

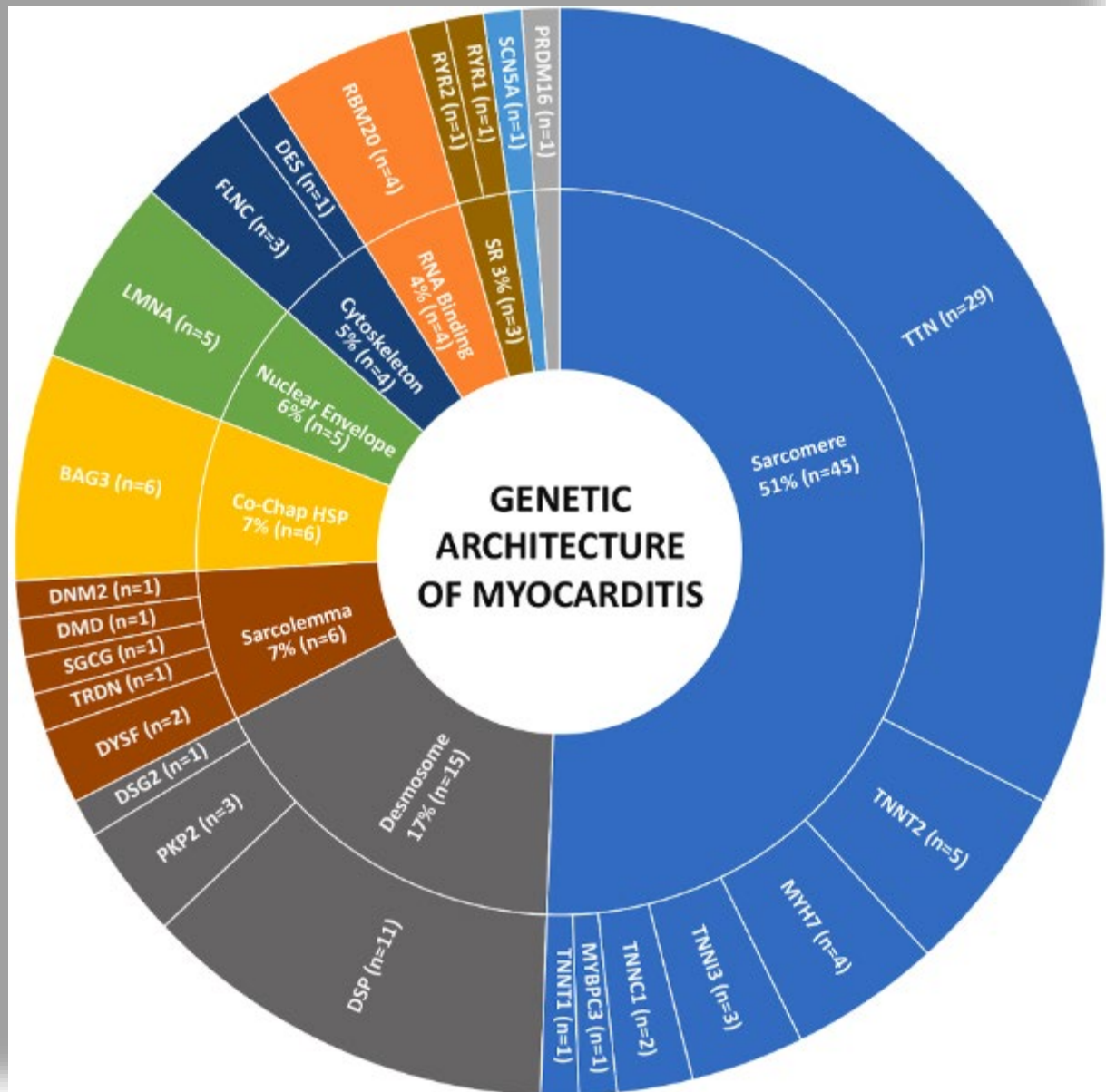
מעבר למיוקרדיטיס: גילוי גנטי מציל חיים באריתמיה בפעוטות

