

# The Post-Mortem Evaluation: What is the Yield?

February 7, 2020

Peter Aziz, MD

Director, Inherited Arrhythmia Clinic



**Cleveland Clinic Children's**



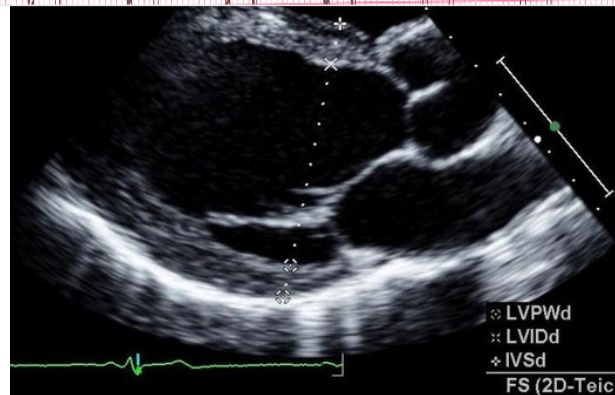
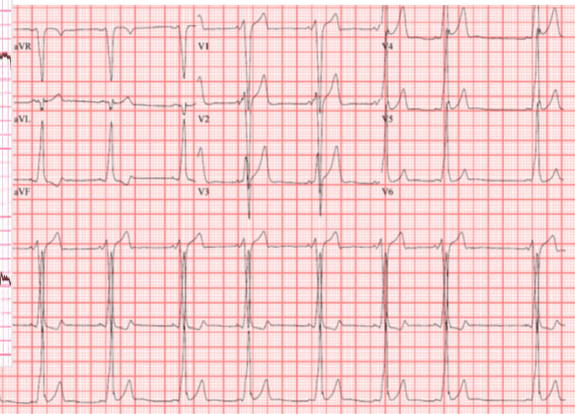
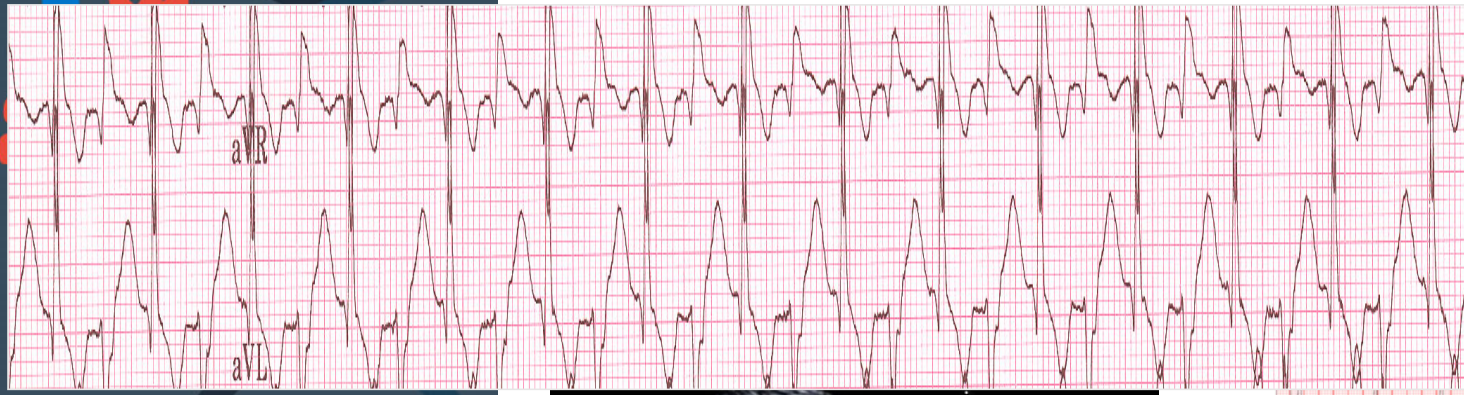
# The Cardiac Family History

## Identifying the at Risk Relative

- Sudden death <50yo (including MIs)
- Unexplained drowning
- Seizures
- Syncope
- Congenital deafness
- ICDs/pacemakers *at an early age*



# Low Hanging Fruit



# Inherited Arrhythmia Clinic



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## Inherited Arrhythmia Clinic

### Doctors

Director: [Peter Aziz, MD](#)

