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Family Evaluation After Sudden Death

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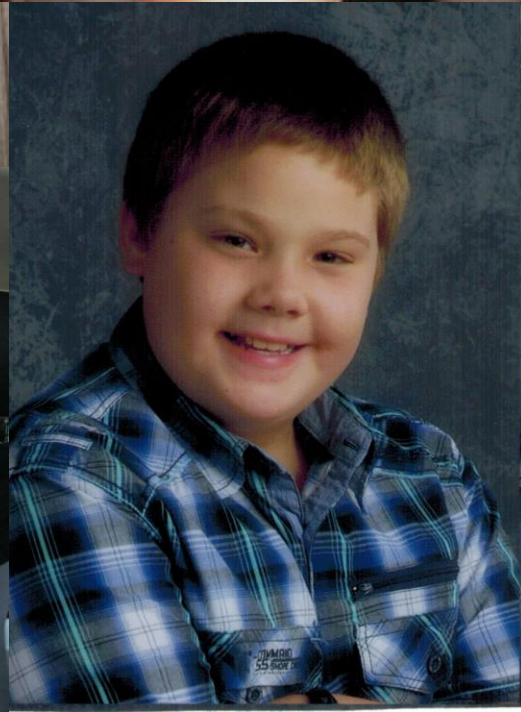
Conflict of Interest

- Consulting: Medtronic
- Research: Boston Scientific, Medtronic
- People of BC: Clinical Income
- Families of Sudden Death Victims: Paul Brunes
Chair in Heart Rhythm Disorders

I am an open access person – for slides, e mail akrahn@mail.ubc.ca
(I will repeat that at the end)

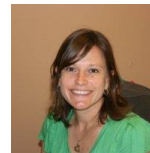
Families We Have to Meet







Storm Trackers



Family Member with Cardiac Arrest



- Seattle – 2.7 fold increased risk of SCD if a 1st degree relative had SCD before 65, after adjusting for CV risk factors (Circulation 1998;97:155)
- Paris study – parental SCD increased risk 80%, both parents 9-fold (Atherosclerosis 2002;162:211)
- Finnish autopsy study – VF with MI increased risk of family member with SCD compared to no VF (Circulation 2006;114:1462)
- Dutch – VF with MI – familial OR 2.7 (Circulation 2006;114:1140)

Why is the Cause of Sudden Death not Always Apparent?

Tragedy strikes

- Coroner - inquiry and autopsy
- May consult special pathologist
- Very small structural things can be difficult to detect
- Wires are no longer active (no reanimation)



Cardiac Arrest or Sudden Death? Its Luck (or lack thereof)

“the difference between cardiac arrest and sudden death is often 5 minutes by ambulance”



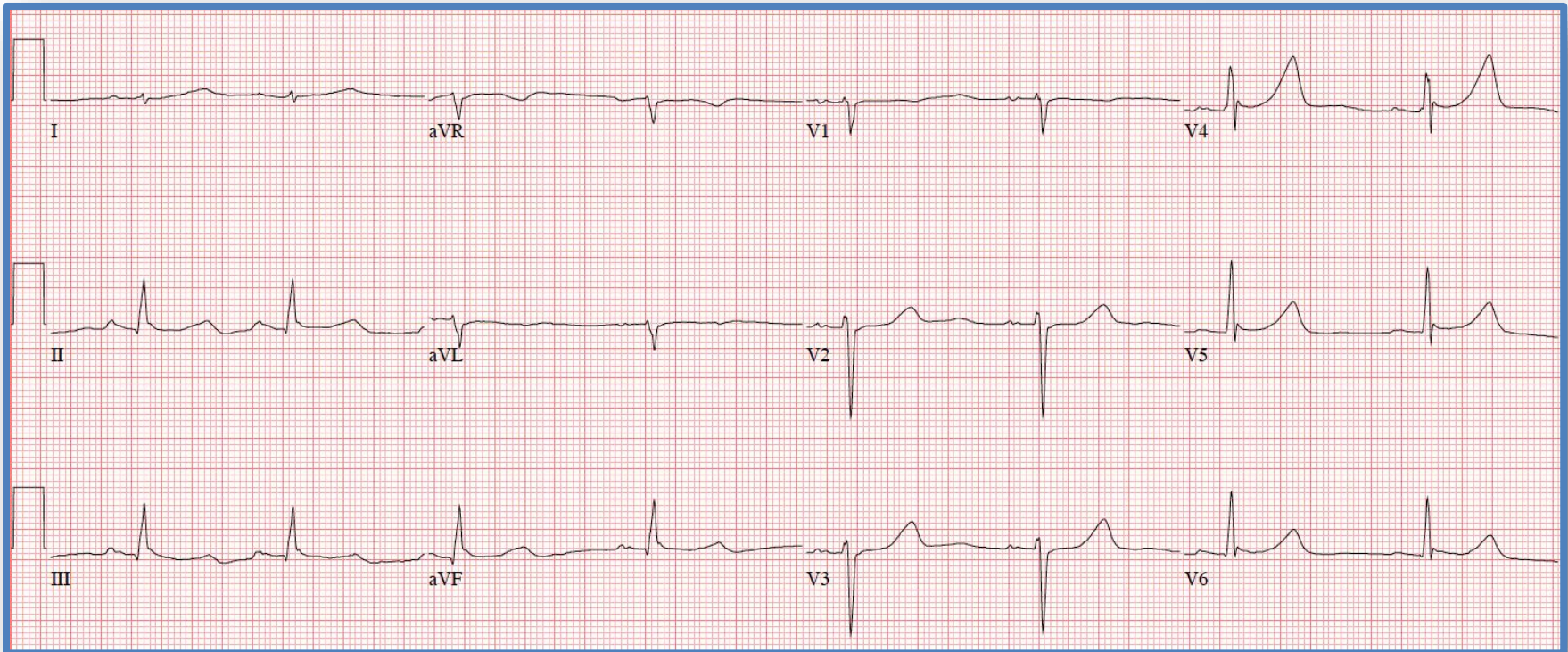
Case Presentation

- 62 year old Asian woman
 - Cardiac arrest getting ready for work
 - Husband hears a thump, works as a fireman
 - 13 year old daughter died suddenly at a church youth group, preceded by epilepsy for 2 years
 - PMH HTN on 25 mg HCTZ, recent URTI