אילת שאואר, סמדר הורביץ, ברנרד בלאסן,

הפרעות קצב תורשתיות

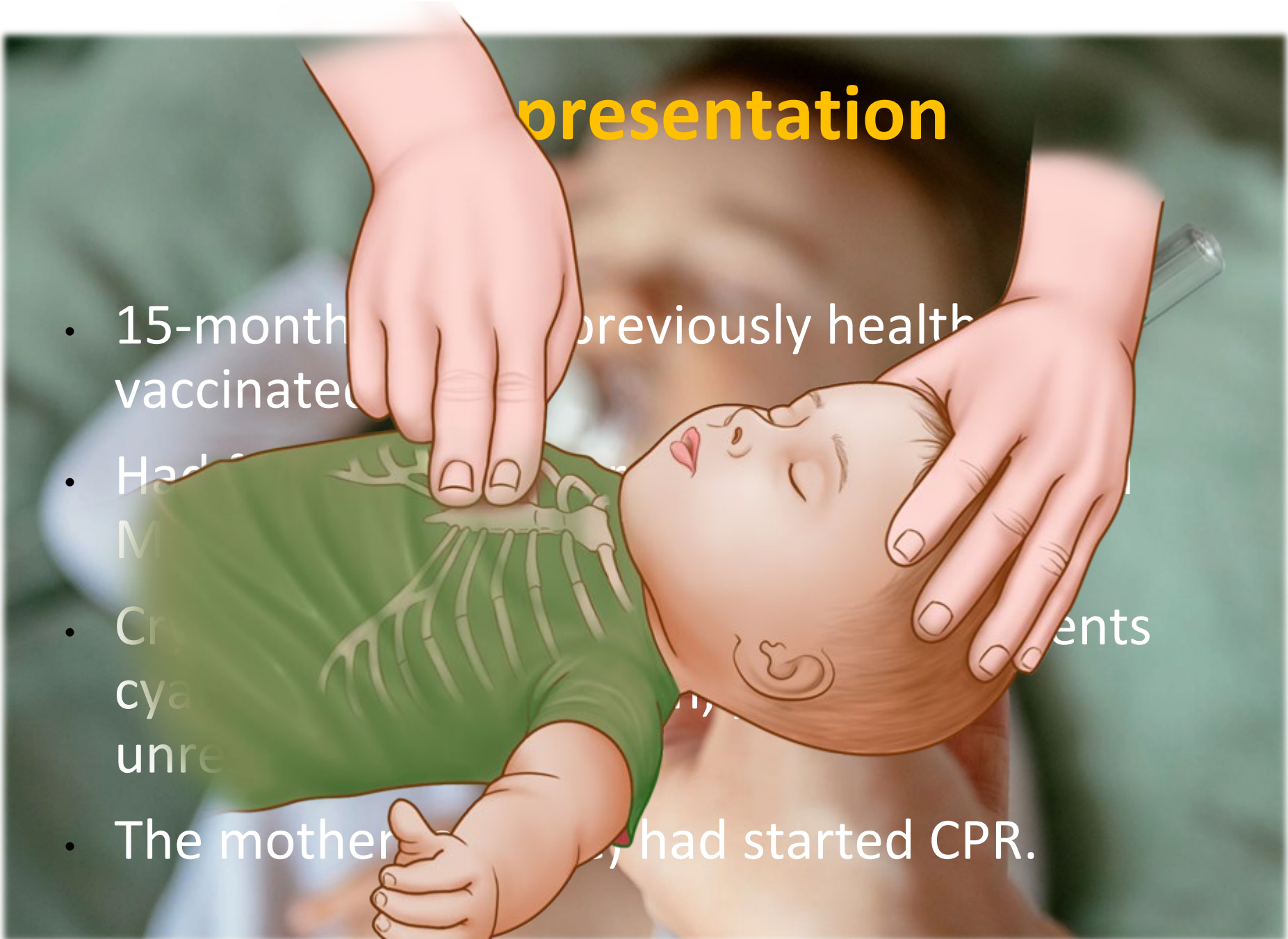
מערך הלב הדסה





presentation

- 15-month-old, previously healthy, unvaccinated child
- Had a seizure at home
- Mother brought child to the ED
- Child was unresponsive
- The mother had started CPR.

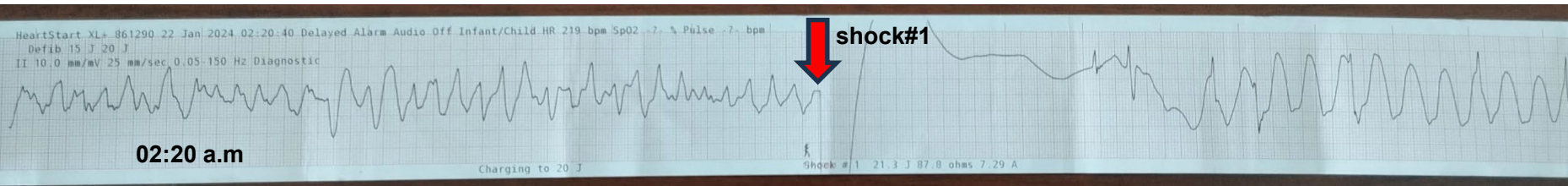
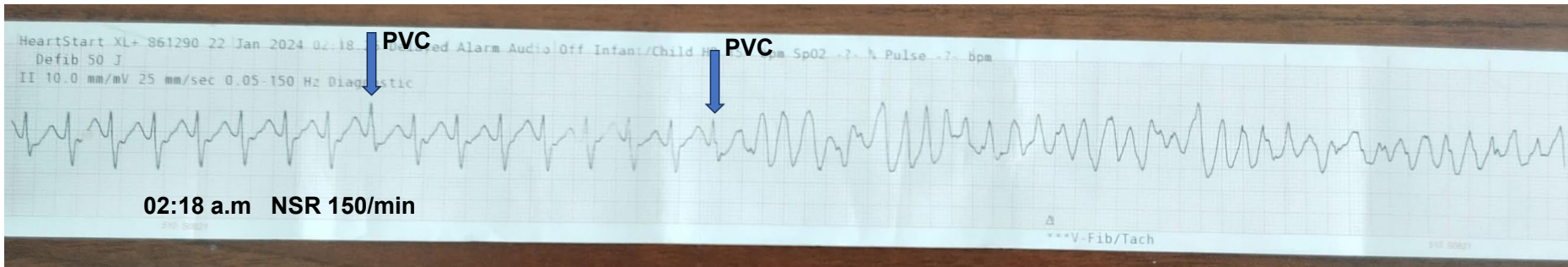
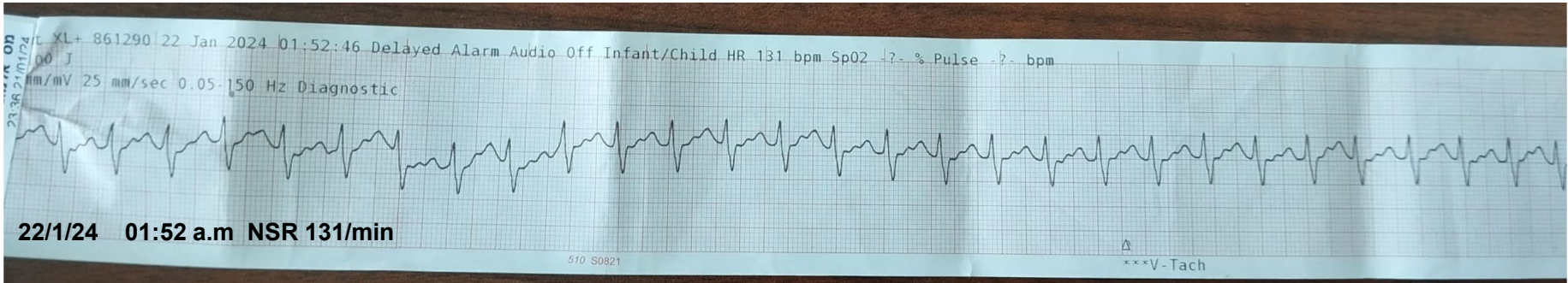


In Emergency Room..