### Staff Members

#### Center for Pediatric and Congenital Heart Disease

- [Peter Aziz, MD](#) - Pediatric Electrophysiology and Pacing, Director
- [Elizabeth \(Tess\) Saarel, MD](#) - Pediatric Cardiology (Department Chair), Section of Electrophysiology and Pacing

#### Miller Family Heart and Vascular Institute

- [Bruce Lindsay, MD](#) - Cardiovascular Medicine, Section of EP and Pacing
- [Mina Chung, MD](#) - Cardiovascular Medicine, Section of EP and Pacing
- [Patrick Tchou, MD](#) - Cardiovascular Medicine, Section of EP and Pacing
- [Niraj Varma, MD](#) - Cardiovascular Medicine, Section of EP and Pacing

#### Genetic Counseling

- Diane Clements, LGC
- Brittany Psensky, MS, LGC
- Christina Rigelsky, MS, LGC

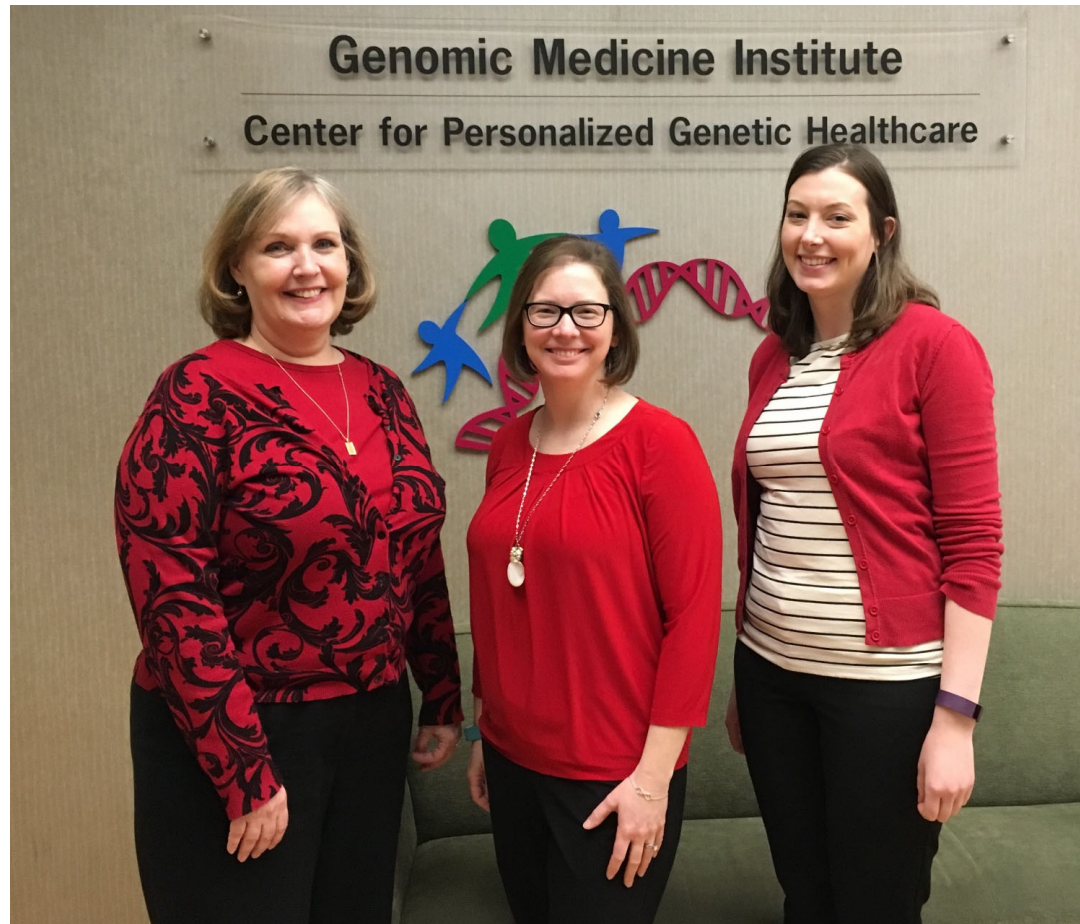
Learn more about the [Center for Personalized Genetic Healthcare](#).