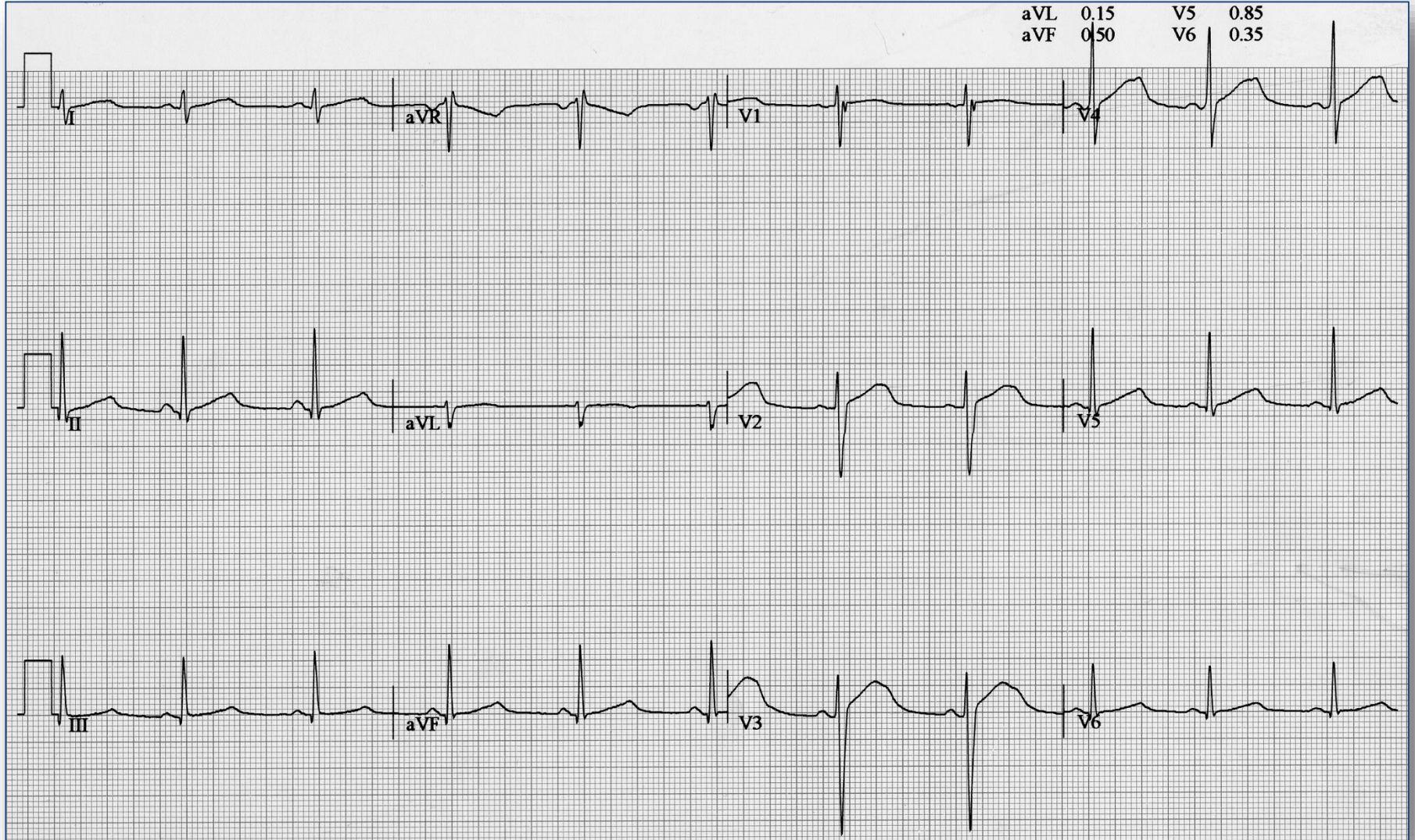
Evaluation

- Defibrillated in the field
- Cooled x 48 hours, wakes up
- Cath normal, echo normal



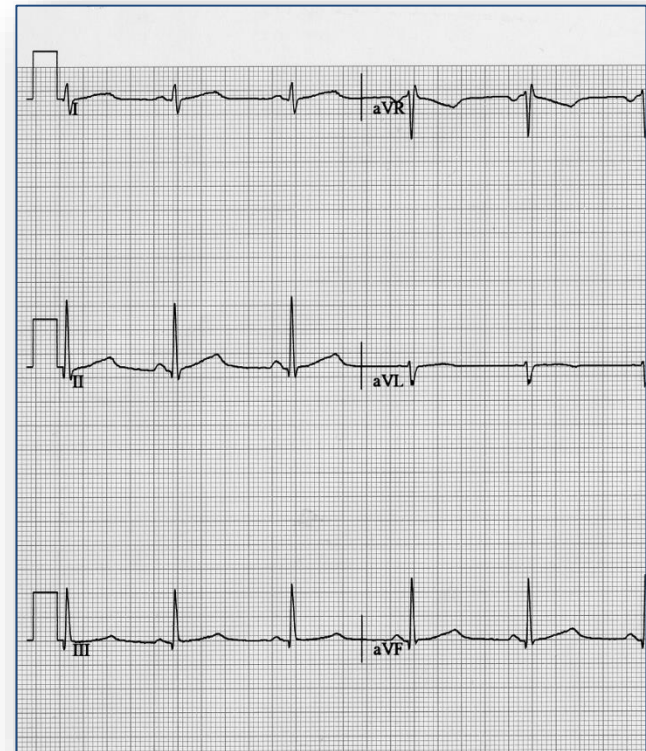
Cool ECG (literally)

Rest

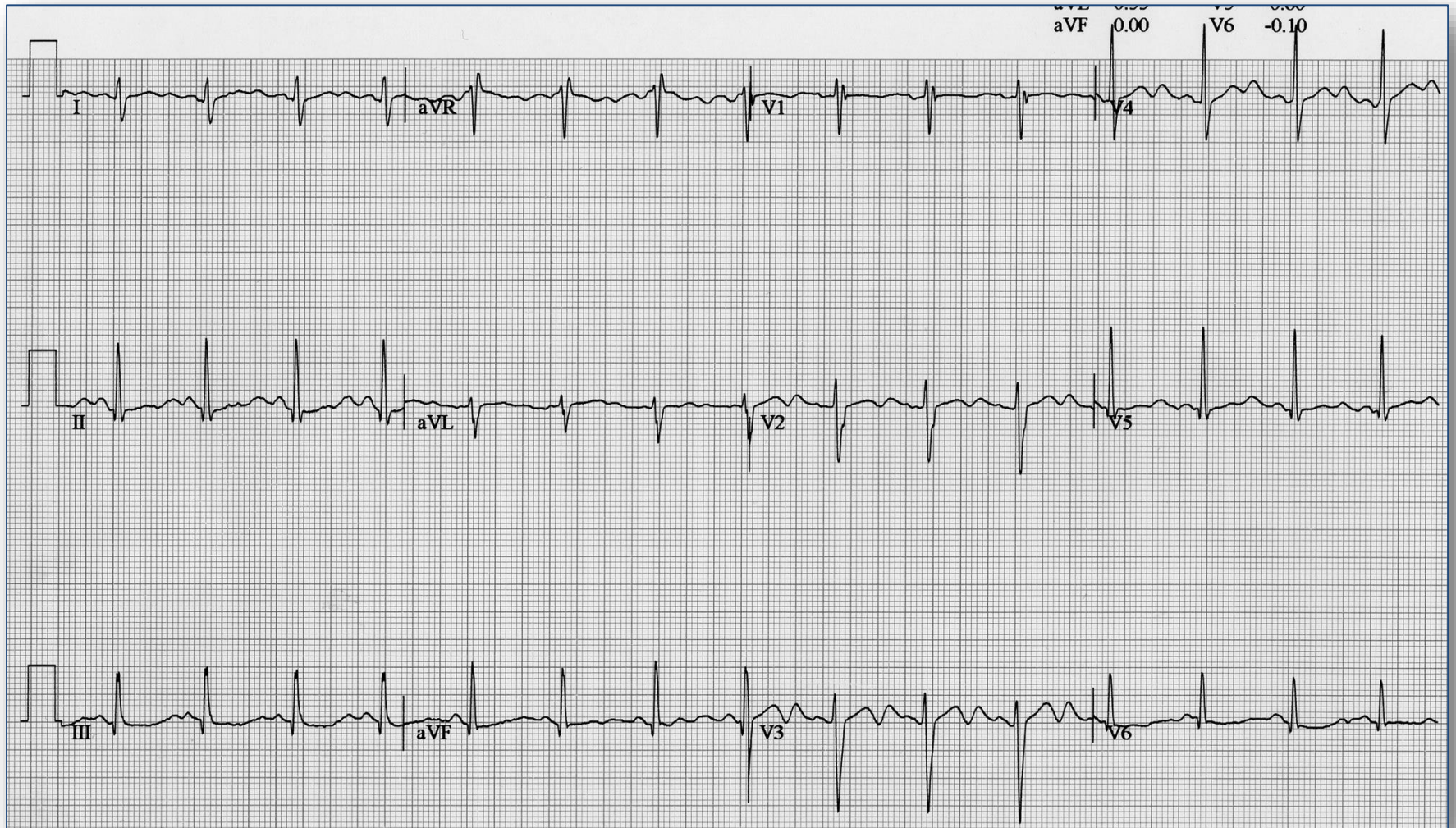


What is the most likely diagnosis?