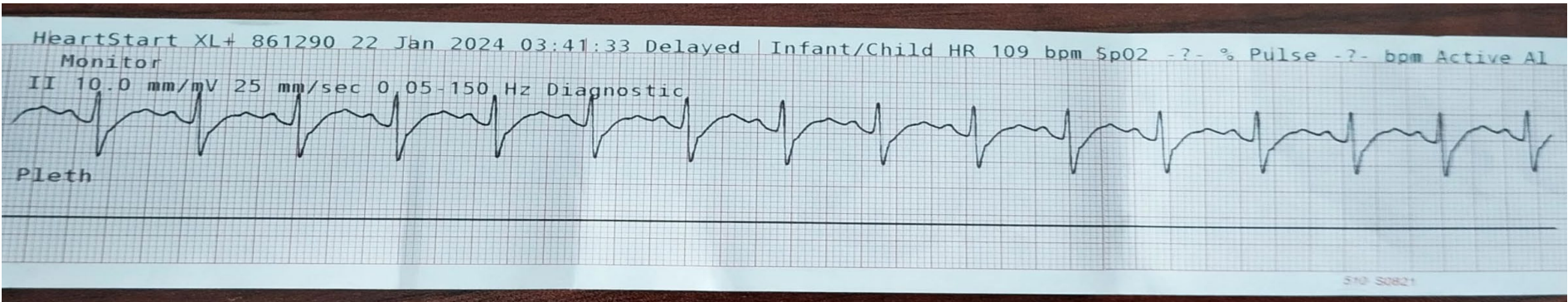
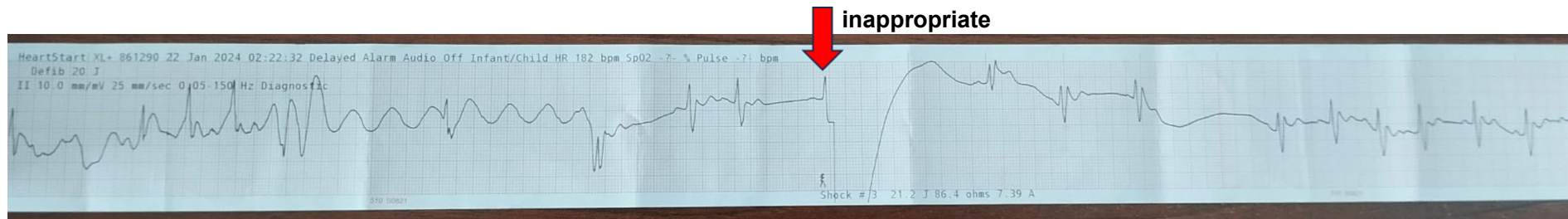
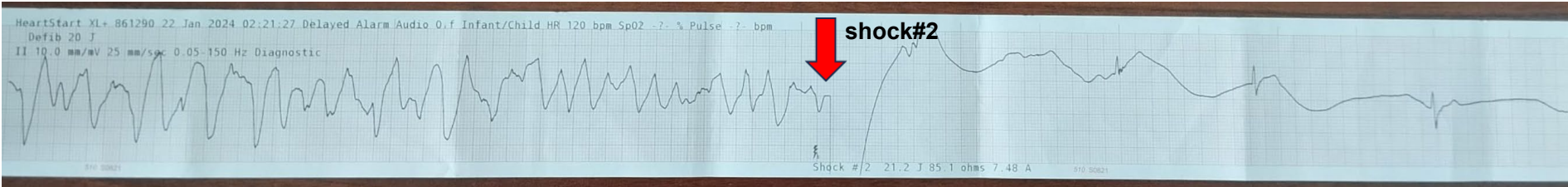
- Conscious, crying, then went unresponsive again



