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

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General aim
What are the health-related implications for patients with autoantibody-mediated congenital heart block?

Background
Ro/SSA autoantibodies are commonly present in women with Sjögren’s syndrome. During pregnancy, the autoantibodies are transported across the placenta and are associated with a risk of congenital heart block (CHB) in the baby. CHB carries substantial morbidity and mortality; affected children generally require pacemaker implantation. However, little is known regarding health-related implications in later life in this group of patients.

Study design
Medical records (1987-2010) for a cohort of patients with CHB (n = 119) born to Ro/SSA-positive mothers and their siblings (n = 128) were retrieved from the Swedish national patient register. Data was also extracted for matched healthy controls (n = 1,190) and their siblings (n=1,071). Cox proportional hazard modeling was used to estimate the risk of diseases for patients with CHB and their siblings.

Results
Individuals with CHB had significantly increased risks of arrhythmias, cardiomyopathy and/or heart failure. Moreover, the hazard ratio of cerebral infarction in patients with CHB was 39.9 (95% CI: 4.5–357.3). Patients with CHB also had elevated hazard ratios for infections, which was partly explained by pacemaker implants. Both patients with CHB and their siblings had an increased risk to develop autoimmune diseases with hazard ratios of 5.7 (95% CI: 2.8-11.6) and 3.6 (1.7-8.0), respectively.

Conclusion
Our findings suggest that patients with CHB carry a risk of disease of several organs and etiology greater than the general population. This increased morbidity is present in several areas, but most apparent within cardiovascular and inflammatory diseases, of which the latter risk is also shared with their siblings.

Atrial fibrillation and flutter 46.7 (10.1–216.1)
Heart failure 34.4 (9.5–125.2)
Cerebral infarction 39.9 (4.5–357.3)
Diseases of arteries, arterioles and capillaries 7.6 (1.7–34)

Figure 1 Cohort design

Figure 2 Hazard ratios (95% CI) of cardiovascular disease for CHB patients

Figure 3 Cumulative hazard of developing autoimmune disease (right side figures denote hazard ratios (95% CI))