1. Long QT syndrome
2. CPVT
3. ARVC
4. Early repolarization
5. Coronary spasm



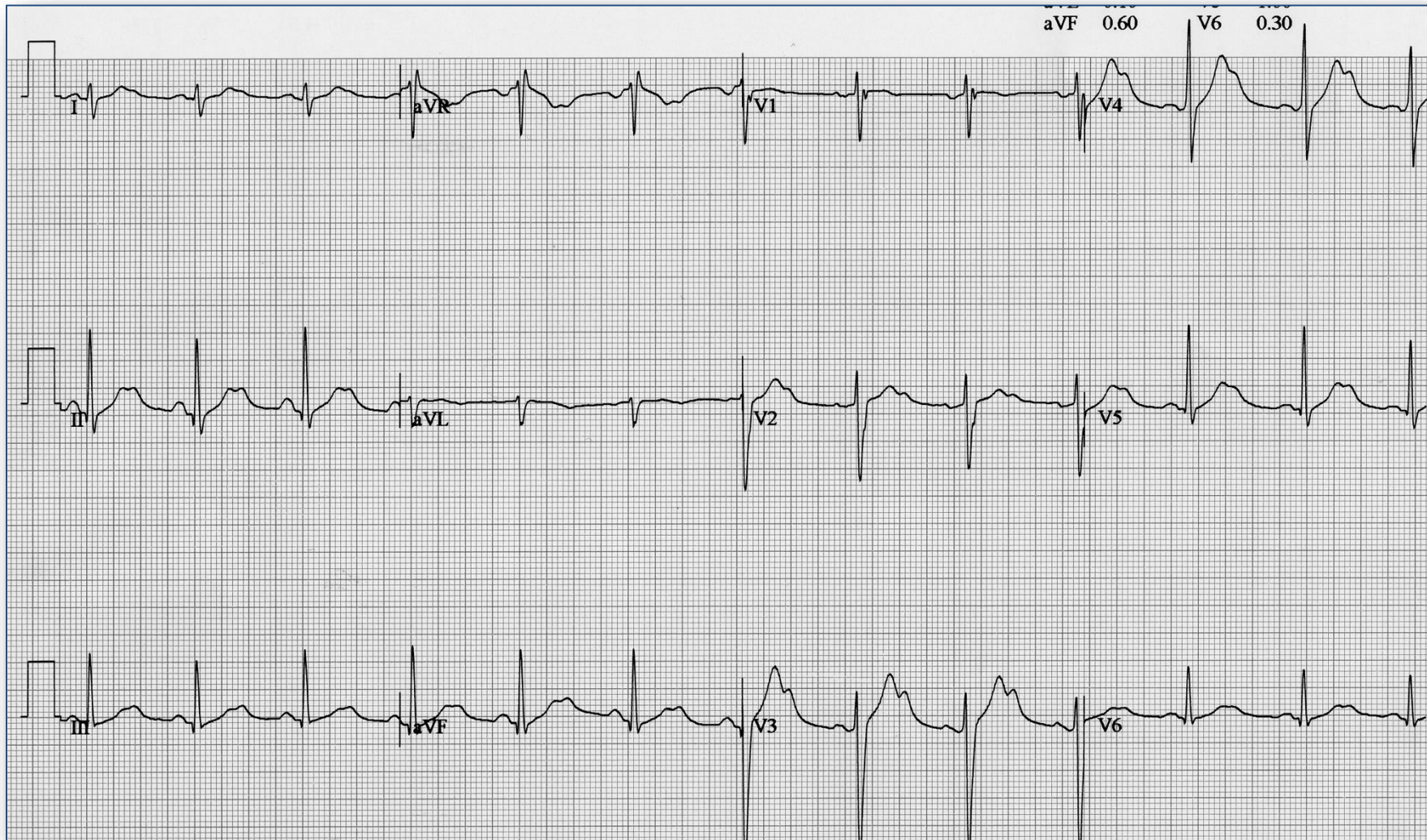
Standing and Early Exercise



Abnormal T Wave Shape And Duration



Mid Recovery



KCNH2 Channel- I_{Kr}

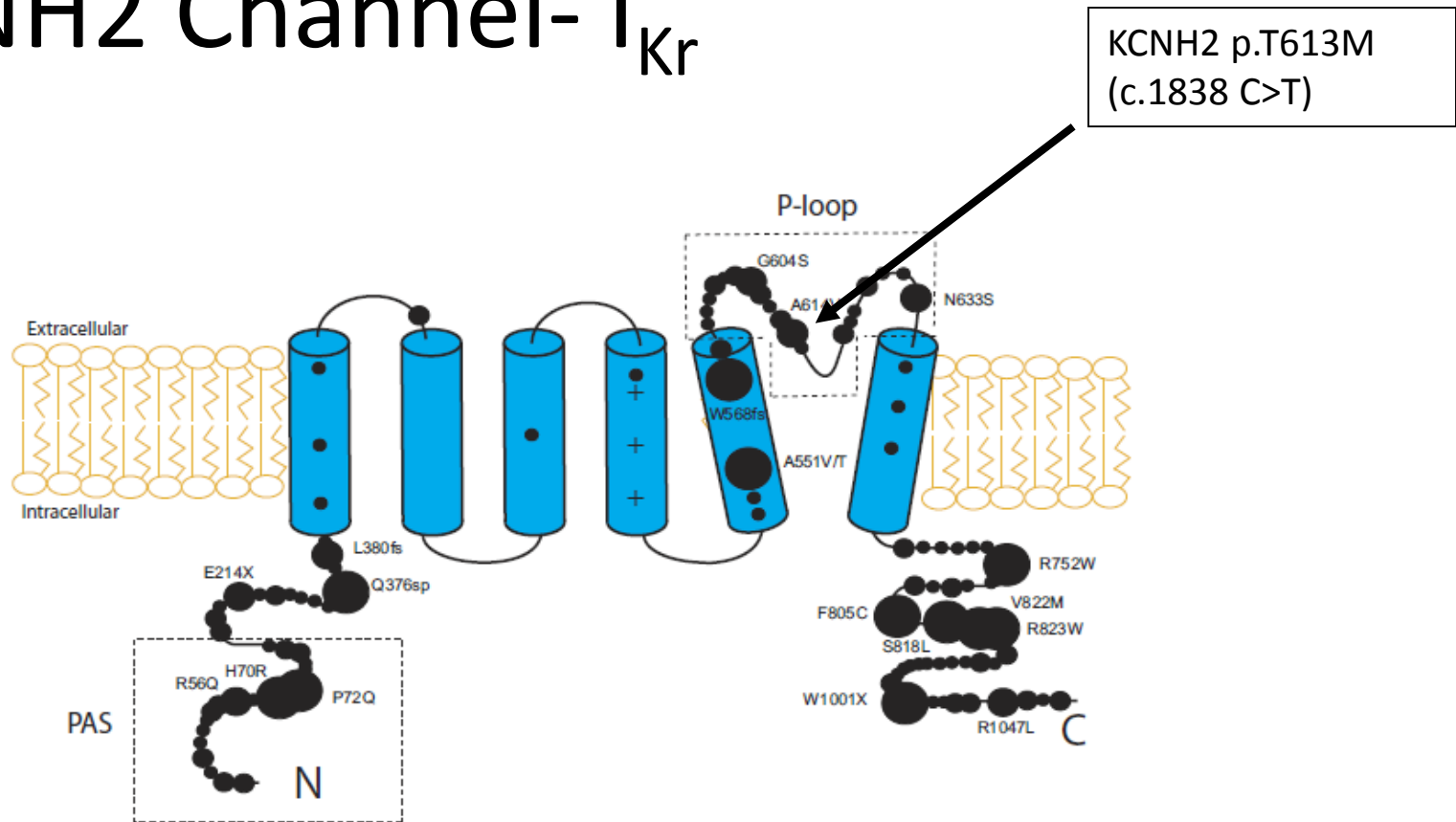
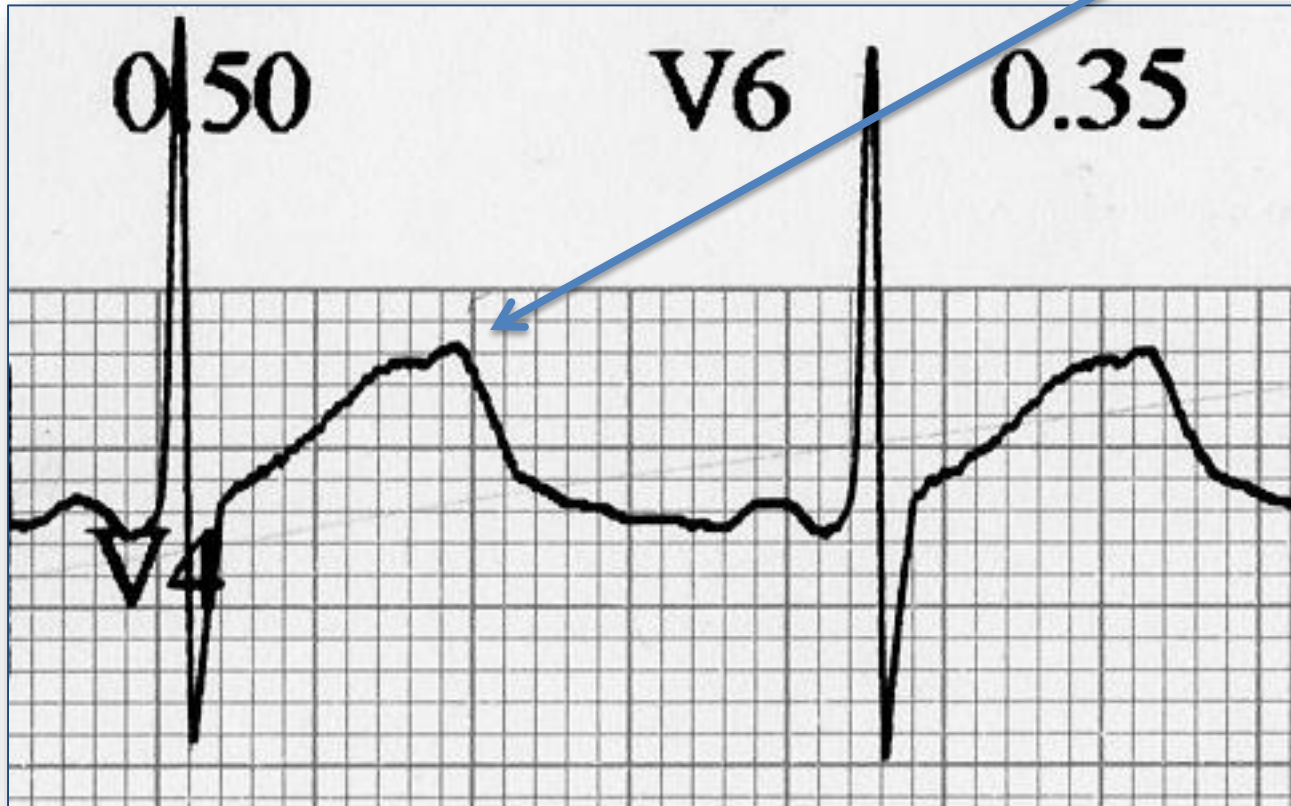


Figure 1 Distribution of mutations in the *KCNH2* potassium channel among study patients.

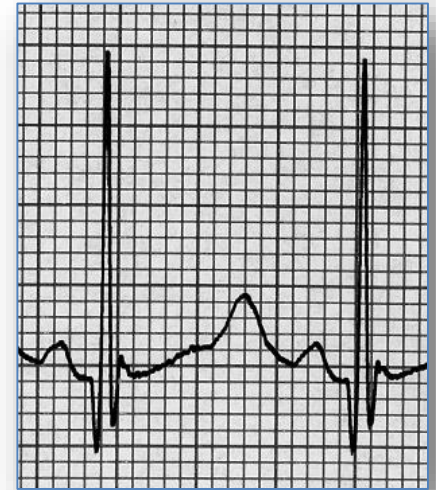
Mutation T613M located in the pore-loop (p-loop), between S5 and S6