- Structured to evaluate the family, any age
  - All first degree relatives
  - Pediatric and adult EP
  - Consistency in care/management
- Monthly meetings
- Liaise with pathologists and coroners



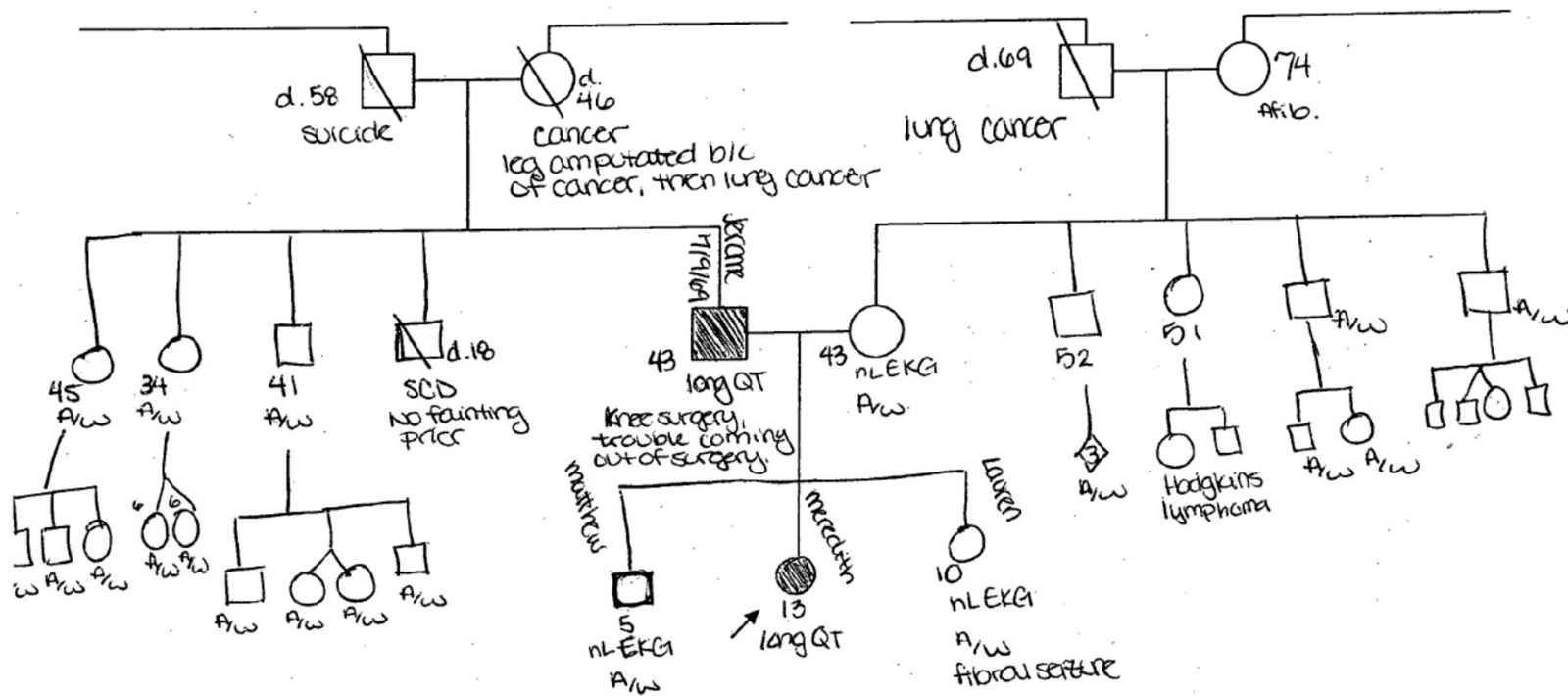


# Involve Genetic Counselors



# Pedigree

Ethnicity: Irish, Czech



