Noncompaction Cardiomyopathy in an Infant for Laparoscopic Surgery
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INTRODUCTION
Noncompaction cardiomyopathy is diagnosed by prominent ventricular trabeculations and deep recesses on echocardiogram. These trabeculations represent poor myocardial formation during embryologic development and can result in a dilated and hypertrophied heart. There is a varied clinical presentation ranging from stable cardiac function to frank heart failure. Symptoms can present at any time from newborn to adults, however, early presentation is associated with more severe cardiac dysfunction. The disease has only recently been identified as a distinct classification by the American Heart Association.

CASE
The patient is a 5 week old male infant with non-compaction cardiomyopathy presenting for laparoscopic pyloromyotomy. He was born at 37 weeks with no complications and discharged home after a routine stay. When he was 21 days old he became cyanotic and short of breath. He was brought to a community hospital and was diagnosed with cardiogenic shock, metabolic acidosis, and multi-organ dysfunction. The initial echocardiogram showed severely depressed left ventricular function with a right to left shunt through a patent PDA. His trachea was intubated and both a prostanladi E2 and milrinone infusion were initiated, which stabilized cardiac function. He underwent an evaluation for coexisting syndromes that was negative. He was transferred to Mount Sinai Medical Center for an evaluation as a potential cardiac transplant recipient. Furosemide and hydrochlorothiazide were started to treat moderate pulmonary edema. Oxygen delivery was supplemented through initiation of erythropoietin treatment and transfusions. The prostanladi E2 infusion was discontinued because subsequent echocardiograms showed an improvement in the right to left shunt. As the patient’s health stabilized, mechanical ventilation was no longer necessary. He had several bouts of non-bilious emesis after feeding which led to a diagnosis of pyloric stenosis. At the time of surgery he weighed 3.8 kg. His most recent cardiac evaluation indicated severe left ventricular dysfunction with a shortening fraction of 9%. On exam, he appeared adequately nourished and hydrated. He had breath sounds with crackles bilaterally. His abdomen was soft, non-tender to palpation, and slightly distended. His heart rate was 125, blood pressure 70/40, respiratory rate 28, and room air saturation 99%. A complete blood count and comprehensive metabolic panel were within normal limits.

DISCUSSION
Treatment for patients with noncompaction cardiomyopathy is dependent on clinical presentation. In this case, the patient was in cardiogenic shock and was treated with an inotropic infusion and diuretics. The patient was optimized prior to surgery as evident by his physical exam findings. He tolerated laparoscopic surgery well, after adjusting management for an increase in end tidal carbon dioxide and a drop in blood pressure. We chose to extubate the patient’s trachea because he was stable throughout the procedure. Pediatric cardiomyopathy is a relatively rare condition with an incidence of 1.13 cases per 100,000 children aged 18 years or younger. However, it is associated with high rates of death or transplantation. In this case, the patient needed to have his pyloric stenosis resolved prior to heart transplantation. He had no associated complications with laparoscopic pyloromyotomy and later received a heart transplant.

REFERENCES