Presentation and follow up of Brugada Syndrome in children.

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Background
Brugada syndrome (BrS) is a congenital channelopathy that usually presents in adulthood. Childhood characteristics are less well-known, impetus for this report.

Methods
Data were collected retrospectively from 1996 and prospectively from 2006 by the Brugada team of the University Hospital of Brussels. Patients aged ≤16 years with documentation of spontaneous or drug induced coved type 1 Brugada ECG pattern were included.

Results
Inclusion criteria were met for 43 patients. Median follow up was 2 1/2 years (1 month to 172 months). Six patients were lost from follow up.
Seventeen patients were symptomatic at presentation (M 12/ F 6). Median age was 10 years (7 months to 16 years). Presenting symptoms were: syncope (13 patients - six having an associated conduction disorder) and (aborted) sudden cardiac death (4). Abrupt syncope occurred in 10 cases. In the remaining 3, cause was less clear and implantable cardioverter defibrillator (ICD) implantation was recommended on individual basis, considering the familial background and the repetitive character of the syncope. The circumstances at the occurrence of the symptoms were extremely variable. An ICD was implanted in 15 patients. Appropriate shocks were administered in three patients. Three patients received inappropriate shocks.
Screening of asymptomatic family members revealed 27 patients with BrS. Five were subsequently symptomatic. Three patients had vasovagal syncope so no ICD was recommended. Two patients were referred for ICD implantation.
Ajmaline testing precipitated ventricular fibrillation in 3 patients, requiring CPR.
An electrophysiological study was performed in 28 patients of whom 17 were asymptomatic family members. VT was non inducible in any of the asymptomatic patients, although inducible in 2 out of eleven symptomatic patients.

Conclusion
Symptomatic patients were mostly ≤ 12-year-old boys. Indication for ICD implantation may be tasking, as most frequent presentation was syncope. Fever did not stand out as a precipitating factor. Ajmaline infusion is potentially dangerous and should be reserved for academic settings. 1/3 was previously diagnosed with atrial conduction disturbances, such as sick sinus syndrome or atrial standstills, suggesting a common denominator. Clinical follow up of asymptomatic family members is useful.