Experience with foetal supraventricular arrhythmias

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This study was to review experience and outcomes of supraventricular (SV) arrhythmias in fetus Methods: Cases were divided in groups: SVPB= premature SV beats, NSSVT= non-sustained SV tachycardia, SSVT= sustained SV tachycardia, and AF= atrial flutter. Heart failure (HF) was defined as foetal hydrops or isolated effusion (pericardial or pleural or ascitis). Outcome was favourable if arrhythmia resolved or stabilized until full-term birth, not-favourable if premature birth or foetal death occurred.

Results: 188 fetuses were included: 89 in SVPB (47.3%), 31 in NSSVT (16.5%), 60 in SSVT (31.9%), 8 in AF (4.3%), aged at diagnosis 30.8±4.5 weeks (no difference between groups). Foetus HR at diagnosis was 241±30 bpm in SSVT vs 226±26 in AF. Antiarrhythmic therapy was administered in sustained tachycardia (83% of SSVT and 71% AF): 28 had 1 medication, 25: 2 medications, 2: 3 medications. Complication occurred in 29 cases, all in SSVT and AF (29 of 64±43%): 18 hydrops, 5 ascitis, 4 pericarditis, 1 pleural effusion and 1 LV dysfunction+MR, was more frequent in SSVT (86%: hydrops in 30%) than in AF (51%: no hydrops), p=0.08. Fetal HF was associated with HR at diagnosis: 251±25 bpm in hydrops vs 228±31 bpm in no-hydrops (p=0.025). Outcome was favourable in SVPB and NSSVT, in 45 of SSVT+AF (79%): Tachycardia resolved in 36, more frequently in SSVT (57%) than in AF (25%). HR only decreased in 9 cases. Premature birth occurred in 10% foetal death in 2. Outcome was not associated with HR or weeks of gestation at diagnosis. Defavourable outcome was more frequent in hydrops or isolated effusion (57%) than in uncomplicated cases (10%, p=0.0002). Resolution occurred in 45% hydrops vs 66% of non-hydrops cases. Digoxine decreased from 79% of cases before 2000 to 33% after 2000, while flecaïne increased from 14% to 48.5%. There was no relationship between therapy or number of medications and outcomes.

Conclusion: Fetal SSVT more frequently resolves but has worse outcome than AF, especially if HR at diagnosis is high and hydrops occurs. Larger scale prospective studies are needed to evaluate the efficacy of flecaïne compared to digoxine therapy.

Risk markers of cardiac events in patients with Marfan syndrome diagnosed during childhood

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Risk markers of cardio-vascular events in children with Marfan syndrome remain little known. Aortic root z-score measurements have been recently updated. We assessed the prognostic value of aortic root z-score in patients with Marfan syndrome diagnosed during childhood.

Methods: From the French multicenter database, 457 patients with Marfan syndrome, diagnosed before 18 y.o., without a history of cardiac event were prospectively included in this cohort study. Echocardiographic measurements of aortic root diameters were performed at each visit. We calculated the Z-score of aortic root measurements using the Bichat formula for each visit. Mean z-score was defined as the mean of the z-score (MeanZS15) calculated for each measurement before the age of 16.

Results: Median age at end of FU was 15.9 years (interquartile 10.9-20.3). FU was complete for 69.5% of patients. Median FU was 4.6 years. A cardiovascular event occurred in 17 patients (3.7%, prophylactic aortic surgery n=14, aortic dissection n=1 and deaths n=2). Survival free of cardiac events was 85.1% in patients with a meanZS15 of the Valsalva diameter <3 and 56.4% in patients with a meanZS15 of the Valsalva diameter ≥3 (p<0.0001 by log-rank test). In univariate analysis, Valsalva meanZS15 ≥3, age at inclusion in the database, a lower heart rate and an increased arm/height ratio were associated with an increasing risk of cardiac events (p=0.0001, p=0.04, p=0.01 and p=0.04 respectively). After multivariate adjustment using a cox proportional hazards model, only Valsalva meanZS15 ≥3 and lack of FBN1 or TGBB3 mutation identified were associated with an increasing risk of cardiac events (p=0.0001 and p=0.04 respectively).

Conclusion: Valsalva meanZS15 may help to predict whose children with a Marfan syndrome will have a cardio-vascular event. Therefore a particular attention to beta-blockade treatment observance and a regular follow-up may be suggested in high-risk children.

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