# מעבר למיוקרדיטיס: גילוי גנטי מציל חיים באריתמיה בפעוטות

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## oresentation

# 15-month vaccinate

previously health

cya unre

The mother

had started CPR.

ents

## In Emergency Room..

Conscious, crying, then went unresponsive again



## Initiation of VF and shock #1







## Shock #2







### **ECG on Admission**

#### Sinus Tachycardia 161bpm and RBBB



# **Admission Course**

- Intubated and ventilated
- WBC=2300 Trop T=150
- Echo=normal LV and RV function and no structural abnormality
- Adenovirus positive

## **Diagnosis: Myocarditis**

- Mild neurologic deficit
- Seizures Started levetiracetam (Keppra) resloved

### **ECG on Admission**

#### Sinus tachycardia 161bpm RBBB with Coved ST elevation in V1



#### **RBBB and 4-5mm coved ST elevation**



#### **ECG 4 Days Later** RBBB and minimal ST elevation



## **RBBB and 2mm ST elevation in lead V1**



## **Genetic test results**

LIKELY PATHOGENIC

סיווג

SCN5A:c.273+1G>T chr3-38674525 C>A NM\_000335.5

# SGN5/A





# SCN5A:c.273+1G>T

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## **Splice Site Mutations**



# Brugada Syndrome in the Pediatric Population



#### Arrhythmic Event in Brugada Pediatric Population is Rare!



Milman et al; Age of First Arrhythmic Event in Brugada Syndrome *Circ Arrhythm Electrophysiol.* 2017;10:e005222. DOI: 10.1161/CIRCEP.117.005222

#### The Rate of Recurrent Arrhythmic Event is Higher at age<12yo than 13-20yo



## **Risk Factors for Recurrent AE**

#### TABLE 3 Risk Factors for Recurrent AE

		Event Rate With Variable*	Event Rate Without Variable†	HR	95% CI	p Value
/	Age ≤12 yrs					
	SND	7/7 (100)	7/14 (50)	3.3	1.1-9.7	0.03
	Atrial arrhythmias	9/9 (100)	6/13 (46)	3.0	1.07-8.70	0.04
	IVCD	9/9 (100)	6/13 (46)	3.4	1.2-9.8	0.02
	Large S in ECG lead I	6/6 (100)	8/14 (57)	3.1	1.04-9.30	0.04
	Either vs. none of the above	12/12 (100)	3/10 (30)	6.9	1.9-25.2	0.004
1	Age 13-20 yrs					
	SCN5A mutation	8/9 (89)	3/8 (38)	5.0	1.03-23.80	0.045

\*Number of patients with the variable and recurrent AE/total number of patients with the variable (%). †Number of patients without the variable and recurrent AE/total number of patients without the variable (%).

 $CI = confidence interval; HR = hazard ratio; IVCD = intraventricular conduction delay (QRS <math>\geq$ 110 ms); SND = sinus node dysfunction.

#### Patients with Symptoms + Spontaneous Type Brugada are at Higher Risk for Arrhythmic Events



## **Quinidine or ICD??**



#### **ICD complications**

Serious ICD-related complications occurred in 9 of 22 patients (41%), including lead failures (n = 4), inappropriate shocks (n = 4), endocarditis with reimplantation (n = 2), and hemothorax (n = 1). Two (9%) patients had both lead failure and inappropriate shocks or endocarditis. All complications were related to endocardial devices except the hemothorax, which appeared in the early postoperative period after epicardial ventricular lead implantation.

Andorin et al New Data on Brugada Syndrome in the Young Heart Rhythm, Vol 13, No 6, June 2016

#### ABSTRACT

**BACKGROUND** Young patients presenting with symptomatic Brugada syndrome have very high risks for ventricular arrhythmias and should be carefully considered for implantable cardioverter-defibrillator (ICD) placement. However, this therapy is associated with high rates of inappropriate shocks and device-related complications.

**OBJECTIVES** This study investigated clinical features, management, and long-term follow-up of young patients with Brugada syndrome and ICD.

**METHODS** Patients diagnosed with Brugada syndrome, who underwent implantation of an ICD at an age of  $\leq$ 20 years, were studied.

**RESULTS** The study included 35 consecutive patients. The mean age at ICD placement was  $13.9 \pm 6.2$  years. Ninety-two percent were symptomatic; 29% presented with aborted sudden cardiac death and 63% with syncope. During a mean follow-up period of 88 months, sustained ventricular arrhythmias were treated by the ICD in 9 patients (26%), including shocks in 8 patients (23%) and antitachycardia pacing in 1 patient (3%). Three patients (9%) died in an electrical storm. Seven patients (20%) experienced inappropriate shocks, and 5 patients (14%) had device-related complications. Aborted sudden cardiac death and spontaneous type I electrocardiogram were identified as independent predictors of appropriate shock occurrence.

**CONCLUSIONS** ICD therapy is an effective strategy in young patients with symptomatic Brugada syndrome, treating potentially lethal arrhythmias in >25% of patients during follow-up. Appropriate shocks were significantly associated with previously aborted sudden cardiac death and spontaneous type I electrocardiograms. However, ICDs are frequently associated with complications and inappropriate shocks, both of which remain high regardless of careful device implantation and programming. (J Am Coll Cardiol 2018;71:148-57) © 2018 by the American College of Cardiology Foundation.

# Conclusions

- Fever and ventricular fibrillation in a baby is NOT always myocarditis
- When seizures are present post ventricular fibrillation consider arrhythmia as the cause
- Consider in-hospital genetic testing and results on "emergency basis"
- Pediatric population with Brugada is at higher risk for recurrent arrhythmic events
- Quinidine may not prevent arrhythmia completely in the pediatric population, thus an ICD should be implanted.

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## Case

- 29yo, previously healthy
- Admitted for Ajmaline test due to family Hx of Brugada.
- A father of a boy who had CA at 15mo

LIKELY PATHOGENIC

SCN5A:c.273+1G>T chr3-38674525 C>A NM\_000335.5

## Baseline ECG 1



## **Baseline ECG 2 High leads**



## Ajmaline baseline ECG



## Ajmaline 50mg



## Ajmaline 60mg



#### Monomorphic VT originating from LV inferior wall



## **Atrial fibrillation post resuscitation**



## ECG 1 day post Ajmaline test



## ECG 2 day post Ajmaline test



## ECG 3 day post Ajmaline test