Initiation of VF and shock #1



Shock #2



ECG on Admission

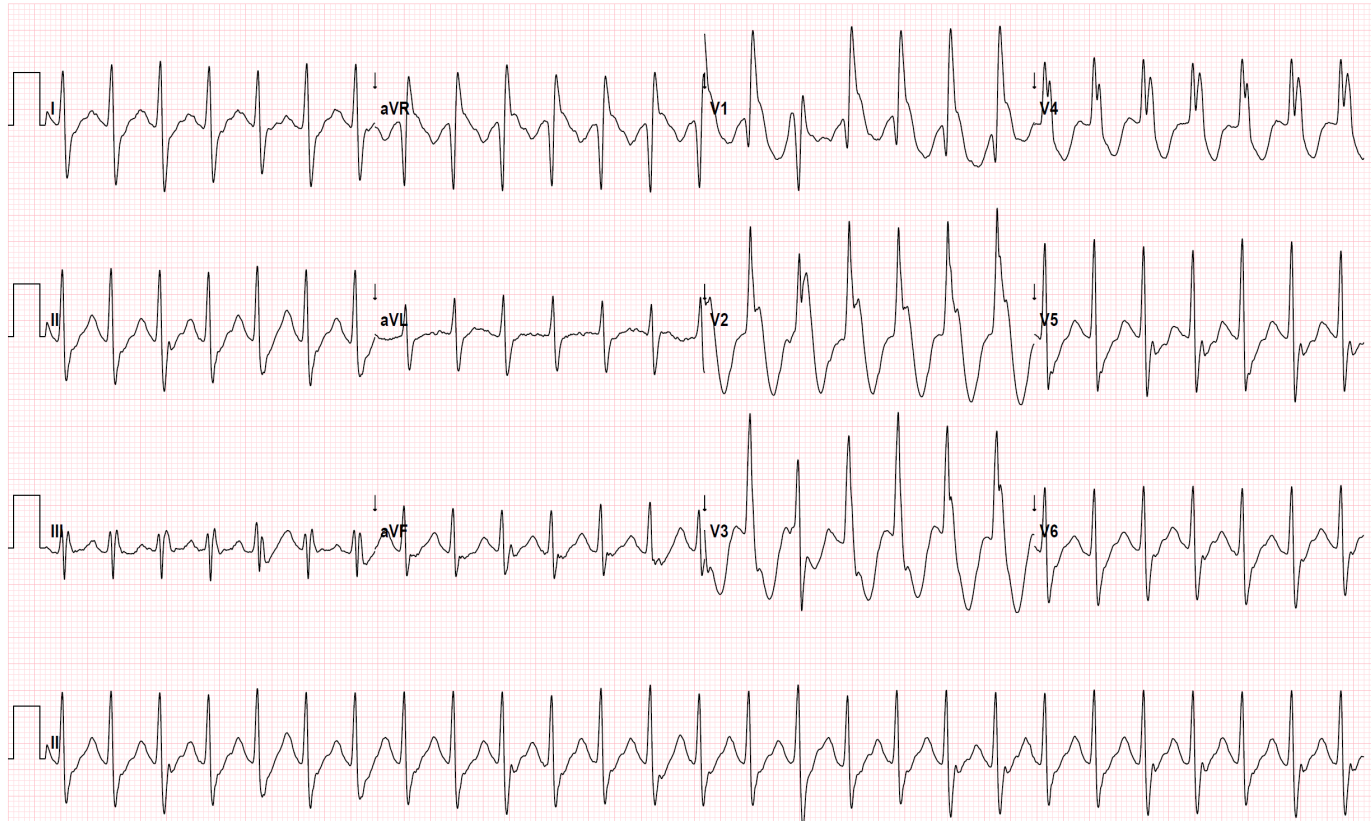
Sinus Tachycardia 161bpm and RBBB

21-Jan-2024 23:59:15

Vent rate: 161 BPM
PR int: * ms
QRS dur: 127 ms
QT/QTc: 293 / 382 ms
P-R-T axes: * 25 58
Avg RR: 372 ms
QTcB: 480 ms
QTcF: 407 ms

..PEDIATRIC ECG INTERPRETATION
UNCERTAIN IRREGULAR RHYTHM
RIGHT BUNDLE BRANCH BLOCK [QRS >= 110ms, no S IN V1, 1-15yr]
ABNORMAL ECG

