Background: Williams-Beuren syndrome (WS) is an autosomal dominant disorder characterized by dysmorphic faces, cardiovascular diseases, mental retardation and idiopathic hypercalcemia. Cardiovascular defects are found in up to 80% of the pts, the most common of which is supravalvular aortic stenosis, being present in 48.8% of pts. Other frequent cardiovascular manifestations are stenosis of pulmonary arteries, mitral valve anomalies, stenosis of peripheral arteries, atrial and ventricular septal defects. Only little information exists about the occurrence of arrhythmias in the setting of this syndrome.

Methods: Data about WS pts were retrospectively revised with the aim to identify conditions that required invasive procedures for arrhythmic events.

Results: In the last twelve months, two patients affected by WS underwent radiofrequency catheter ablation (RFCA) for atrio-ventricular nodal re-entry tachycardia (AVNRT) at our institution. Pt #1, a 17 years old boy with a familiar history of thalassemia and thyropathy, suffered from recurrent episodes of palpitations, requiring repeated admission in emergency. In both patients slow-fast AVNRT was induced during electrophysiologic study with a single extrastimuli protocol (cycle length 322±29.6msec). RFCA of slow pathway was successfully performed in both pts, without any complications.

Conclusions: Besides congenital heart disease, potential morbidity in WS pts can derive also from AVNRT.

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Atrioventricular nodal re-entry tachycardia: a potential morbidity condition in Williams syndrome

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In the last twelve months, two patients affected by WS underwent radiofrequency catheter ablation (RFCA) for atrio-ventricular nodal re-entry tachycardia (AVNRT) at our institution. Pt #1, a 17 years old boy with a familiar history of thalassemia and thyropathy, suffered from recurrent episodes of palpitations, requiring repeated admission in emergency. In both patients slow-fast AVNRT was induced during electrophysiologic study with a single extrastimuli protocol (cycle length 322±29.6msec). RFCA of slow pathway was successfully performed in both pts, without any complications.

Conclusions: Besides congenital heart disease, potential morbidity in WS pts can derive also from AVNRT.

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Pre-ablation determination of expected power output as indication of lesion size during radiofrequency ablation: verification of an experimental algorithm applied in patients with accessory pathways

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Purpose: to develop an algorithm determining the target temperature (tp) needed for a desired lesion size for any obtained catheter position, by measuring the tp rise to a test pulse (dT) and correlate this to power-output when ablating accessory pathways.

Methods: experimental model: in pigs the catheter tip was positioned in the left ventricle and 0.6 W applied for 5 s and ablation then performed with unchanged tp position. Development of algorithm: dT was measured and ablation then performed at target tp 70 or 60°C and volume determined. Regression analysis between dT and lesion volume was performed and curves extrapolated to targettps 55-85°C. Testing of algorithm: A target volume of 300 mm³ was tested by measuring dT and then ablating with the target tp suggested by the algorithm. Clinical testing: in patients with accessory pathways pre-ablation test pulse was applied and power output during ablation at target tp 70°C was registered.

Results: the algorithm was developed from 34 lesions. Volume ranged from 13-1440 mm³. There was a significant negative correlation between dT and volume for 70°C and 60°C (r = 0.82 and 0.74), and between dT and power output r = 0.97 for target tp 70°C. The algorithm was tested in 6 animals (15 lesions), target volume 300 mm³. Target tp ranged from 62-85°C. Lesion volume was 310±93 mm³. The variance on this lesion volume was significantly smaller (p<0.02) than the variance on lesion volume for target tp of 70°C. Clinical testing was done in 5 patients, 9 test pulses. dT ranged from 0.1-0.9°C. A significant negative correlation was found between dT and power-output, r = 0.73, not significantly different from the correlation found in the animal experiments.

Conclusion: the tp rise to a pre-ablation test pulse can evaluate the impact of electrode tissue contact and cavitory cooling on lesion size. Using this information to choose a target tp allows creation of a more controllable lesion size. Preliminary clinical testing suggests that a similar relation between dT and ablation parameters is present during ablation of accessory pathways in humans.