11 additional presymptomatic family members identified !!!

Little Clue – Funny shaped T wave



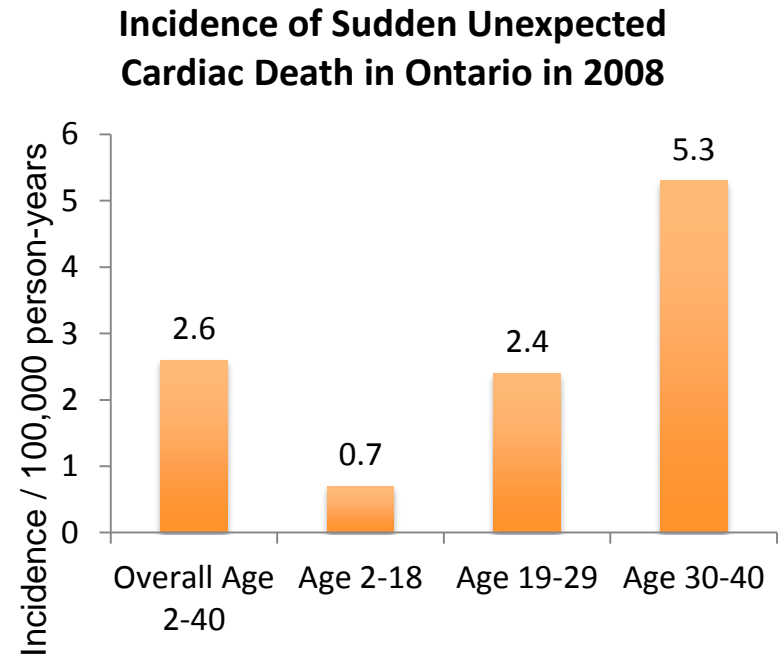
Long QT Syndrome: Our Poster Child

- Exercise or auditory stimuli trigger syncope and sudden death
- Autosomal dominant inheritance
- Prevalence 1:2500 (Schwartz Circ 2009)
- 4-20% lifetime risk of sudden death
 - 10% of sudden infant deaths (SIDS)
 - Drowning, accidents
- 50% have an ECG with normal or borderline QT
- Monogenic cause determined in 75%
- Rx beta blockers and rare use of sympathectomy and ICDs