UNCONFIRMED REPORT



119080001535

SITE 1

Site # 1 Cart # 11 ELI Link 5.0.0.3 Sequence # 06382 25mm/s 10mm/mV 0.05-40 Hz W

Admission Course

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

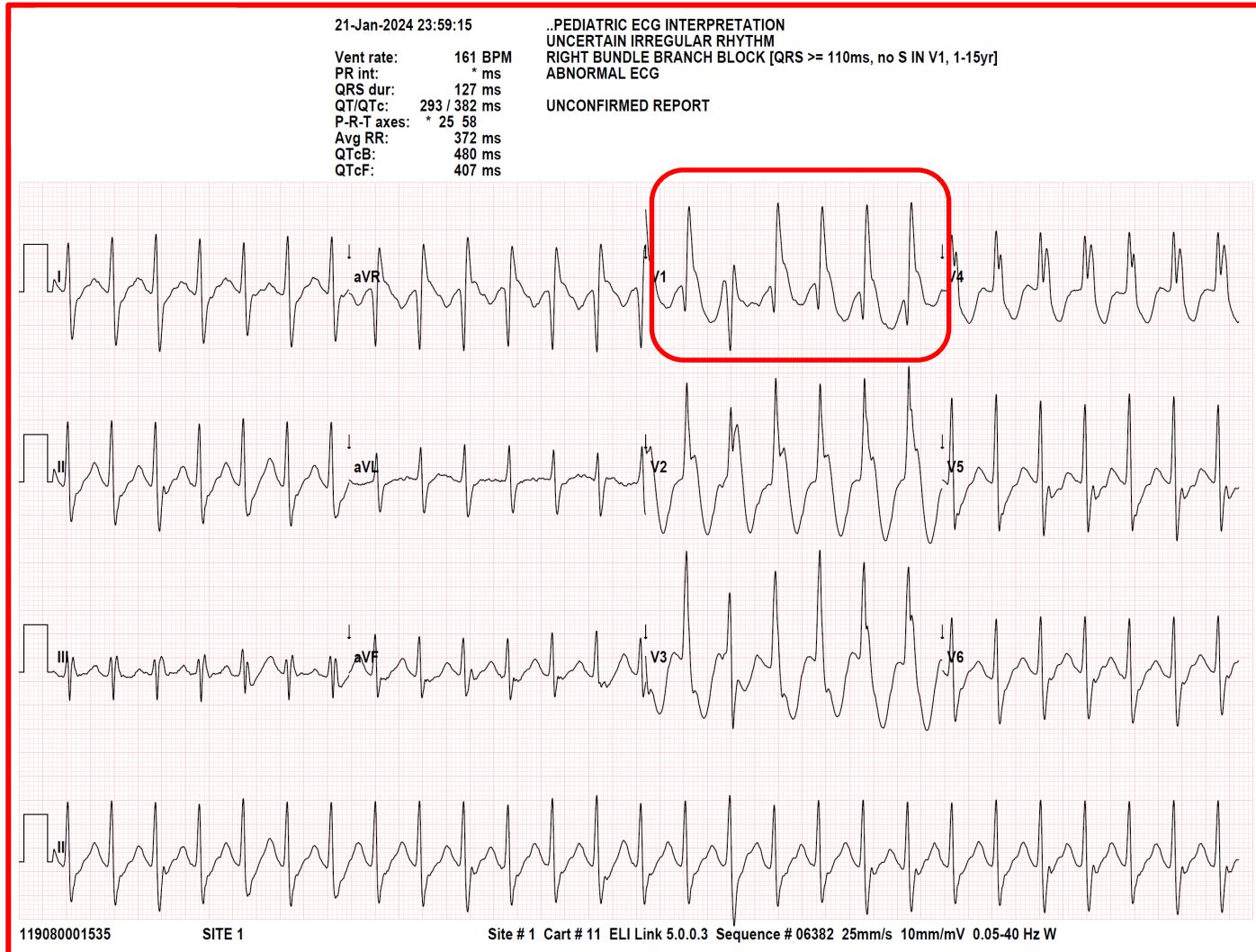
Diagnosis: Myocarditis

- Mild neurologic deficit
- Seizures – Started levetiracetam (Keppra) - resolved

ECG on Admission

Sinus tachycardia 161bpm

RBBB with Coved ST elevation in V1

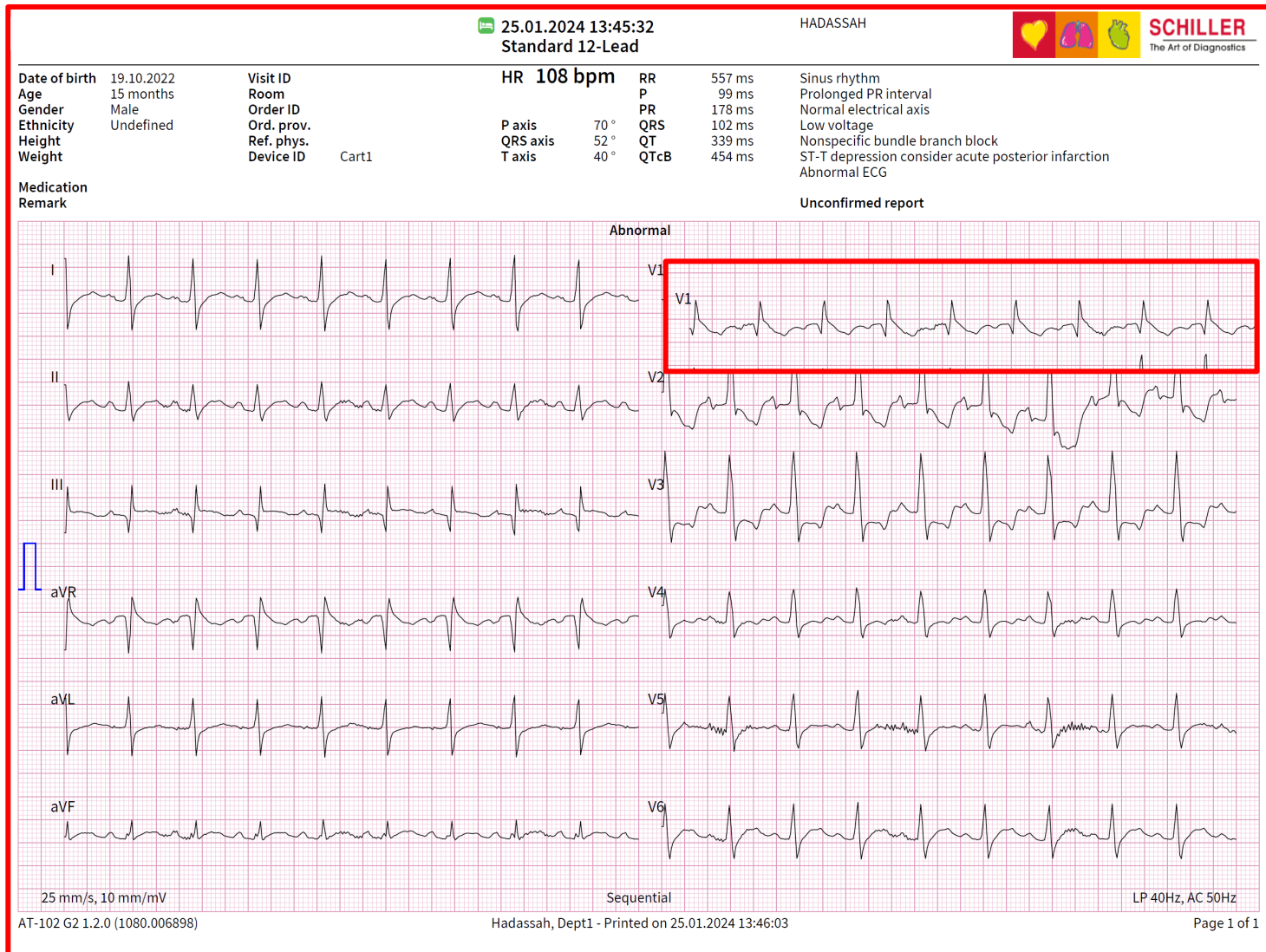


