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**Department of Medicine, Solna**

## **Pregnancy in Ro/SSA positive women**

**AKADEMISK AVHANDLING**

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## ABSTRACT

Congenital heart block (CHB) may develop in the fetus during pregnancy in Ro/SSA positive women. It is a rare but life-threatening disease and the most serious manifestation of neonatal lupus erythematosus (NLE). The frequency of CHB in Ro/SSA autoantibody positive pregnancy is 1-2%, with a recurrence rate of 12-17%. Development of CHB may be monitored by Doppler/echocardiography examinations during gestational week (GW) 18-24 when early signs of CHB are most likely to occur. The surveillance enables early treatment and may reverse the inflammatory process in the fetal heart. Although this has been known for a decade, little is known about how Ro/SSA autoantibody positive mothers experience their pregnancy when being at risk for having a child with CHB or actually give birth to a child with CHB. Furthermore little is known about the neurodevelopment in the children born with CHB. The aim of this thesis was therefore to investigate Ro/SSA autoantibody positive women's experience of pregnancy and the postnatal period as well as neurodevelopment in children with CHB.

A main finding of the studies is that the awareness and knowledge of Ro/SSA autoantibodies and associated risk for CHB is low both among health care personnel as well as women/families involved. Further, there is a lack of adequate and sufficient information available. The majority of the participants in the study received the information of their own autoantibody positivity as well as the connection to risk for CHB when they were already pregnant. Most women stated that they wanted the information as soon as the autoantibody positivity was known, if possible before pregnancy in order to prepare and adjust to the situation. The majority of the women also stated that the information would not have influence their decision of becoming pregnant. A majority of the women giving birth to children with CHB stated that they did not receive enough information. Our studies showed that the informant was an important factor predicting how the women experienced the information, where information communicated by someone with specialization in pediatric cardiology was significantly more often experienced as sufficient and understood by the women. The serial Doppler/echocardiography examinations did not induce anxiety, but rather coming to a highly specialized center and getting the possibility to interact and to get information and support from the staff was experienced as a benefit by the women. Women giving birth to a child with CHB often wished for more support. When investigating neurodevelopment, our data indicate that in addition to well established factors such as male sex and being born preterm, both maternal systemic lupus erythematosus (SLE) and CHB may influence neurodevelopment.

In conclusion, the result from this study highlights the vulnerable situation that women and their families experience when there is a risk for the child to develop CHB. We conclude that it is important to develop structured programmes for the surveillance of pregnancy in women who are SSA/Ro52 positive and to refer these women to specialized centers where the experience to manage the situation and with the possibility of maintaining updated information, surveillance and treatment is available. Such programme should also include guidelines for the involved personnel in the chain of care and make relevant information accessible for the women/families. Offering professional psychological support to the women and families should also be considered. In particular, for the women that gave birth to a child with CHB, the surveillance programme should range into the post partum period, and also involve the primary child care center to bridge the gap between different care givers. In addition, follow up of neurodevelopment should be considered for children with CHB, especially if the mother is diagnosed with SLE. An early diagnosis is one way to help these children overcome their difficulties during childhood and school years and make sure that they obtain the support needed.