Incidence of SCD in Ontario

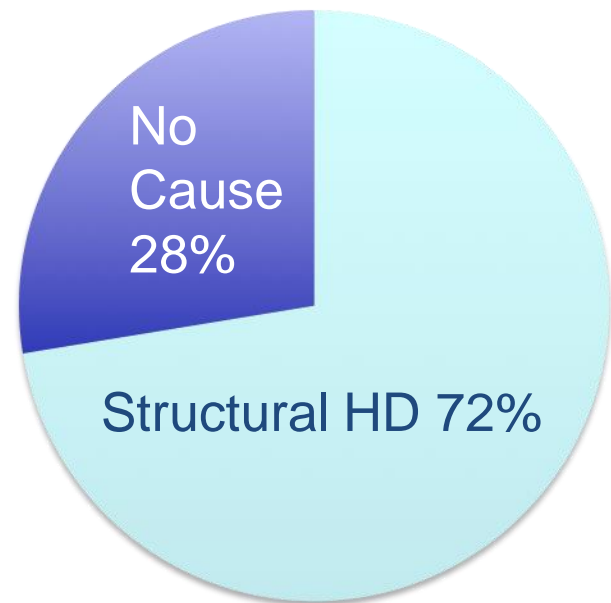
- Incidence of SUD/SCD:
 - Incidence reported per 100,000 person-years
 - Overall: 2.6
- Incidence increases with age:
 - Age 2-18: 0.7
 - Age 19-29: 2.4
 - Age 30-40: 5.3



Cause

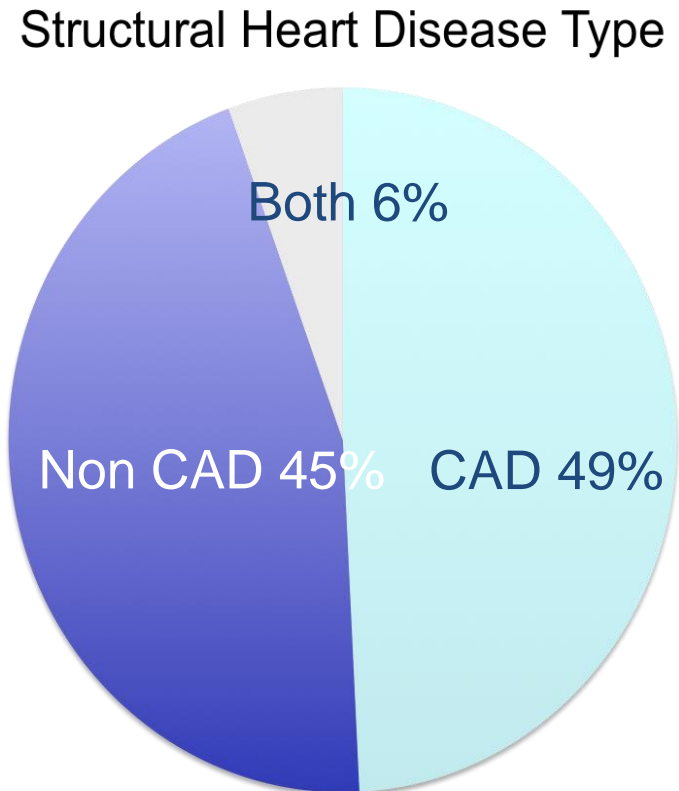
- Of 174 cases:
 - 126 has structural heart disease identified on autopsy
 - 48 had no identifiable anatomic or toxicologic cause of death
- Age differences:
 - <30 were more likely than those aged 31-40 to have SUD (OR=3.3, $p=0.0003$)

Attributed Cause of Death, all cases



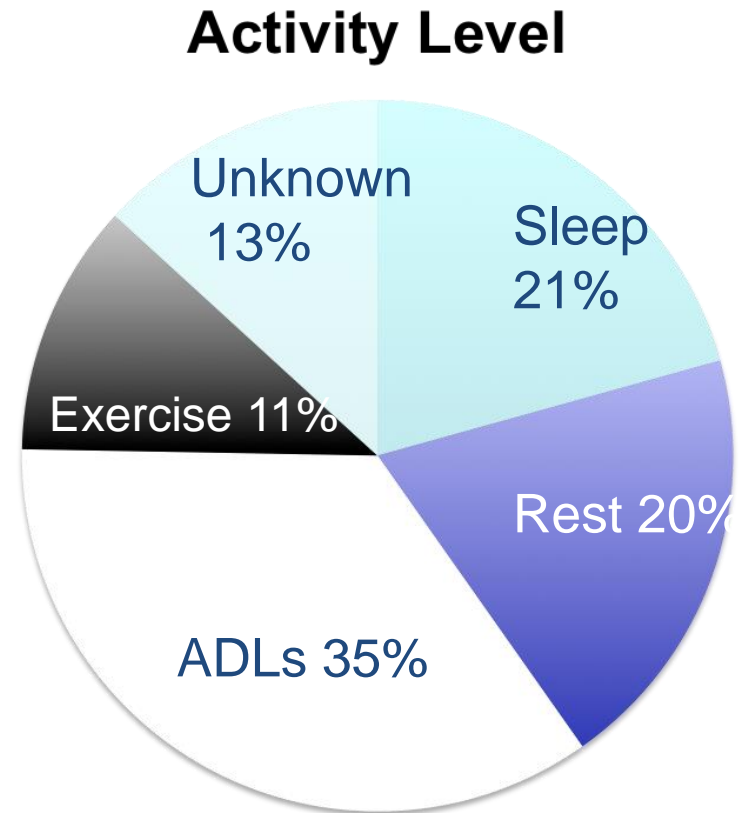
Structural Heart Disease

- CAD/ischemic
 - Single most common pathology
- Non-CAD/non-ischemic
- 78% of SHD was unrecognized (n=98), discovered on autopsy