RBBB and 4-5mm covered ST elevation



ECG 4 Days Later

RBBB and minimal ST elevation



RBBB and 2mm ST elevation in lead V1



Genetic test results

LIKELY PATHOGENIC

סינוג

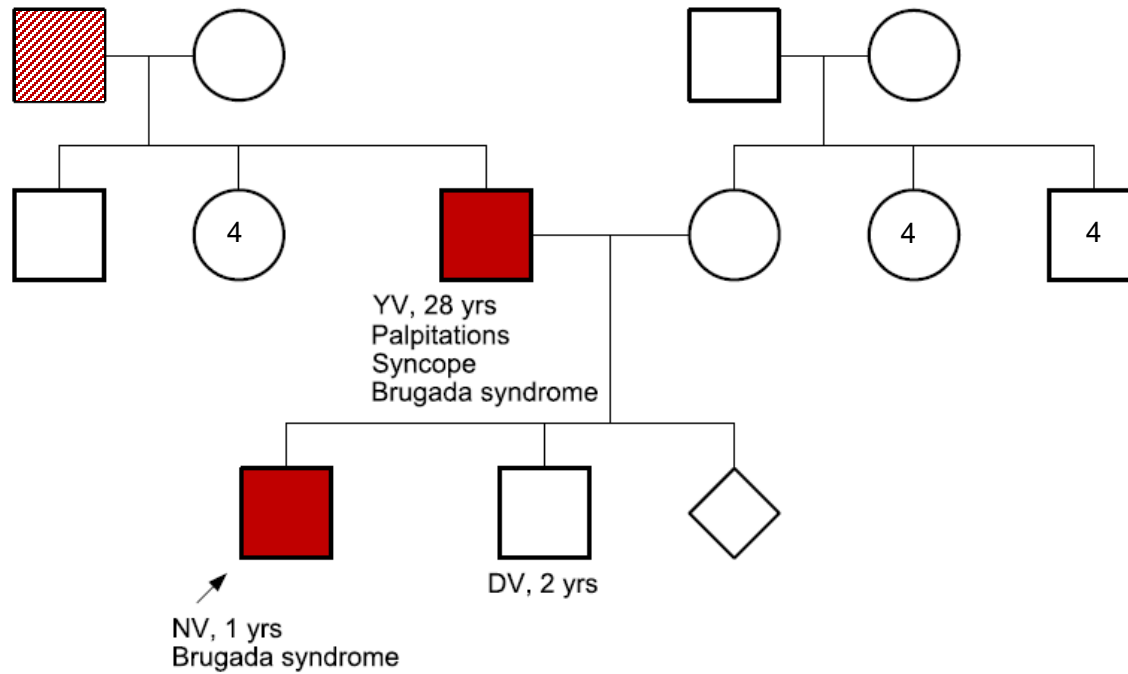
SCN5A:c.273+1G>T

chr3-38674525 C>A NM_000335.5

SCN5A



Family Tree

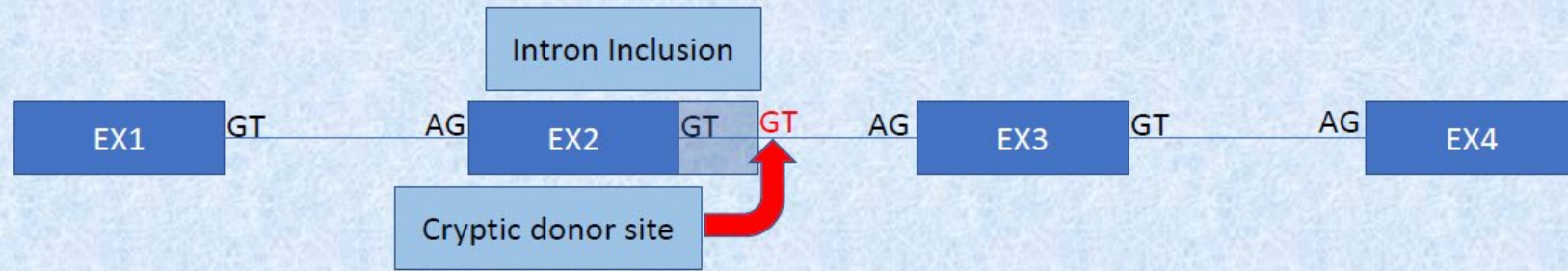
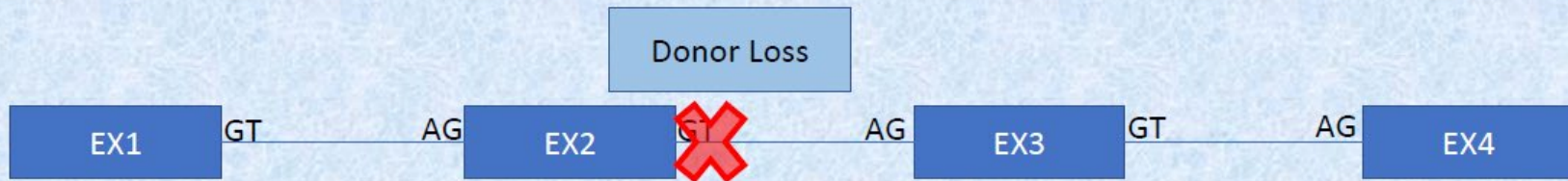
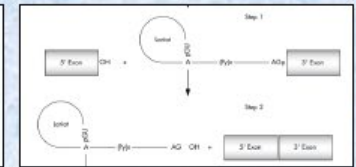
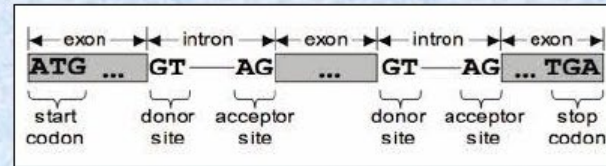


