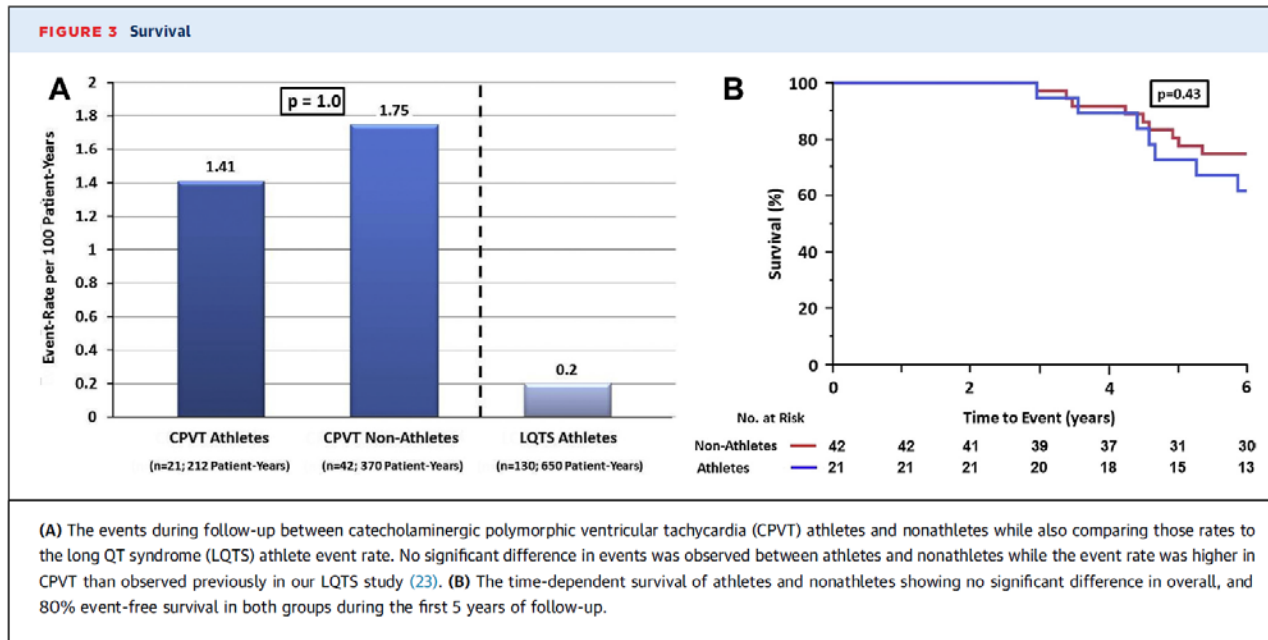
**Background:** Patients with Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) are at increased risk of sudden death with exertion. Based on expert opinion, current guidelines advise disqualification from nearly all sports.

**Objective:** To determine the outcome of continued sports participation in patients with CPVT

**Methods:** Retrospective record review on patients diagnosed with CPVT after 6 years of age. A well-informed decision for sports participation was shared between the patient/family and physician.

### **Results:**

- Among 63 CPVT patients (34 females, mean age at diagnosis  $16.6 \pm 12.9$  years, 53 had a positive genotype), 21 patients continued sports participation after treatment implementation.
- Prior to treatment implication, 76% of the athletic group had prior cardiac events vs 43% in the non-athletic group.
- 3 CPVT-triggered events occurred in the athlete group (14%) compared to 7 events in the non-athlete group (14%,  $P=1.00$ ). Following the triggered events, these patients were re-evaluated and their treatment was modified. There were no reported deaths.
- The 5 year event-free survival in both groups was 80%.



**Conclusion:** After correct diagnosis and implementation of a robust CPVT-directed treatment program, the likelihood of a nonlethal cardiac event in this study was the same whether the CPVT patient was an athlete or not.

**Comments:**

This study is a pilot retrospective review. The number of patients with positive genetics and a negative phenotype among the study group were not identified.

Overall, the prevalence of cardiac events on treatment in the study group is lower than those reported in other studies (14% vs 25-27%)<sup>1,2</sup>. Given the small number of patients and the known accumulative events over years in CPVT patients, larger studies with longer follow-up are needed to establish safety participation in CPVT patients.