# The Evidence



# A Prospective Study of Sudden Cardiac Death among Children and Young Adults

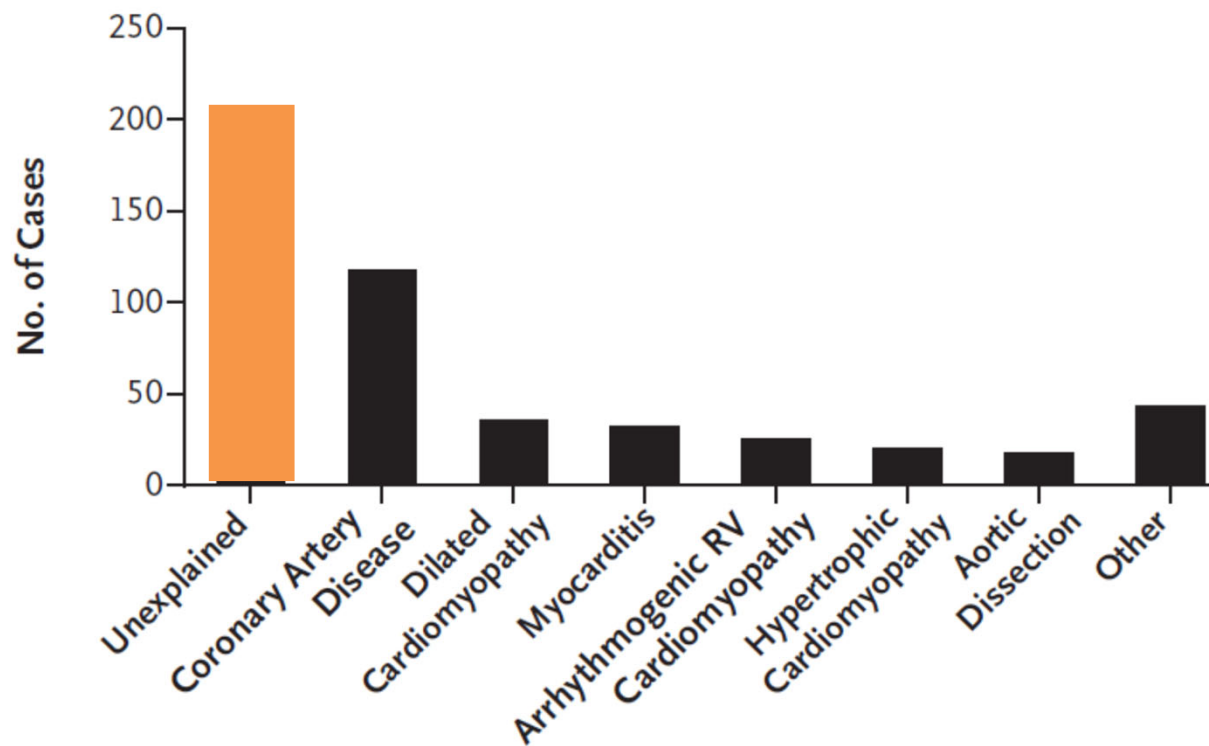
- SCD (1 to 35 years)
- Australia and New Zealand from 2010-2012
- 490 cases identified
  - 72% male
  - Initially evaluated with autopsy