LEGEND

■ Brugada syndrome

Splice Site Mutations

Alternative Splicing - Types

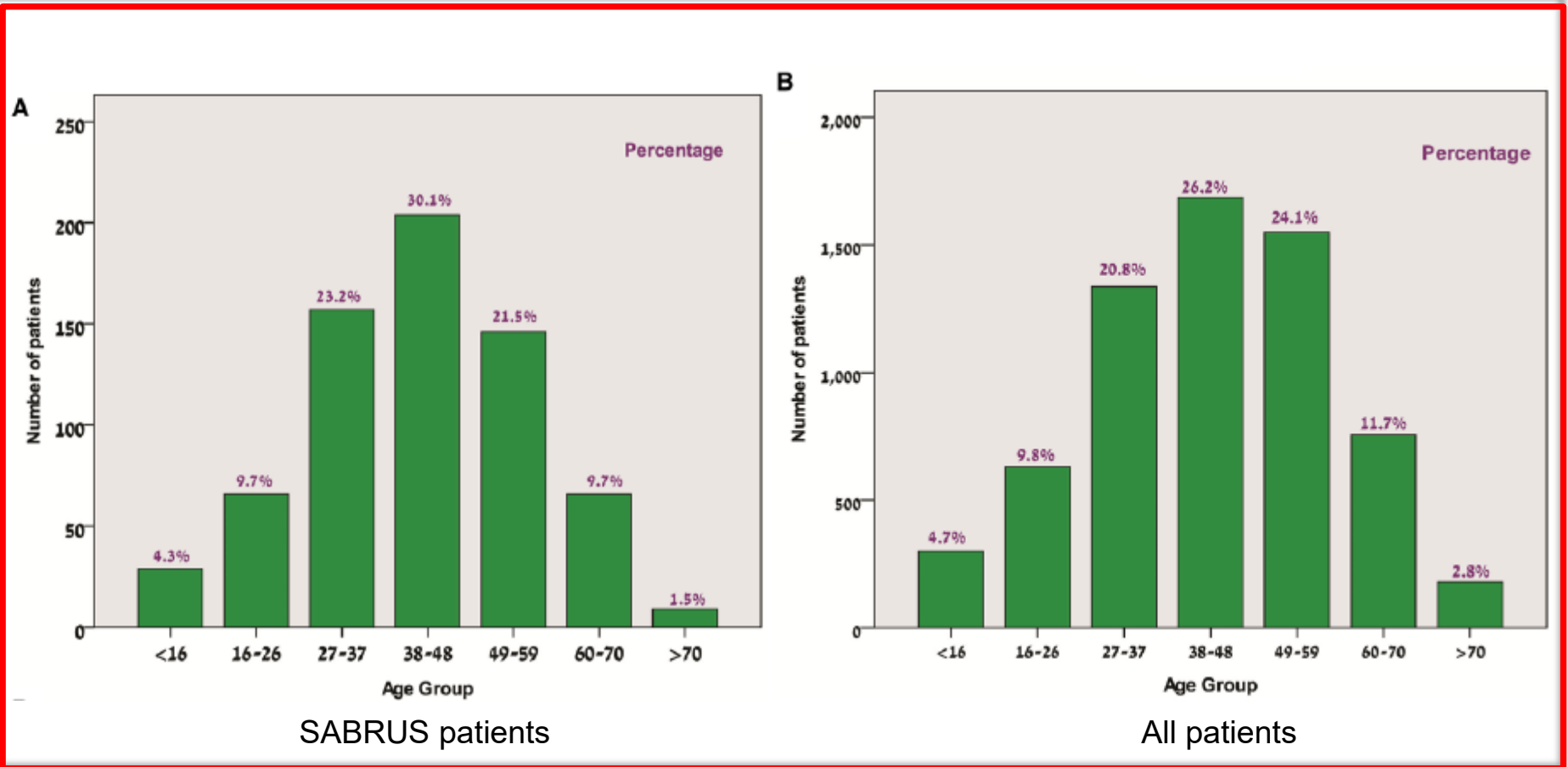


Byrne CD, Schwartz K, Lawn RM. Loss of a splice donor site at a 'skipped exon' in a gene homologous to apolipoprotein(a) leads to an mRNA encoding a protein consisting of a single kringle domain. *Arterioscler Thromb Vasc Biol.* 1995 Jan;15(1):65-70. doi: 10.1161/01.atv.15.1.65. PMID: 7749817

Brugada Syndrome in the Pediatric Population



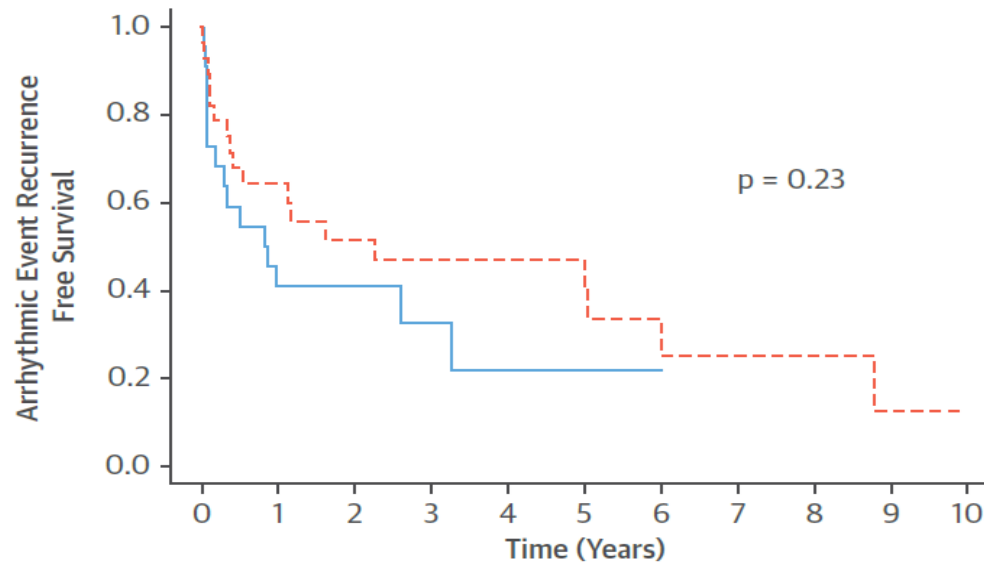
Arrhythmic Event in Brugada Pediatric Population is Rare!



