

Cardiac Neonatal Lupus: Long-Term Data Show More Problems with Aging

Written by Wayne Kuznar

According to long-term follow-up of patients from the Research Registry for Neonatal Lupus (RRNL), patients with cardiac manifestations of neonatal lupus (NL) are more likely to develop an enlarged heart and the need for pacemaker replacement as they age. The findings emphasize the need for continued long-term cardiac evaluation of patients with cardiac-NL. They were presented by Amit Saxena, MD, New York University School of Medicine, New York, New York, USA, the lead investigator of the analysis [Saxena A et al. ACR 2012 Poster 290].

NL is a model of passively acquired autoimmunity. It occurs when anti-SSA/Ro and/or SSB/La antibodies cross the placenta and make their way into the fetal circulation. The greatest morbidity and mortality occurs from irreversible damage to the heart, for example due to congenital heart block or cardiomyopathy, said Dr. Saxena. Long-term cardiac and rheumatologic outcomes as well as those related to neurodevelopment have not been formally assessed.

Family members of 75 children from the RRNL with cardiac-NL completed questionnaires, as did 35 families of children with cutaneous NL and 74 unaffected siblings. Twenty-four percent of the children were 0 to 5 years old at latest follow-up, 27% were 5 to 10 years old, 11% were 10 to 15 years old, 21% were 15 to 20 years old, and 17% were >20 years old. Eighty-five percent of the children had third-degree heart block.

Questionnaire responses revealed that 74% of the children had a pacemaker implanted and 65% needed a replacement pacemaker. Forty-one percent of family members had ever been told that their child had a dilated heart and 20% had ever been told that their child had congestive heart failure. One child had undergone heart transplant.

When cardiac outcomes were examined by age group, older age was associated with a reported history of an enlarged heart ($p=0.016$), a greater need for pacing ($p=0.001$), and a greater likelihood of having a pacemaker replaced ($p=0.003$; Table 1). Of the 37 children with cardiac-NL who were aged >10 years, 19% required cardiac medications (ie, digoxin, angiotensin-converting enzyme inhibitors, or β -blockers) compared with 5% of those aged <10 years ($p=0.086$).

Table 1. Long-Term Data May Show More Problems with Aging.

Age (years)	n	Total CHF	TDH*	Paced*	PPM Replaced*	Cardiac Meds†
0-5	18	4 (22%)	5 (28%)	10/17 (59%)	0 (0%)	1 (6%)
5-10	20	2 (10%)	4 (20%)	11/17 (65%)	8/11 (73%)	1/18 (6%)
10-15	8	3 (38%)	6 (75%)	6 (75%)	5/6 (83%)	2/7 (29%)
15-20	16	3 (19%)	9 (56%)	13 (81%)	10/12 (83%)	2 (13%)
>20	13	3 (23%)	7 (54%)	13 (100%)	9/10 (90%)	3/11 (27%)

*Median age higher in those reporting enlarged heart ($p=0.016$), needing a PPM ($p=0.001$), and PPM replacement ($p=0.003$). †Of 37 children >10 years, 19% required cardiac medicine (digoxin, ACE inhibitor, or β blocker) versus 5% of 36 children <10 years ($p=0.086$). CHF=congestive heart failure; PPM=permanent pacemaker; TDH=total dilated heart.

Children who had dilated cardiomyopathy or valvular disease on their fetal echocardiogram had significantly more pacemakers replaced as they got older compared with those without these features on their fetal echocardiogram ($p<0.05$).

Twelve percent of children with cardiac-NL had delayed motor milestones compared with only 3% of unaffected siblings ($p=0.027$), but there was no difference in intellectual delay between these groups (19% vs 15%, respectively).

Fluorinated Steroids in Cardiac Neonatal Lupus Do Not Impact Survival

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Exposure to fluorinated steroids does not seem to have an impact on fetal survival in cardiac neonatal lupus (NL), with the possible exception of fetuses with hydrops. Available data are discordant regarding the efficacy of fluorinated steroids in the prevention of fetal mortality in cardiac-NL. In a study in which patients with fetal complete atrioventricular (AV) block were treated with dexamethasone, mortality at 1 year was 10%, compared with a mortality rate of 54% in historical controls, although the historical controls used clearly had much higher rates of poor prognostic factors [Jaeggi ET et al. *Circulation* 2004]. In contrast, no significant effect of treatment with fluorinated steroids on mortality was observed in a retrospective multicenter study of 175 fetuses diagnosed with second- or third-degree AV block [Eliasson H et al. *Circulation* 2011].

Similarly, the available data on the prevention of cardiac-NL with the use of fluorinated steroids are limited, with one study that showed effective prevention of congenital