Aetiology and Outcomes of Paediatric Out-of-Hospital Cardiac Arrest Survivors

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Heart Rhythm Congress

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Background

- SCD accounts for ~400 deaths per year in UK population (1-34 years old).
- Undiagnosed **inherited cardiac conditions** cause ~50% of SCD cases.
- Aetiology of paediatric SCA specifically is unknown.
  - Lack of studies
  - Differences in research design, inclusion criterias, etc.

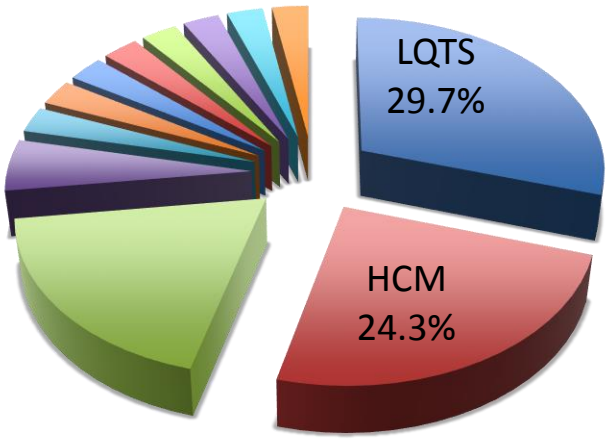
Aim: What is the aetiology of out-of-hospital cardiac arrest in paediatric survivors?

Methods

- Single-centre, retrospective study
- 37 patients (0-16 years) – survived out-of-hospital VF arrest between Aug 1996 and Oct 2017
- Significant clinical notes and investigation findings reviewed and recorded
- 21 out of 37 (57%) were males
- Median age at arrest was 11.0 years (IQR = 6-13 years)
- Exclusion criteria:
  - SCA secondary to a non-cardiac cause
  - Previously known relevant cardiac condition

Results

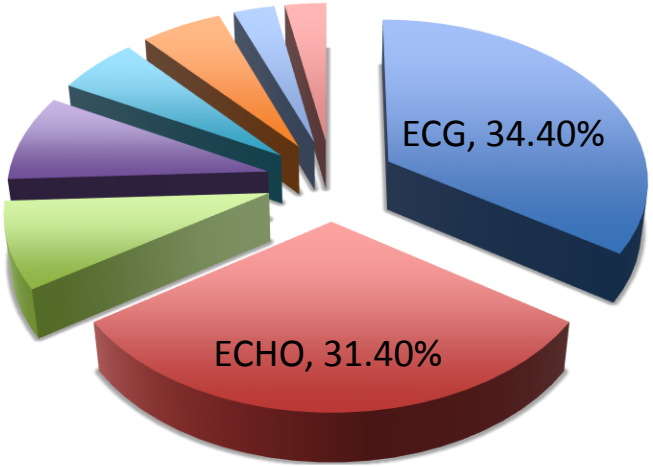
Diagnoses



- LQTS
- HCM
- CPVT
- Idiopathic VF
- ARVC
- SQTS
- WPW Syndrome
- Subaortic Stenosis
- Bi-ventriclular NCC
- ERS
- BrS
- DCM

- 33/37 patients (89.2%) diagnosed with inherited cardiac condition.
- LQTS and HCM most commonly diagnosed condition.

Investigations used to make diagnosis



- ECG
- ECHO
- Holter Monitoring
- ECG & ACT
- EST
- Genetic Test
- MRI
- ECG & MRI

- ECG and ECHO most frequently used investigation to confirm diagnosis.

Genetic Testing

- 25/37 genetic results available → 17 causative mutations found

Diagnosis	Number of patients identified with a gene mutation/total number of patients tested (% yield)	Gene Mutations
LQTS	4/9 (44%)	a) SCN5A (LQT3) b) KCNH2 (LQT2) c) CACNA1C (LQT8)
HCM	5/7 (71.4%)	a) MYBPC3 b) TNNI3
CPVT	6/7 (85.7%)	a) RYR2
SQTS	1/1 (100%)	a) SLC22A5
BrS	1/1 (100%)	a) SCN5A

- Genetic testing important as a diagnostic tool

Conclusions

- Inherited cardiac conditions the most common cause of out-of-hospital cardiac arrest
- LQTS and HCM most commonly diagnoses
- ECG and ECHO most useful investigations
- Highlights importance of genetic testing.

Clinical Implications

- Full clinical and genetic workup of proband is needed
- Important to screen first-degree relatives