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

FIGURE 1 Kaplan-Meier Curve for Recurrent Arrhythmic Events Among Pediatric and Adolescent Patients



Number at risk:

Pediatric	22	9	7	4	2	2	1	0	0	0	0
Adolescents	28	17	12	10	9	7	4	3	3	1	1

— Pediatric - - - Adolescents

In both age groups, a high recurrence arrhythmic event rate (59.1% and 35.7% at 12 months, respectively; $p = 0.27$) was observed with median time to recurrence of 9.9 months and 27.2 months, respectively.

Risk Factors for Recurrent AE

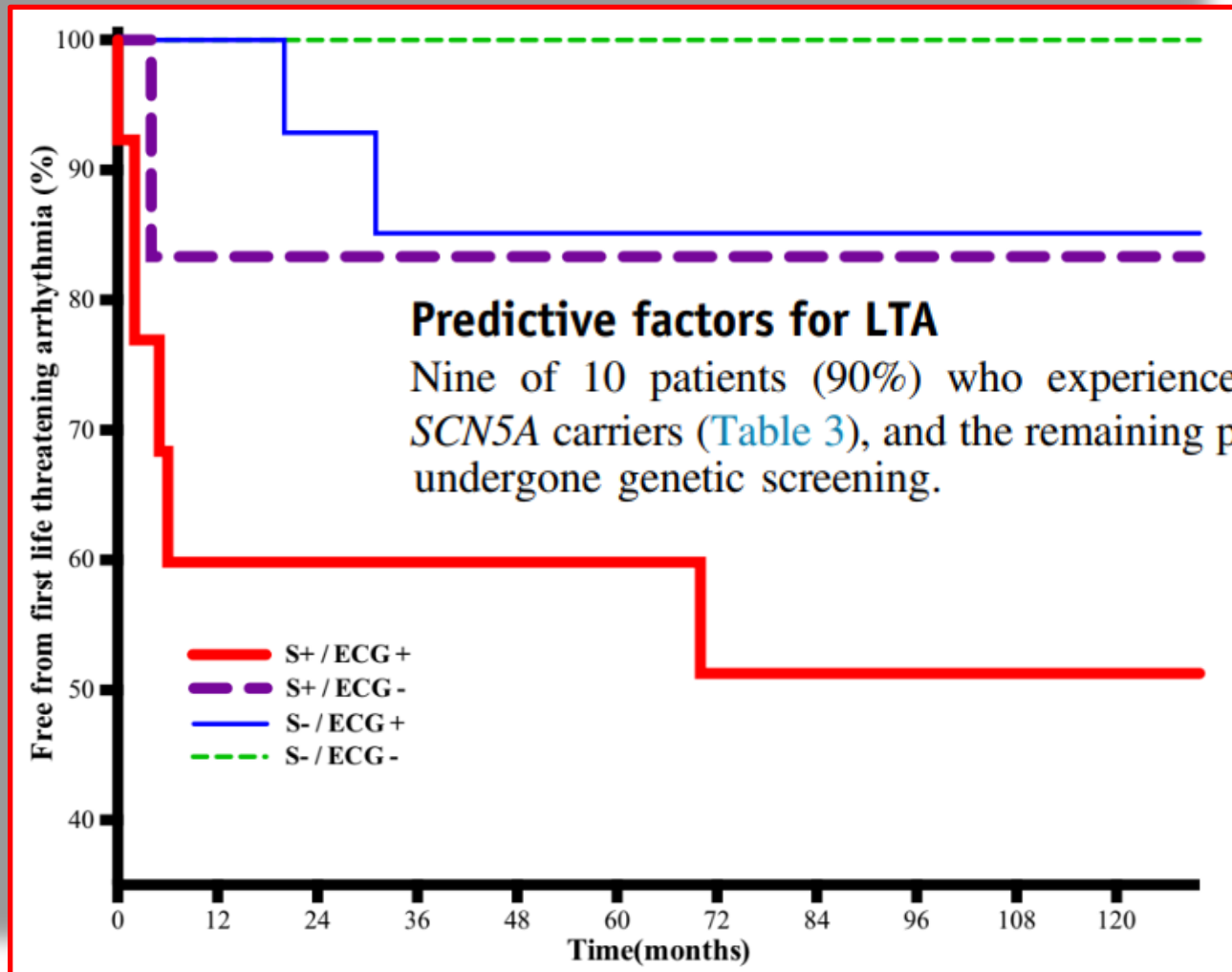
TABLE 3 Risk Factors for Recurrent AE

	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
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 ≥110 ms); SND = sinus node dysfunction.

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



Quinidine or ICD??



Conclusions

- Fever and ventricular fibrillation in a baby is NOT always myocarditis
- When seizures are present post ventricular fibrillation consider arrhythmia as the cause
- EP/inherited arrhythmia specialists consultation
- Consider in-hospital genetic testing and results on “emergency basis”

THINK

OUTSIDE

THE

