P-2

Fever In Children At-Risk For The Brugada Syndrome

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Introduction (or Basis or Objectives): Brugada syndrome (BrS) is a rare inherited arrhythmia syndrome. Of the genes associated with BrS, variants in the SCN5A gene are most frequently described. Usually however, no associated genetic variant is identified. BrS patients, especially children, are at risk for arrhythmic events during fever. Fever is known to unmask the Brugada-type-1 pattern. Children with a positive family history for BrS, genotype-positive, and phenotype-positive children are considered at-risk for Brugada-type-1 pattern and cardiac events (CE). They are thus advised to record an ECG during fever, which is a stressful experience.
To evaluate the current policy, we aimed to identify risk factors for Brugada-type-1 pattern during fever (F-type-1) and CE by evaluating demographics and genetic background in an at-risk paediatric population.

Methods: Children with a positive family history for BrS, genotype-positive, and phenotype-positive children with ≥1 available fever-ECG from three tertiary medical centres were included and divided into two groups depending on the occurrence of F-type-1 during follow-up. Demographics and ECGs during fever were retrospectively retrieved and analysed. Chi square test and applicable post-hoc analyses with Bonferroni correction were used.

Results: The study population consisted of 68 children, 21 were allocated to the F-type-1 positive group and 47 to the F-type-1 negative group. The groups differed significantly in CE; in the F-type-1 positive group 7 (33,3%) children had CE during follow-up in comparison to 0 in the F-type-1 negative group (p=0,000). Five of them had symptoms during fever. The two groups also differed significantly in genetic background (p=0,002, See Figure 1). In the F-type-1 positive group 14 (43,8%) were SCN5A variant carriers in comparison to 1 (4,2%) in the genotypic negative children (p=0,001). When children diagnosed with BrS at first presentation after CE (n=3) or type-1-ECG at baseline (n=1) were excluded, this result remained unchanged.

Conclusions: Our results suggest that carrying a SCN5A variant is a risk factor for F-type-1 in children. F-type-1 is also associated with symptoms in this population. Children carrying SCN5A variants associated with BrS should be closely watched during febrile episodes and an ECG should be recorded.

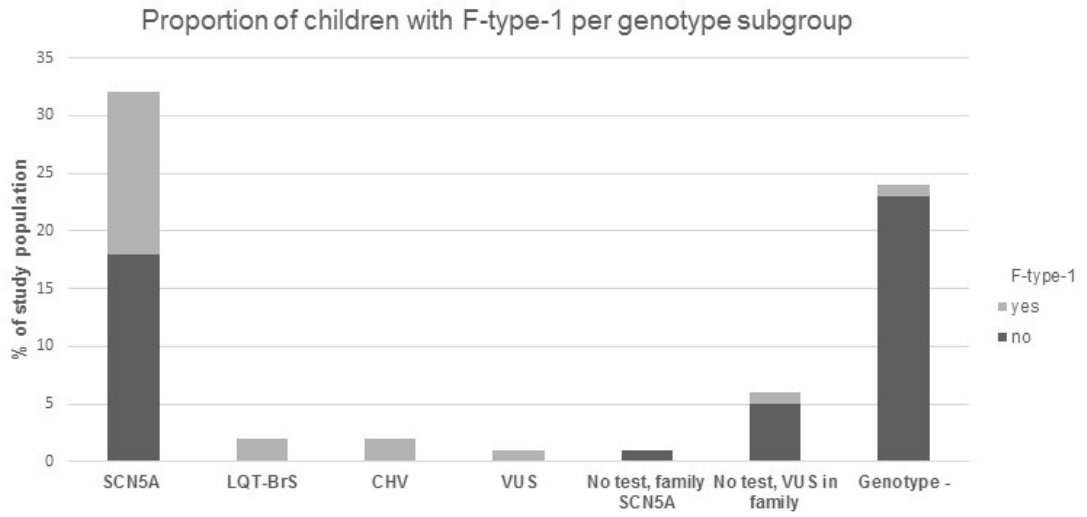

Figure 1: Proportion of children with F-type-1 per genotype subgroup. SCN5A: SCN5A pathogenic variant carrier; LQT-BrS: overlap syndrome variant carrier; CHV: compound heterozygous SCN5A variant carrier; VUS: variant of unknown significance