# A Prospective Study of Sudden Cardiac Death among Children and Young Adults

C Causes of Sudden Cardiac Death

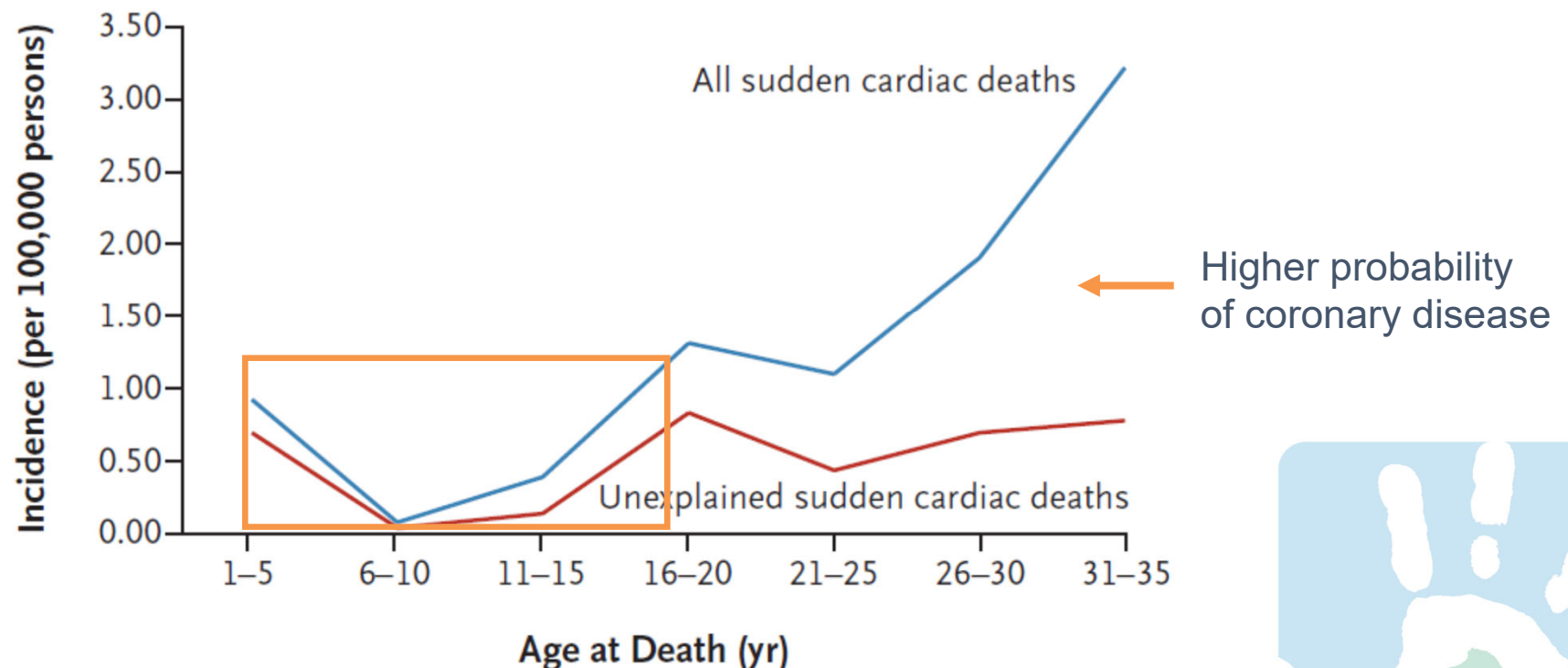


Bagnall RD, *NEJM* 2016



# A Prospective Study of Sudden Cardiac Death among Children and Young Adults

A All Sudden Cardiac Deaths and Unexplained Sudden Cardiac Deaths



Age at Death (yr)  
Bagnall RD, ..., Skinner JR, et al *NEJM* 2016

# Sudden Unexplained Death

## Heritability and Diagnostic Yield of Cardiological and Genetic Examination in Surviving Relatives

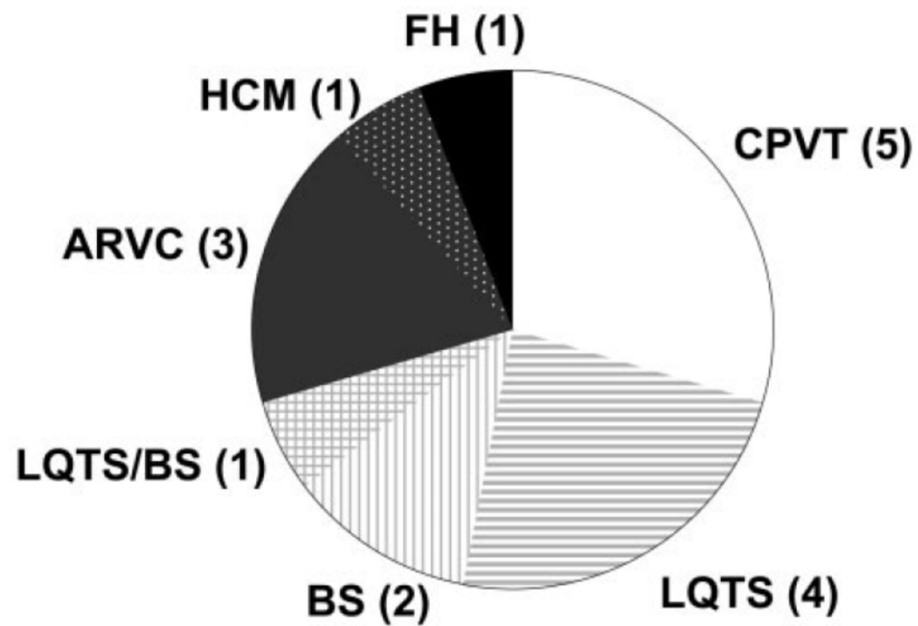
- 43 Families with sudden unexplained death <40 years of age
- First-degree relatives were evaluated
  - 183 surviving relatives

