

Dizygotic twin with congenital AV block

To the Editor,

The incidence of congenital atrioventricular (AV) block is estimated to be about 1 in 15,000-20,000 births. Although it is a rare disease, it increases fetal, neonatal, and childhood morbidity and mortality, and 60%-65% of all cases are reported to require pacemaker implantation before reaching adulthood (1). This disease can be idiopathic, but it is mostly caused by transmission of auto-antibodies, such as anti-Ro/SS-A and anti-La/SS-B antibodies, from mothers affected by an autoimmune disease, which damages the cardiac conduction system. Half of these mothers are asymptomatic or are not diagnosed until after their delivery.

Dizygotic twin sisters, aged 22 years with congenital AV block and a permanent pacemaker implanted in their adolescence, presented to our hospital for a routine check-up. A two-chamber permanent pacemaker was implanted in twin A and was left in DDD mode. A two-chamber permanent pacemaker was also implanted in twin B, but it was left in VDD mode. The electrocardiograms showed that the pacemaker rhythm (atrial sense, ventricular pacing) and transthoracic echocardiograms of both twins were normal. Twin A had a history of removal of the pacemaker due to infection, implantation of a new pacemaker 4 years ago, and coil embolization of patent ductus arteriosus (PDA) 3 years ago; otherwise, both twins were healthy.

Complete heart block can occur as an isolated entity, or it can accompany other congenital heart defects, like transposition of great arteries, PDA, or atrial septal defect (2). The history of PDA in one of the twins may suggest maternal infection or drug abuse during pregnancy, but there was no evidence to support this hypothesis. In addition, the mother of the twins did not have any autoimmune disease. Her anti-Ro/SS-A and anti-La/SS-B antibodies were negative. Herein, we present the first dizygotic twins in the literature with congenital AV block. Killen et al. (3) reported a similar case in chorionic diamniotic twins exposed to maternal anti-Ro/SS-A and anti-La/SS-B antibodies. However, in that report, one twin had sinus rhythm and the other had Mobitz type 1 second-degree AV block (Wenckebach). The treatment of congenital heart block includes intrauterine steroid (4) and intravenous immunoglobulin (IVIG) (5) and implantation of a permanent pacemaker after birth.

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