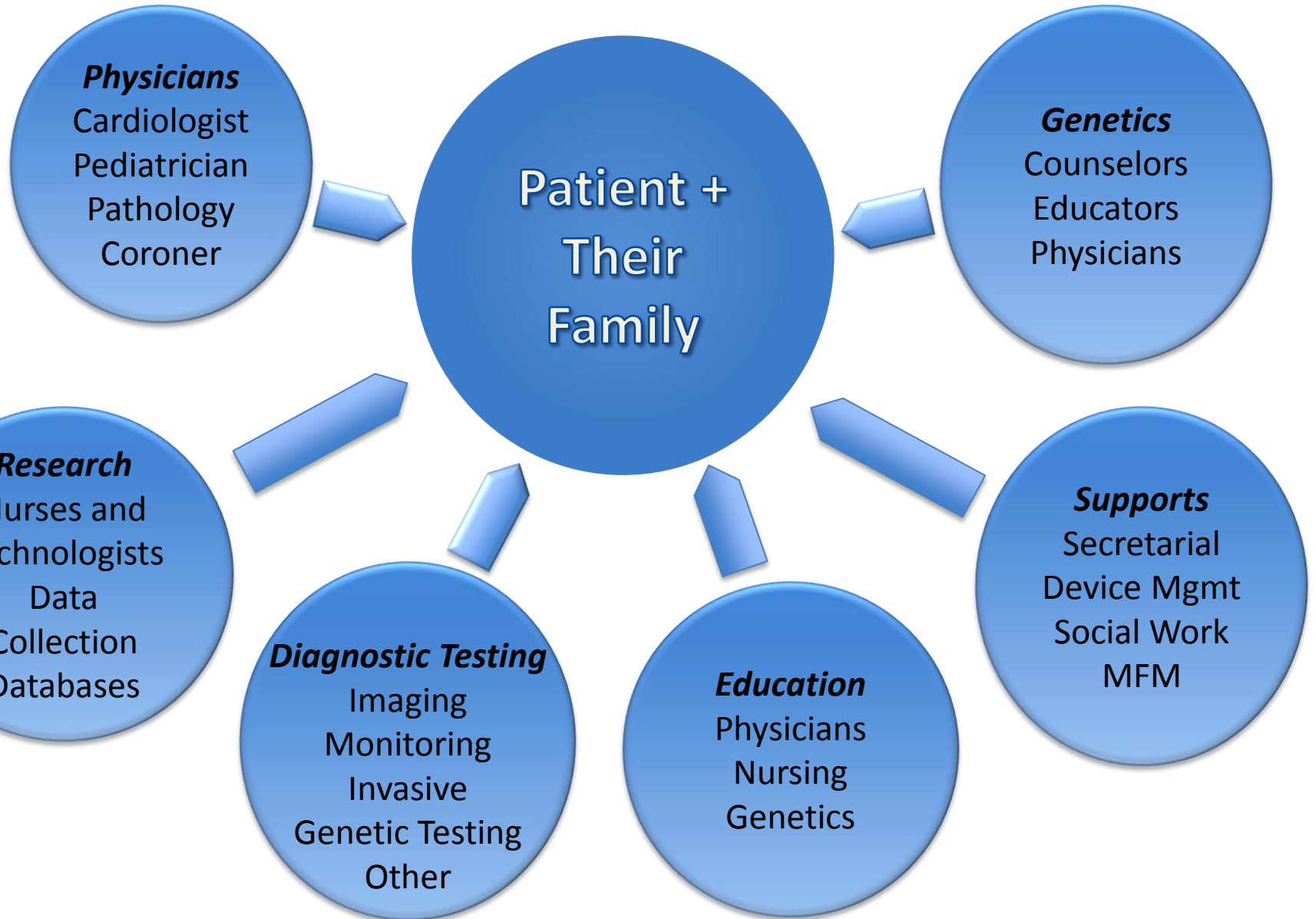


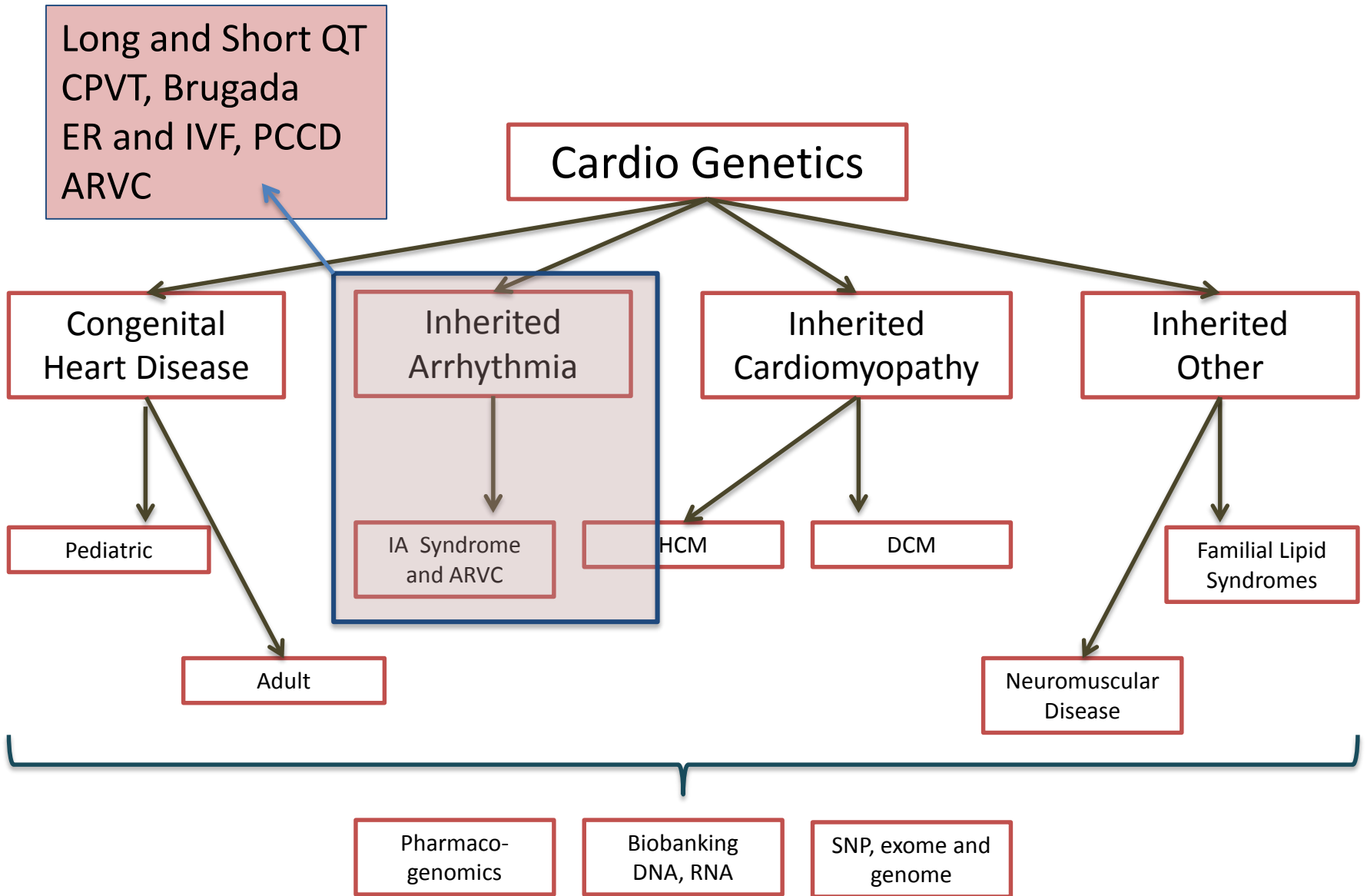
Activity Level

- Majority occurred at low activity levels
- Only 11% occurred during moderate or vigorous exercise
 - Age differences:
 - Age 2-18: 33% (6 of 18)
 - Age 19-40: 9% (14 of 156)
- No deaths during organized competitive sports



Inherited Arrhythmia Clinic





Inherited Arrhythmia Clinic



- Deliberately slow process
- Family oriented
- Key element is a thorough history – not just syncope and sudden death
 - Miscarriages, stillbirths, SIDS, drownings and accidents, triggers
- Systematic testing
 - Monitoring, imaging, provocation, genetic testing

Diagnostic Cascade – Family Member

ECGs

Regular ECG

High lead ECG

Signal Averaged ECG

Holter

Imaging

Echocardiogram

MRI (Cardiomyopathy)

