

Comorbidity and long-term outcome in patients with congenital heart block and their siblings exposed to Ro/SSA autoantibodies in utero

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Objective: Congenital heart block (CHB) may develop in fetuses of Ro/SSA autoantibody-positive women. Given the rarity of CHB, information on comorbidity and complications later in life is difficult to systematically collect for large groups of patients. We therefore used nation-wide health care registers to investigate comorbidity and outcomes in patients with CHB and their siblings.

Methods: Data from patients with CHB (n=119) and their siblings (n=128), all born to anti-Ro/SSA-positive mothers, and from matched healthy controls (n=1,190) and their siblings (n=1,071), were retrieved from the Swedish National Patient Register. Analyses were performed by Cox proportional hazard modeling.

Results: Individuals with CHB had a significantly increased risk of cardiovascular comorbidity, with cardiomyopathy and/or heart failure observed in 20 (16.8%) patients versus 3 (0.3%) controls, yielding a hazard ratio (HR) of 70.0 (95% CI 20.8-235.4), and with a HR for cerebral infarction of 39.9 (95% CI 4.5-357.3). Patients with CHB also had a higher risk of infections. Pacemaker treatment was associated with a decreased risk of cerebral infarction but increased risks of cardiomyopathy/heart failure and infection. The risk of systemic connective tissue disorder was also increased in patients with CHB (HR 11.8, 95% CI 4.0-11.8), and both patients with CHB and their siblings had an increased risk to develop any of 15 common autoimmune conditions (HR 5.7, 95% CI 2.83-11.69 and 3.6, 95% CI 1.7-8.0, respectively).

Conclusions: The data indicate an increased risk of several cardiovascular, infectious and autoimmune diseases in patients with CHB, with the latter risk shared by their siblings.

Key words: Congenital heart block, pacemaker, SLE, Sjögren's syndrome