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ST segment depression after Norwood/systemic-pulmonary artery shunt

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A three-month-old girl with double inlet left ventricle (S;D;D), hypoplastic outlet ventricle, restrictive bulboventricular foramen, d-transposition of great arteries and interrupted aortic arch underwent Norwood stage 1 operation, systemic to pulmonary artery (PA) shunt and atrial septectomy, and developed ectopic atrial tachycardia that responded to digoxin therapy. She developed fussiness lasting two hours, not associated with cyanosis, and not relieved by feeding. Her pulse oximetry was 88% while breathing room air, and she had no differential blood pressure gradient. Apart from single second heart sound and a grade IV/VI ejection systolic murmur, the examination was normal. Her electrocardiogram and echocardiogram were unchanged. A Holter monitor revealed ST segment depression and T wave inversion upon increasing the heart rate beyond 140 beats/minute (Figs. 1 and 2). A cardiac catheterization showed patent systemic to PA shunt, patent native aorta, and no evidence of coarctation of aorta. After undergoing the second stage operation, her diastolic runoff flow was normal.

Figure 1. Absence of ST segment depression with a heart rate <140 beats/minute in a patient with Norwood/systemic to pulmonary artery shunt with diastolic runoff flow.

Figure 2. ST segment depression and T wave inversion with a heart rate >140 beats/minute in a patient with Norwood/systemic to pulmonary artery shunt with diastolic runoff flow.
and symptoms of fussiness disappeared and a
surveillance Holter monitor showed better-
looking ST segments and T waves even at higher
heart rates (Figs. 3 and 4). Three months later,
the patient is doing well and is awaiting Fontan
completion.

Despite many advances in the field of pediatric
cardiology, the interstage mortality rate remains
high [1]. The use of the systemic to PA shunt in
the Norwood operation has been complicated by
wide pulse pressure, diastolic runoff flow [2,3],
even distribution of pulmonary blood flow [4],
and increased early mortality [5]. Providers need
to keep a high index of suspicion for the adverse
effects of the diastolic runoff that accompanies
the presence of systemic to PA shunts.

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