Left atrial inexcitability in pediatric patients with congenital lupus induced complete atrioventricular block

Abadir S. (1,2), Vobecky S.J. (1) Rohlicek C. (2) Fournier A. (1)
CHU Sainte Justine, Montreal, Canada (1)
Montreal Children’s Hospital, Montreal, Canada (2)

Introduction
Atrial standstill and atrioventricular (AV) conduction blocks have been described in systemic lupus erythematosus adult patients and older children. In newborns from women with anti-Ro/SSA antibodies, AV block and ventricular cardiomyopathy are well known immunologic complications; to our knowledge, no cases of atrial inexcitability or atrial standstill have been described in this setting.

Methods
We recently encountered four pediatric patients with maternal lupus induced complete AV block meeting the criteria for cardiac pacing, who demonstrated left atrial inexcitability and/or interatrial conduction block. Clinical and pacemaker data, electrocardiograms and echocardiograms were reviewed.

Results
Diagnosis of maternal lupus induced AV block was made prenatally (n=2), at birth (n=1) and at 34 months (n=1). All 4 female patients underwent epicardial dual chamber pacemaker implantation (mean age 28.5 ± 25.4 months) using steroid eluting leads, through a left thoracotomy. In 3 patients, no appropriate left atrial appendage (LAA) or left atrial capture could be achieved despite high outputs. The atrial lead was thus fixed on the right atrial appendage (RAA), along with a left ventricular (LV) lead. LAA biopsy performed in one patient was unremarkable. One patient developed cardiomyopathy soon after pacing was initiated. Upgrade to a biventricular pacemaker system did not improve function. This patient is awaiting heart transplant.
The fourth patient underwent LAA and LV lead placement. Follow up demonstrated early LV dysfunction and increased delay between surface P wave and intracardiac atrial depolarisation raising suspicion for interatrial conduction delay. The pacemaker was upgraded to a biventricular system, the LAA lead moved to the RAA, resulting in LV function improvement.

Conclusion
In patients with congenital lupus induced complete AV block, pacemaker implantation can be challenging due to left atrial inexcitability and/or interatrial conduction delay, suggesting an associated atrial myopathy, yet to be characterized. Longer follow up will be needed to better understand the true significance of this finding in the setting of congenital lupus induced complete AV block. Further preoperative delineation should be done, possibly using echocardiography, to guide surgical approach.