

Combination of sinus node dysfunction and intestinal pseudo-obstruction in children and young adults: about 11 cases of this undescribed association

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INTRODUCTION: Sinus node dysfunction (SND) is rare in children and is encountered mainly as a late complication of congenital heart surgery. It is sometimes found in patients with a structurally normal heart and can be inherited. Chronic intestinal pseudo obstruction (CIPO) is a rare disabling isolated condition in children. It is idiopathic in most cases. The combination of CIPO and SND has not been described.

RESULTS: We report 11 French Canadian children (5) and young adults (6) that manifested a combination of symptomatic CIPO and SND (between 01/1998 and 03/2010).

In the pediatric population (non related patients), first signs of CIPO developed at mean age of 11.1 yrs (6.6-17yrs). CIPO preceded SND by 4.2 and 5.5 yrs in 2 patients, CIPO and SND were simultaneous in 2 and SND preceded CIPO by 2 yrs in 1 patient. There were 4/5 boys. Three patients with severe symptomatic bradycardia, junctional rhythm and pauses required a pacemaker (PM) 1-6 months post diagnosis. They remain well 0.5-7.5 yrs post PM implant. The other 2 patients manifested tachy/brady syndrome requiring antitachycardia medication and did not need a PM. The 5 patients had dilated left atrium and ventricle on echo with normal systolic function, normal anatomy and no mitral regurgitation. One patient had left diastolic dysfunction. One patient died at 18.8 yrs in the post-operative period of multiorgan transplantation. During the same period, 6 other patients (2/6 boys), were diagnosed with CIPO in our institution without manifestations of SND. No family members of our affected pediatric patients suffered of CIPO or SND.

Among the adults (mean age 35yrs, range 23-43yrs; 4/6 men), all have CIPO (all diagnosed before age of 15yrs) and have cardiac symptoms. All but 2 have PMs and tachycardias and the 2 others have sinus bradycardia and supra-ventricular tachycardias. Within these adults, there are 2 pairs of siblings. Genetic testings of patients and families are currently underway.

CONCLUSIONS: SND developed in 11 patients diagnosed with CIPO in our institution, mainly during childhood (7/11) or young adulthood (4/11). Genetic investigations are currently underway and a founder effect is so far most probable.