

Multiple Cardiac Rhabdomyomas Causing Left Ventricular Outflow Obstruction

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Clinical Image

A term female infant was born via an urgent C-section for fetal arrhythmia. Postnatally, the heart rhythm was irregular and an EKG showed a normal sinus rhythm and frequent premature atrial complexes. There was cardiomegaly noted on a chest X-ray. A loud systolic murmur was appreciated. An echocardiogram showed multiple myocardial tubers (Figure 1). One tuber was attached to the septal leaflet of the tricuspid valve and another was located right below the pulmonary valve. However, the most significant one was located just below the aortic valve and measured 12×7 mm causing a moderate to severe left ventricular outflow tract obstruction (LVOTO) (50-60 mmHg peak gradient) (Figure 2). A T1 weighted brain MRI showed sub-ependymal hematomas (Figure 3) and cortical tubers consistent with tuberous sclerosis. This was further confirmed by gene sequencing showing a mutation in the *TSC-2* gene. The patient is being monitored closely as an outpatient for the possible deterioration of the LVOTO or the development of arrhythmias.

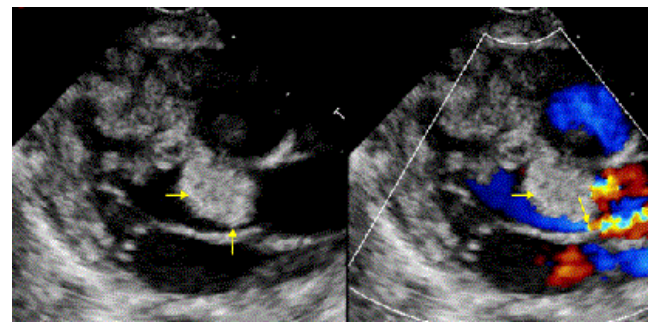


Figure 2: A parasternal long axis view.

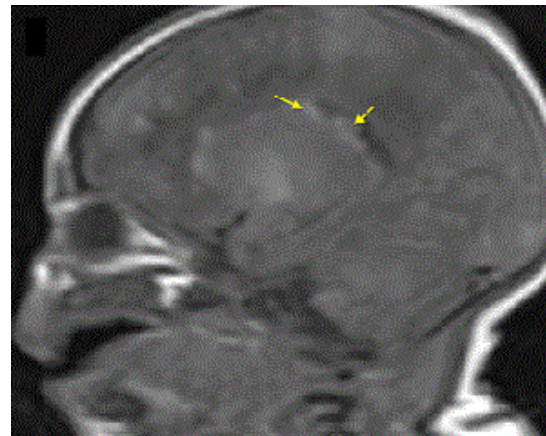


Figure 3: MRI showing sub-ependymal hematomas.

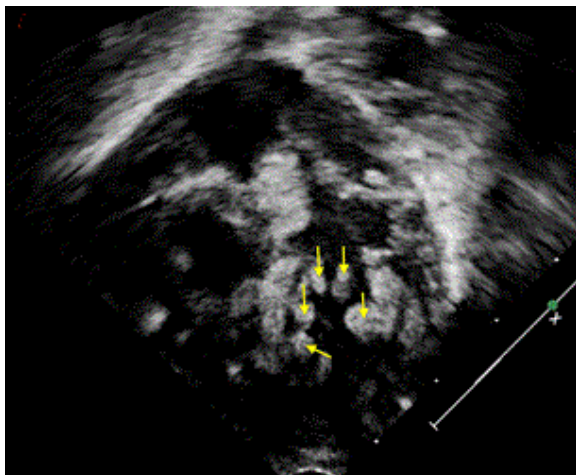


Figure 1: An echocardiogram showed multiple myocardial tubers.