Congenital ↑QT_c Syndrome (Channelopathy)

How?

3 Major $LQTS_{1,2,3}$ Mutations

↓

Impaired $Ion_{K>>Na}$ Channels

↓

↑Action Potential (↑QT_c), Repol Abn

↓

Early Afterdepolarizations

- ↓K, Mg, Ca ↓ • ↑Sympa
- Meds ↓ -Exercise Stress
  -Acute Arousal

(@jackpenner Acquired $LQTS_c$)

⭐Polymorphic VT (Tdp)⭐

Who?

- >1 in 2,000
- Penetrance Variable, Low As 10%
- Dx by 30 yo
- Family History

What?

- Majority Without Sx @ Dx And Life
- Triggered Syncope +/- Seizure
- Incidental ↑QT_c
- Sudden Cardiac Arrest
- Rarely 1st Event SCD
- ↑Postpartum ♡ Events

Arrhythmia?

- PVT/T_dP (Most Common)
  - -Typically Short Lived
  - -Progress Syncope +/- V.Fib
- AV Block (~5%)

Associations?

- SN Deafness (JLN Syndrome)
- Hypokalemic Periodic Paralysis

Dx:

- ↑QT_c → Genetics
- Ekg in Family

Rx:

- Avoid QT_c Meds
- Asymptomatic vs Symptomatic (Type)

Menu

- -Avoid ↑Sympa
- -B-blocker
- -ICD

Edu:

- Many w/ Wrong Dx of Seizure
- Ekg After 1st Seizure
- AEDs w/ Na Channel Blocking ↑SCD
- ICD NOT Needed in Most

Dx
The Clinical Problem Solvers