

**Sudden arrhythmic death syndrome (SADS): Diagnostic yield of comprehensive clinical evaluation of paediatric first-degree relatives**

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**Background:** Sudden arrhythmic death syndrome (SADS) is most often caused by heritable cardiac diseases. Studies in adults have identified evidence of inherited cardiovascular diseases in up to 53% of families, but data on the prevalence of familial disease in children are scarce. The aim of this study was to evaluate the yield of clinical screening in paediatric first-degree relatives of victims of SADS or aborted cardiac arrest (ACA) using a systematic and comprehensive protocol.

**Methods:** All consecutive paediatric patients referred for family screening between 2003 and 2013 after a SCD or ACA of a family member were retrospectively enrolled into the study. Systematic evaluation of the children included clinical examination, family history, ECG, echocardiogram, 24-hour tape and signal averaged ECG. Older patients also underwent exercise testing, cardiac MRI and ajmaline provocation testing.

**Results:** A total of 110 children from 63 consecutive families were included in the study. An inherited cardiac disease was identified in 12 1st-degree children from 11 (17.5%) families (7 children were diagnosed with Brugada syndrome, 2 with long QT syndrome, 1 with CPVT, and 2 had late potentials on signal averaged ECGs).

**Conclusions:** These data show a high prevalence of inherited heart disease in paediatric first-degree relative in families with a history of SCD or ACA. The results highlight the importance of a systematic, comprehensive approach and ongoing screening of paediatric family members.