Tan HL *Circulation* 2005



# Sudden Unexplained Death

## Heritability and Diagnostic Yield of Cardiological and Genetic Examination in Surviving Relatives



Tan HL *Circulation* 2005

- 17/43 (40%) were diagnosed
- Through familial screening, 151 surviving relatives were affected (8.9 per family)



# Sudden arrhythmic death syndrome: familial evaluation identifies inheritable heart disease in the majority of families

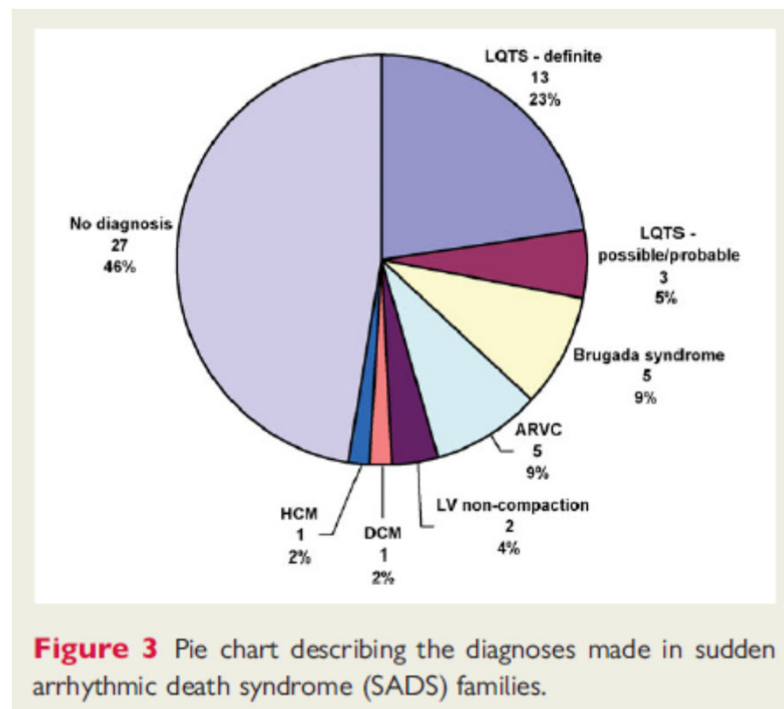
- 57 families with an unexplained death (SADS)
  - Mean age 26 years
  - 61% male
- First degree relatives underwent comprehensive evaluation
  - 184 relatives (3.2 per family)

Behr ER, *European Heart Journal* 2008



# Sudden arrhythmic death syndrome: familial evaluation identifies inheritable heart disease in the majority of families

- 30/57 (53%) a definite or probable cause was identified in the family



Behr ER, *European Heart Journal* 2008



# Practical Considerations



# Common Barriers



- Conversations agitate old wounds
- Evaluations often necessitate future correspondence
  - Phone calls
  - Information exchange
- Families can feel probed in the process



# Common Barriers



- Inevitable relative that refuses to be evaluated
  - Some family members simply don't want to know
- Institutions must be equipped to evaluate patients of any age
- Not everyone lives in Cleveland
  - And they won't come in the winter



# Ethical Considerations



Quenin, *Circulation EP* 2017



# Key Takeaways

- The answer is often not attainable in the deceased
- The family provides important diagnostic clues
- A structured systemic approach is required to adequately assess all family members
- Though our role is to provide answers, we must respect the emotional burden the family carries





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# Sudden arrhythmic death syndrome: familial evaluation identifies inheritable heart disease in the majority of families

**Table 4** Predictors of a diagnosis of inherited heart disease in evaluated first-degree relatives and families: first degree relatives are considered initially as individuals and then as part of families

	Diagnosis present	Diagnosis absent	Statistical significance?
Evaluated first-degree relatives			
Total no.	46 (25%)	138 (75%)	—
Age (years) mean	31.0	36.8	ns
Male	39%	41%	ns
Symptomatic	46%	18%	$P < 0.001$
Presyncope	7%	1%	$P = 0.035$
Syncope	20%	4%	$P < 0.001$

Behr ER, *European Heart Journal* 2008