Maternal derived anti SS-A antibody as a possible etiology of secondary dilated cardiomyopathy associated with congenital left bundle branch block.

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Introduction: Maternal derived anti SS-A antibody affects the fetal heart by initiating a series of events leading to cardiac inflammation, fibrosis and calcification, and eventual blockage of signal conduction at the atrio-ventricular node. Herein we report two infants with dilated cardiomyopathy with left bundle branch block (LBBB) associated with maternal derived anti SS-A antibody in whom clinical status dramatically improved after cardiac re-synchronization therapy (CRT).

Methods: We present two cases of infantile cardiomyopathy with LBBB.

Results: Case 1; A 5 months-old baby was addressed with severe heart failure and a 1 month history of poor feeding and failure to thrive (body weight; 4kg). He was referred to our hospital after the medical treatment of acute heart failure for further management of cardiomyopathy. The electrocardiography (ECG) revealed LBBB pattern with QRS duration of 140 ms and the echocardiography showed a typical septal flash (Fig. 1). He underwent CRT pacemaker implantation by thoracotomy. 7 weeks after the CRT, B-type natriuretic peptide decreased to 38 from 2,200 pg/ml and left ventricular ejection fraction improved to 55 from 19%. His ECG at the age of 7 days of life also revealed LBBB pattern with QRS duration of 100 ms. The titer of the anti SS-A antibody was 63 U/ml, and his mother’s titer was over 10,000 U/ml. Case 2; A 3 months baby was referred to our hospital after initial therapy for cardiogenic shock. The ECG revealed LBBB pattern with QRS duration of 120 ms and the echocardiography showed a typical septal flash (Fig 1). She underwent CRT pacemaker implantation by thoracotomy after break away from DIC. After CRT, we successfully weaned off the mechanical ventilation. And now we started medical treatment for chronic heart failure. The titer of the anti SS-A antibody was 299 U/ml, and her mother’s titer was over 10,000 U/ml.

Conclusions: Congenital LBBB associated with maternal derived anti SS-A antibody may cause mechanical ventricular dyssynchrony and heart failure. That means the electrical and mechanical re-synchronization of the ventricle improves clinical status as well as cardiac function in these infants.