1. Hayashi et al. Incidence and Risk Factors of Arrhythmic Events in Catecholaminergic Polymorphic Ventricular Tachycardia. *Circulation*. 2009;119:2426-2434
2. Roston et al. Catecholaminergic polymorphic ventricular tachycardia in children: analysis of therapeutic strategies and outcomes from an international multicenter registry. *Circ Arrhythm Electrophysiol*. 2015 Jun;8(3):633-42

# Cardiac Abnormalities in First-Degree Relatives of Unexplained Cardiac Arrest Victims

## A Report From the Cardiac Arrest Survivors With Preserved Ejection Fraction Registry

*Clinical Trial Registration*—URL: <http://www.clinicaltrials.gov>. Unique identifier: NCT00292032. (*Circ Arrhythm Electrophysiol.* 2016;9:e004274. DOI: 10.1161/CIRCEP.115.004274.)

**Background:** Unexplained cardiac arrest (UCA)/sudden unexplained death (SUD) may be caused by previously undiagnosed inherited arrhythmias. Screening of first degree relatives is recommended; however, the diagnostic yield varies between studies. It is unclear if the same diagnostic approach should be undertaken for family members of UCA survivors and SUD victims.

### **Objectives:**

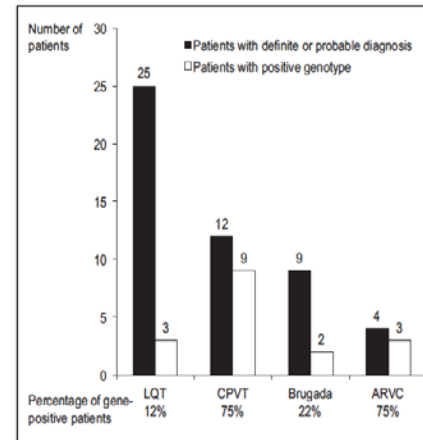
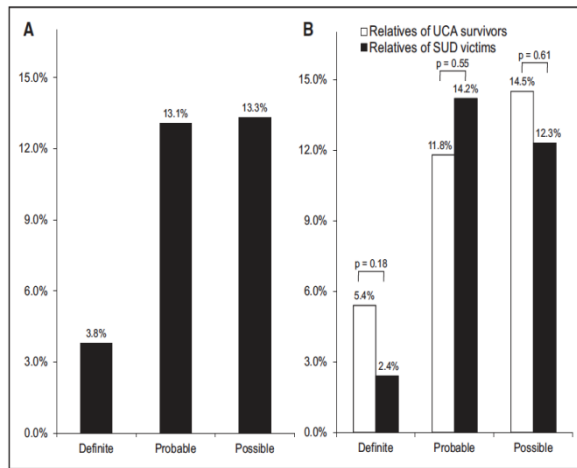
- To determine the prevalence of cardiac abnormalities in first degree family members of UCA/SUD victims
- To compare the diagnostic yield between first degree family members of UCA survivors and SUD victims

### **Methods:**

- CASPER registry captured 398 first degree family members (186 UCA, 212 SUD) who underwent extensive cardiac work-up including ECG, SAECG, Holter, exercise testing, cardiac imaging, provocative drug testing with Epi/procainamide. Genetic testing was phenotype-driven.
- Diagnostic strength was classified as definite, probable, or possible.

## Results:

- Cardiac abnormalities were detected in 30% of first degree relatives of UCA survivors and SUD victims, of which 17% had definite/probable diagnosis including LQTS (13%), CPVT (4%), ARVD (4%), and Brugada (3%).
- Diagnostic yield was similar for first degree relatives of UCA survivors and SUD victims (31% vs. 27%,  $p=0.03$ ).
- Disease-causing mutations were identified in 20/398 first degree relatives (5%), which was 20/96 (21%) of those tested. The most common pathogenic mutations being RyR2 (2%), SCN5A (1%), and KCNQ1 (0.8%).



## Conclusions:

- Systematic cascade screening and genetic testing in asymptomatic individuals will lead to preventative lifestyle and medical interventions with the potential to prevent sudden cardiac death

## Comments:

- Selection/referral bias from large tertiary centers may not reflect diagnostic yield in rural centers
- Overall percentage of genetic testing low, possibly contributing to a lower diagnostic yield
- Study did not address genetic yield per UCA/SUD victim (i.e. multiple family members per UCA/SUD proband)

# Characteristics of premature ventricular contractions in healthy children and their impact on left ventricular function

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[Heart Rhythm](#). 2016 Jul 5. pii: S1547-5271(16)30503-3. doi: 10.1016/j.hrthm.2016.07.002.  
[Epub ahead of print]

**Background:** There is paucity in the pediatric literature on the characteristics of PVCs in healthy children and their impact on LV function.

