

## COMMUNICATIONS ORALES

Samedi 21 novembre de 8 h 30 à 10 h 15

## CO 1

**Pediatric open heart surgery in emerging countries: A 10-year experience at the Maputo Heart Institute, Mozambique**

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**Background** The Chain of hope acts since 1988 to free operate African, Asian and Middle East children with heart diseases. This study aimed to assess the feasibility and results of a 10-year open-heart surgery program at the Maputo Heart Institute, Mozambique.

**Methods and results** Data of all consecutive patients with history of at least one heart surgery performed at the Maputo Heart Institute between Jan. 2001 and Dec. 2011 were opening were analyzed, in regard peri-operative (in-hospital) as well as long-term follow-up. Overall, 891 operations were performed in 776 patients, including 93% performed exclusively by one of the 7 international teams and 7% by Mozambican surgeons alone. The mean follow-up was 2.7±2.8 years (1 month – 11 years). The mean age of patients was 15±6 years, with almost one third of patients living at more than 1000km from the heart institute. Principal indications of surgery was congenital heart disease in 47% (14.1% of ventricular septal defect, 13.4% of tetralogies of Fallot, 5.8% of atrial septal defect, 5.3% of ductus arteriosus and 7.4% of more complex congenital heart diseases), rheumatic heart disease in 33%, endomyocardial fibrosis in 8%. Mean hospital stay was 8 days±12 days. Peri-operative mortality rate was 5.9%, with significant difference between indications: 6.6% for congenital heart diseases, 2.3% for RHD and 6.8% for EMF (P=0.02). Among patients with mechanical prostheses, the mortality associated with the prosthesis was high (18.3%). More than half of patients were lost of follow-up at 3-year, and prevention of rheumatic heart disease non optimal for many children...

**Conclusion** Our findings suggest the feasibility of such a program, with development of local competences and finally the set-up of an independent surgical team with effective intensive care unit. However, our results also emphasize some weaknesses, especially the issue of follow-up of patients who would benefited the most of long-term preventive measures.

**Conflict of interest** The authors have not transmitted any conflicts of interest.

## CO 2

**The impact of clinical and genetic findings on the management of young Brugada syndrome patients**

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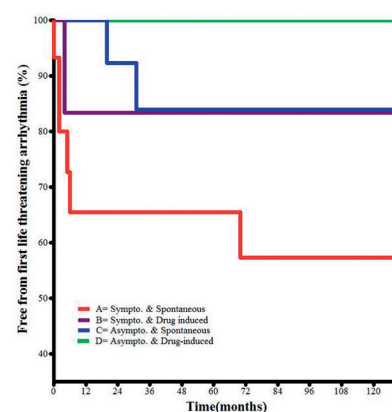
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**Aims** Brugada Syndrome (BrS) is an arrhythmogenic disease associated with sudden cardiac death (SCD) which seldom manifests and is recognized in childhood. We aim to describe the pediatric BrS clinical presentation to identify prognostic factors for risk stratification, and to propose a data-based approach management.

**Methods and results** We studied 106 patients, under 19 years of age at diagnosis with spontaneous (n=36) or drug-induced (n=70) BrS from 16 European hospitals. At diagnosis, mean age was 11.1±5.7 years and most patients were asymptomatic [family screening (n=67), incidental (n=13)] while 15 had experienced syncope, 6 aborted SCD or symptomatic ventricular tachycardia, 2 supra-ventricular tachycardia (SVT), 3 palpitations or presyncope. During follow-up (median: 54 months), 10 patients had life-threatening arrhythmias (LTA) including 3 deaths. Six experienced syncope and 4 SVT. Fever triggered 27% of LTA events. An ICD was implanted in 22 with major adverse events in 41%. Of the 11 patients treated with hydroquinidine, 8 remained asymptomatic. Genetic testing was performed in 75 patients and SCN5A rare variants were identified in 58; among the 32 pediatric probands tested 15 were genotype positive. Of the 10 patients with LTA the 9 with genetic testing were all genotype positive whereas the 17 SCN5A negative patients remained asymptomatic. Spontaneous BrS type 1 ECG (p=0.005) and symptoms at diagnosis (p=0.0015) were predictors of LTA. Time to the first LTA event was shorter in patients with both symptoms at diagnosis and spontaneous BrS type 1 ECG pattern (p=0.01) (figure 1).

**Conclusions** Spontaneous type 1 ECG and symptoms at diagnosis are predictors of LTA events in the young affected by BrS. The management of BrS should become age-specific and prevention of SCD may involve genetic testing, aggressive use of anti-pyretics and quinidine with risk-specific consideration for the ICD.

**Conflict of interest** The authors have not transmitted any conflicts of interest.



**Abstract C02 – Figure 1: Kaplan-Meier curves of LTA events during follow-up in four different groups (global p-value =0.01).**