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Brugada Syndrome in the paediatric population: experience from a single center

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Introduction
Brugada syndrome (BrS) is an arrhythmogenic disease typically diagnosed during adulthood. Few studies exist on paediatric population.
Aim of this study was to analyse the clinical, electrocardiographic (ECG) and genetic characteristics in the paediatric patients of the Brugada Registry of the Piedmont Region of Italy.

Methods
1044 patients were included in the Brugada Registry from 2001 to 2017. Database was analysed looking for patients younger than 18 years old.

Results
53 patients were included in the study. Mean age at diagnosis was 12,8 +/- 4,8 years. 40 patients (75%) were male. Most patients (42, 79%) were asymptomatic at presentation: diagnosis was made during occasional ECG in 26 (62%), familial screening in 11 (26%), ECG made during fever in 4 (10%) or ECG during Ic anti-arrhythmic treatment in one case. Among symptomatic patients (11, 21%) 3 died suddenly (SD); sustained ventricular tachycardia (sVT) and syncopal episodes were documented in one and 7 cases respectively. Mean age at first event was 9 +/- 6 years. Two of the patients with SD had spontaneous type 1 Br ECG in multiple leads and homozygous SCN5A mutations. Type 1 pattern was identified in 30/53 (57%), in 10 during fever; supraventricular arrhythmias were documented in 3 patients (2 atrial fibrillation and one atrial ectopic tachycardia). Genetic testing was performed in 30 cases (57%) and SCN5A mutation was identified in 15 (50%). During follow-up (49 +/- 56 months) 6 patients experienced a new syncopal episode. 7 patients received Hydroquinidine therapy, 4 of them were symptomatic (sVT, syncope) and 3 were asymptomatic with spontaneous type 1 ECG. No patient had symptoms during follow-up. Among symptomatic patients, at presentation or during follow-up, 10/14 (71%) had a spontaneous type 1 Br ECG, while in asymptomatic patients type 1 was identified only in 20/39 (51%, $p< 0.001$).

Conclusions
Spontaneous type 1 Brugada ECG seems to be more frequent in symptomatic patients and SCN5A mutation has been found in a higher proportion of patients than in adults.