## **Objectives:**

- To assess the prevalence of LV systolic dysfunction in children with frequent PVCs (>10%) and to determine if it is associated with certain PVC characteristics
- Characterize evolution of PVCs over time

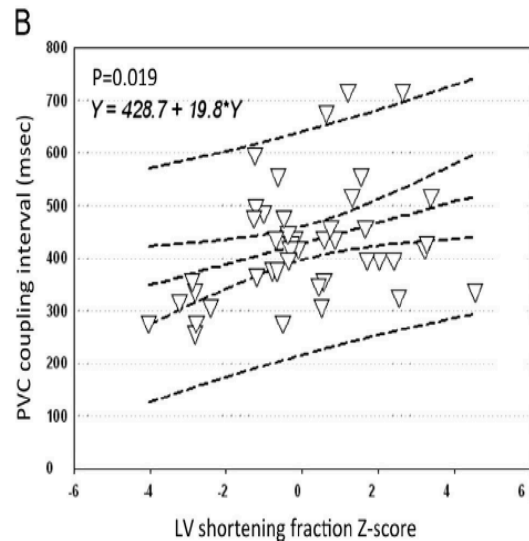
## **Methods:**

- Single center cohort study of children 18 years or younger with structurally normal hearts and no known inherited arrhythmias with a PVC burden  $\geq 10\%$  by a 24-hour Holter monitoring performed over a 5 year time period
- Clinical, arrhythmic, and echocardiographic data were reviewed at baseline and during follow up.
- LV systolic dysfunction was defined as LV SF z score  $< -2$

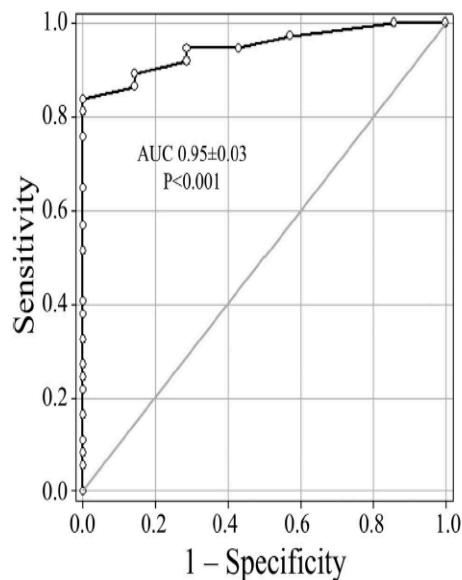


## Results:

- 47 children had a mean PVC burden of 20.9 +/- 11.9% at baseline
- PVC coupling interval averaged 430 +/- 110 msec with a PVC width of 118 +/- 27 msec
- PVCs were monomorphic in 44 patients (94%)
- 7 patients had LV dysfunction defined by LV SF z score < -2 (15%), but no patients had severe CMP
- Strong association between PVC coupling interval and LV dysfunction, with cutoff value of 365 msec having the greatest discriminatory value
- PVC proportion, width, and morphology were not significantly associated with development of LV dysfunction
- During 4.0 +/- 2.8 years of follow-up, the PVC burden decreased from median of 18% to 1.5%



**Figure 1** Association between proportion of PVCs (A) and PVC coupling interval (B) with left ventricular (LV) shortening fraction Z-score. The central straight line in each panel represents the simple linear regression fit to the data. Surrounding curved lines indicate 95% confidence intervals around estimated mean values. Upper and lower straight lines represent 95% prediction intervals. PVC = premature ventricular contraction.



**Figure 2** PVC coupling interval and left ventricular systolic dysfunction. The probability of having left ventricular shortening fraction Z-score < -2.0 as a function of PVC coupling interval is plotted in a receiver operating characteristic curve. Note the very strong association, as reflected by the area under the curve (AUC) (0.95 ± 0.03, P < .001). The diagonal line represents a line of no discrimination. PVC coupling interval < 365 ms yielded the greatest discriminatory potential (Youden J-statistic 0.72, sensitivity 85.7%, specificity 86.5%). PVC = premature ventricular contraction.

## Conclusions:

- PVCs in children with structurally normal hearts is rare and associated with a relatively benign course with spontaneous resolution in most.
- Mild LV systolic dysfunction is observed in 15%, which strongly correlated with a short PVC coupling interval < 365 msec

## Comments:

- Observational study with small sample size prone to type II error