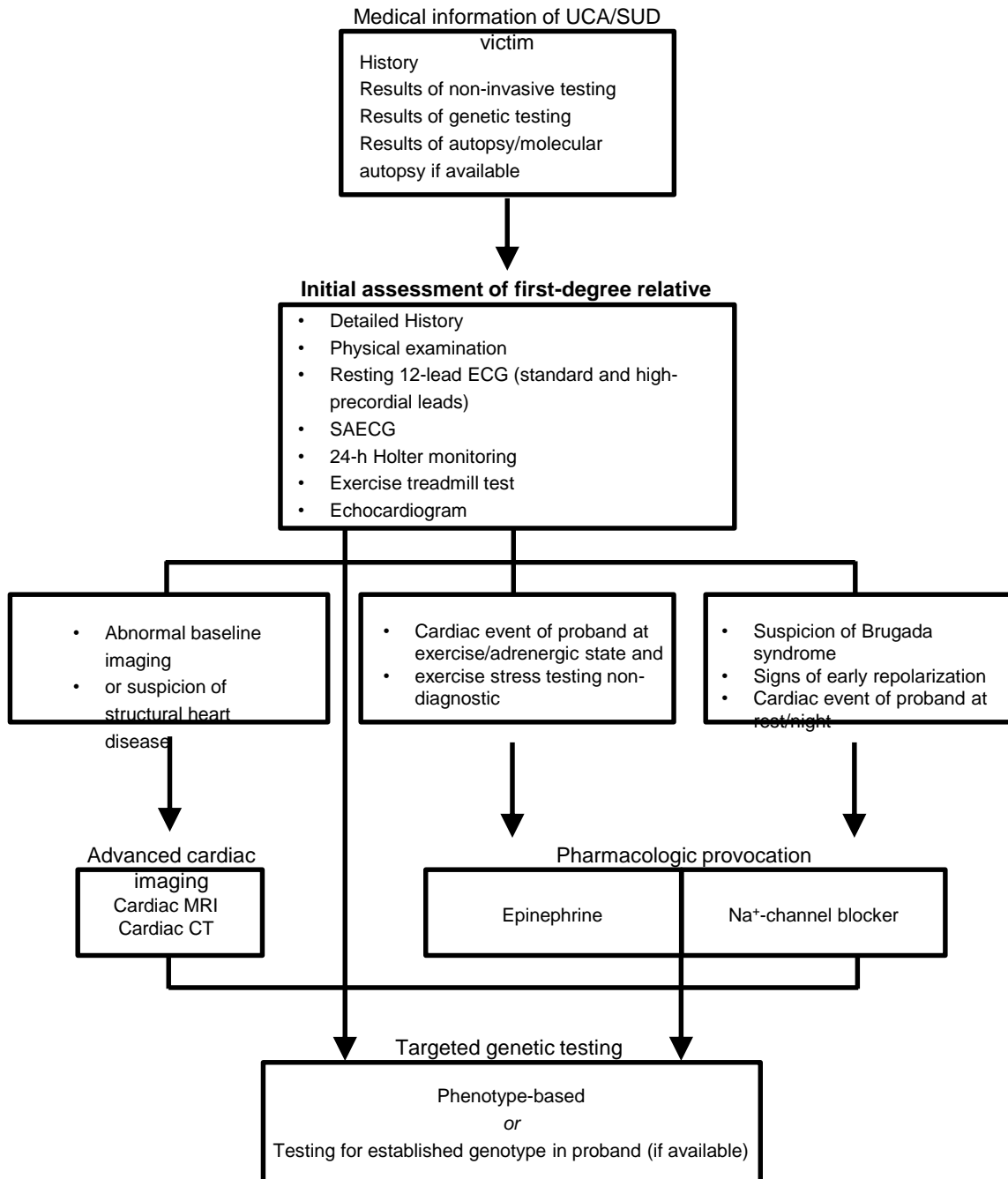
Cardiac CT

Provocation

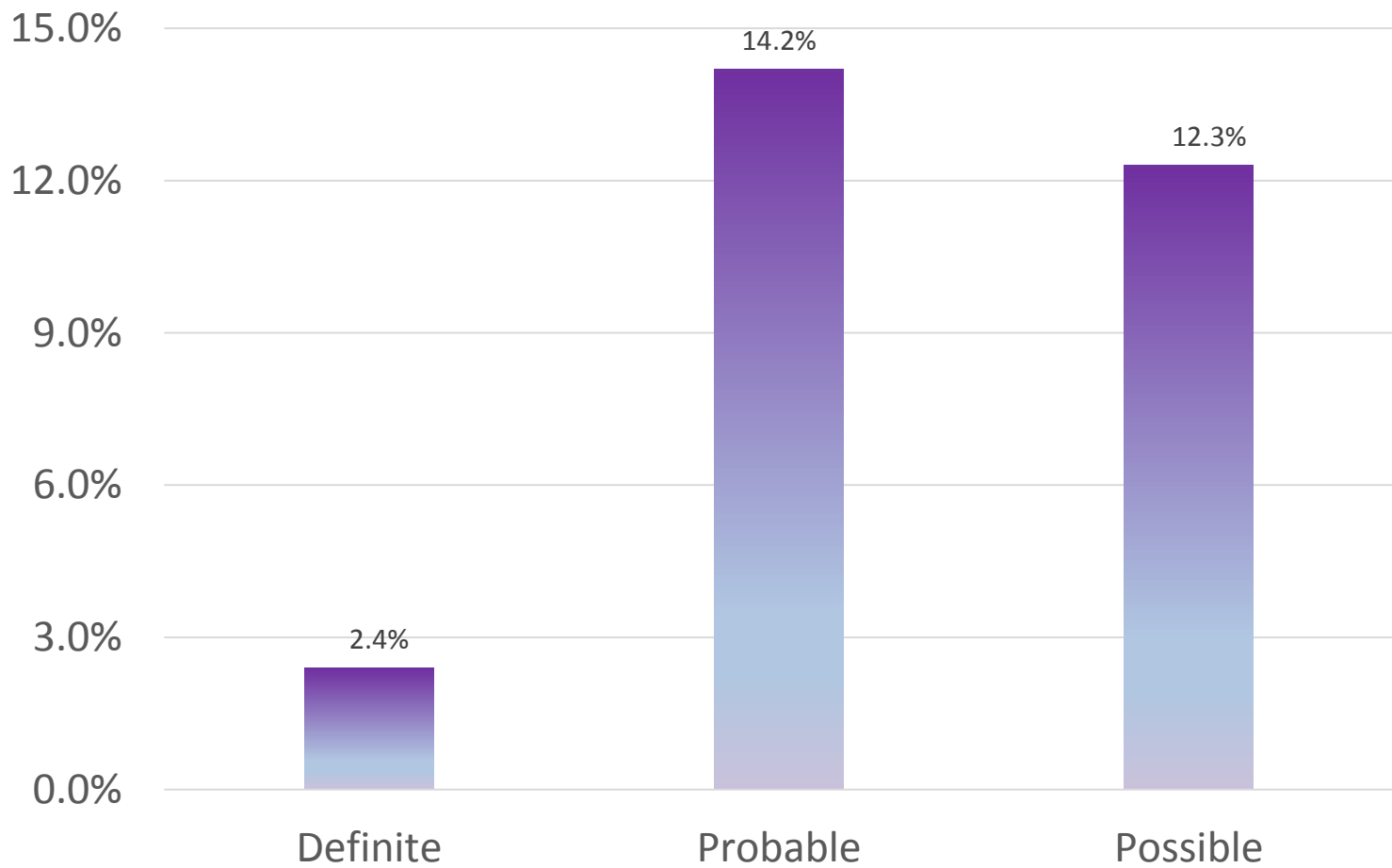
Stress test

Procainamide (Brugada)

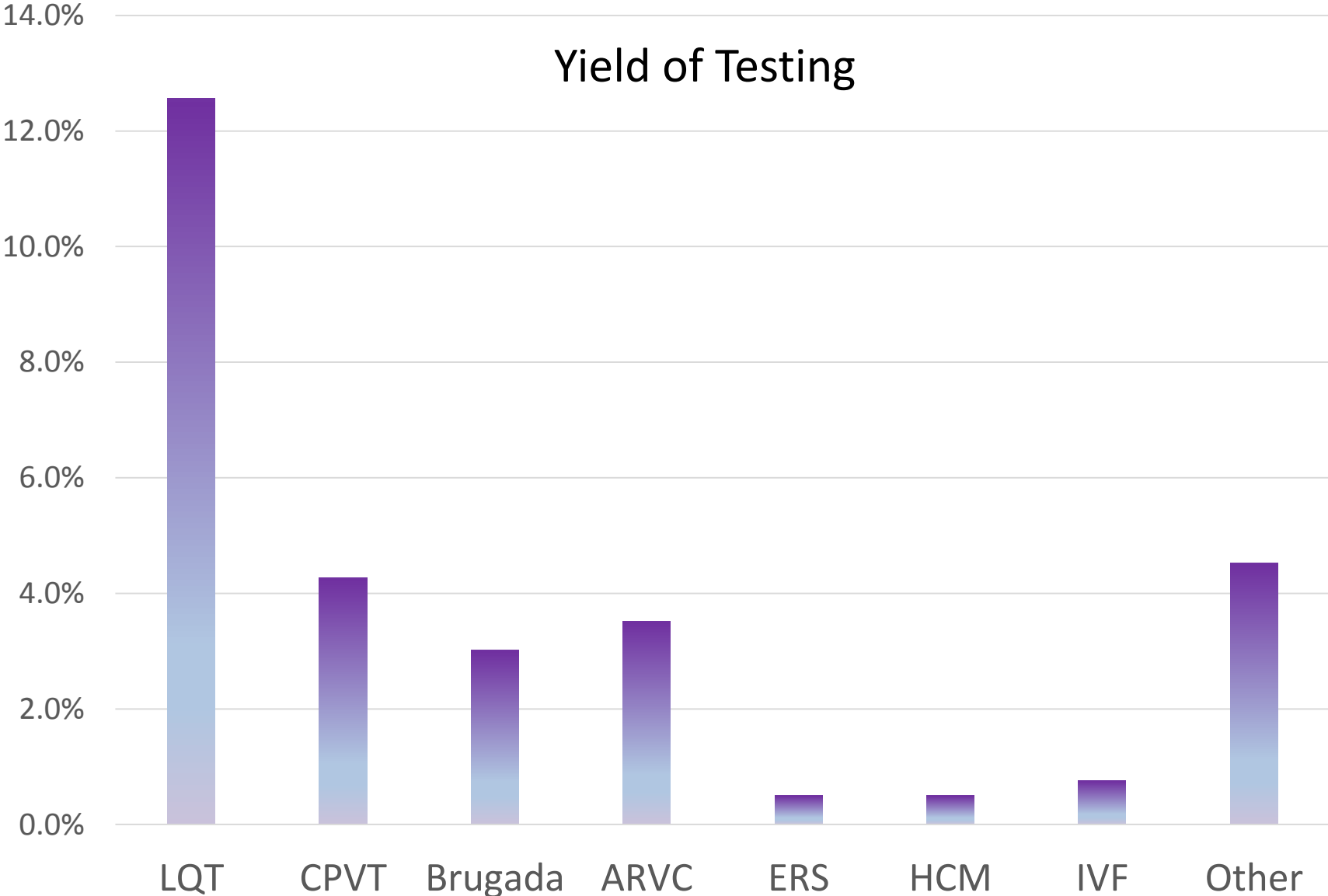
Epinephrine (Long QT and CPVT)



Relatives of SUD victims



Yield of Testing



Steinberg et al, Circ AE 2016

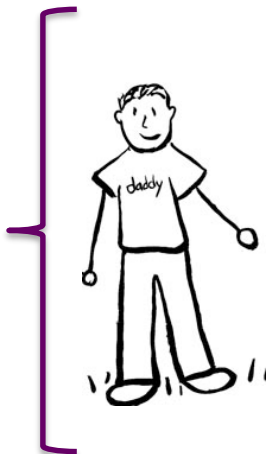
BCIAP Team with Visitors!



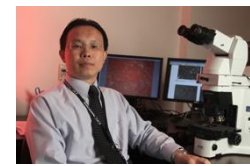
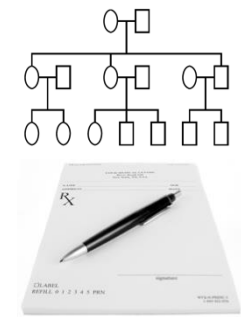
Physician Support has been AMAZING

Sudden Death Victim

- Coronary disease
- Cardiomyopathy
 - ARVC
 - HCM
 - DCM
- Ion Channelopathy
 - LQTS
 - CPVT
 - Brugada
- Other
 - Myocarditis
 - Infiltrative



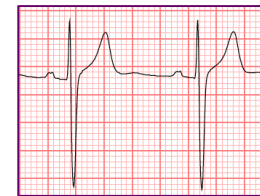
- Family history - sudden death, drowning, single vehicle accidents, crib deaths, heart failure, "heart attack"
- Personal history- syncope, seizures, exercise intolerance chest pain, medications
- Event history - exercise or rest, recent illnesses, fever, new medications
- Autopsy - preliminary and final report , molecular autopsy if no anatomical cause



First degree Relative

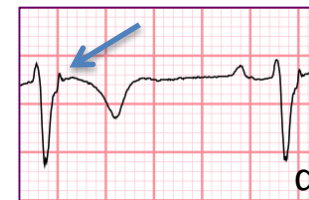
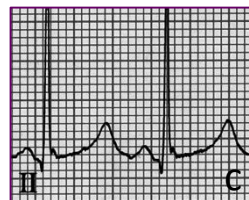
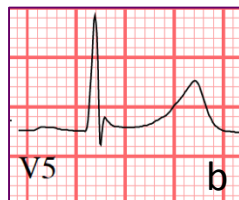
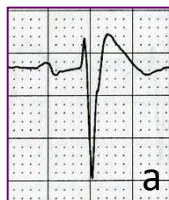


- Personal history - syncope, seizures, palpitations, chest pain, shortness of breath
- Physical examination – murmur, abnormal pulses
- Electrocardiography - ECG, high precordial lead ECG, signal averaged ECG, Exercise test, Holter monitor
- Imaging – echocardiogram
- Special testing – drug challenge, cardiac MRI, genetic testing if sudden death victim has positive testing



ECG Samples:

- a. Type 1 Brugada (lead V₁)
- b. Early Repolarization
- c. Long QT
- d. Epsilon wave (lead V₁)



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2016



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A Touch of Presence
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 Deliverance Matthew@HeartPresence.org
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Conclusions

- Sudden death in the young is a horrible but not that uncommon thing
- 1/3 of these will have a potentially inherited component
- A team approach is necessary to make lemonade out of lemons (and protect those left behind)
- A network of clinics including one in Winnipeg wants to help these families



www.heartsinrhythm.ca

I am an open access person – for slides, e mail akrahn@mail.ubc.ca

Important Websites

- www.heartsinrhythm.ca
- www.qtdrugs.org
- www.brugadadrugs.org
- www.heartrhythmresearch.ca
- akrahn@mail.ubc.ca

