Hybrid Procedure as Treatment for Large Obstructive Left Ventricular Rhabdomyoma
Amanda M. Marshall, MD, Mitchell I. Cohen, MD, Ashish B. Shah, MD, and Christopher L. Lindblade, MD
Phoenix Children’s Hospital/Maricopa Medical Center Residency Program; and Arizona Pediatric Cardiology Consultants, Phoenix, Arizona

We present one of the first reported cases of a neonate with a prenatal diagnosis of large left ventricular rhabdomyoma obstructing the outflow tract that underwent a hybrid procedure with stenting of the patent duct arteriosus and bilateral pulmonary artery banding.

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Cardiac rhabdomyomas are the most frequent neonatal tumors encountered and share an association with tuberous sclerosis in 50% to 80% of cases [1, 2]. The natural history is typically one of spontaneous regression; however, in patients with outflow tract obstruction or hemodynamic instability secondary to persistent ventricular arrhythmias, surgery to resect or partially debulk the tumor may be required [1, 3]. Left ventricular outflow tract (LVOT) obstruction accounts for 47% [4] of rhabdomyomas necessitating surgery, either as a complete or partial resection. However, progressive intrauterine obstruction resulting in single ventricle physiology with hypoplasia of the left ventricle is exceedingly rare. We present one of the first reported cases of a neonate with a prenatal diagnosis of large left ventricular rhabdomyoma obstructing the outflow tract that underwent a hybrid procedure with stenting of the patent duct arteriosus and bilateral pulmonary artery banding.

Prenatal echocardiogram of a 23-week gestational age male fetus revealed the presence of a large (1.2 × 1.5 cm), well-circumscribed, homogeneous cardiac mass in the ventricular septum obstructing left ventricular outflow with severely depressed left ventricular systolic function (Fig 1A). There was severe subaortic obstruction with retrograde flow demonstrated in the ascending aorta. There were also frequent conducted atrial extrasystoles observed on serial fetal echocardiograms. The patient was delivered by induced vaginal delivery at 38 weeks gestation at a tertiary care center, immediately started on prostaglandins, and transferred to the cardiovascular intensive care unit. Postnatal echocardiogram confirmed a large homogeneous mass (Fig 1B) creating severe left ventricular outflow tract obstruction; however, there was a minimal pressure gradient across the outflow tract in the presence of severely diminished left ventricular systolic function. There were multiple smaller echogenic masses within the left ventricular myocardium as well as the anterior wall of the right ventricle. The left ventricle was non-apex forming. Retrograde flow was present in the transverse aorta with mild hypoplasia of the ascending and transverse aorta. Postnatal cardiac magnetic resonance imaging revealed a 20.2 cm × 21.6 cm mass within the outflow tract (Fig 2) resulting in a left ventricular end diastolic volume of 23.6 mL/m² and a narrowed left ventricular outflow tract of 1.3 mm × 1.6 mm. Brain magnetic resonance imaging confirmed the presence of multiple subependymal nodules supporting the diagnosis of tuberous sclerosis. An electroencephalography at 7-days old showed abnormal background activity and 1 subclinical seizure indicative of poor neurologic prognosis with high risk of transforming into infantile spasms. Topiramate was initiated and no further clinical seizures were observed. An electroencephalography repeated at 4 months of age showed no evidence of subclinical seizures.

With the exception of intermittent premature atrial beats and bigeminy, no significant arrhythmias developed; however, the patient’s initially decreased left ventricular contractility worsened at 3 weeks of age with the development of significant pulmonary edema, decreased oxygen saturations, and poor renal oximetry. He underwent a hybrid procedure with patent ductus arteriosus stenting using a 6 mm × 15 mm Palmaz Genesis biliary stent (Cordis Corporation, Bridgewater, NJ) and atrial septostomy, both performed by cardiac catheterization, and surgical banding of the branch pulmonary arteries using 3-mm Gore-Tex strip (Gore, Flagstaff, AZ) to achieve acceptance of a 1.5-mm Hegar dilator. Post procedure, the patient required several days of inotropic support. Over the next 3 weeks the patient showed marked clinical improvement. However, a routine follow-up echocardiography performed demonstrated a new finding of severe right ventricular dysfunction, although the patient remained asymptomatic. The patient was started on epinephrine and milrinone and eventually transitioned to an oral anticongestive heart failure regimen. At the time of discharge at 45-days old, the echocardiogram revealed a non-apex forming LV with akinetic septal wall motion and moderately reduced global function. There was qualitatively more antegrade flow across the hypoplastic LVOT with echocardiographic evidence of mild tumor regression. The right ventricle remained dilated with severe systolic dysfunction. Over the next 5 months the patient thrived, with excellent weight gain and no evidence of clinical seizures. The right ventricular systolic function normalized on heart failure medications. The LV remained markedly diminutive and non-apex forming despite partial regression of the LVOT tumor. Given his improved right ventricular function and stable neurologic status, the patient underwent a successful hybrid takedown with creation of a right superior
vena cava to right pulmonary artery cavopulmonary anastomosis along with left pulmonary patch augmentation at 6 months of age.

Comment

Rhabdomyomas, as functional hamartomas, represent the most common primary neonatal cardiac tumor, with approximately 50% of cases undergoing spontaneous regression by age three years [2]. The estimated time before significant postnatal regression can be observed remains unknown. Resection of obstructive right ventricular tumors is possible through the tricuspid valve or a limited ventriculotomy. Tumors obstructing the LVOT are more surgically challenging given the relatively small neonatal aortic annulus and risk of injury to the conduction system. Other less desirable antegrade surgical approaches risk injury to the mitral valve apparatus. Given the natural history of tumor regression, partial surgical extirpation has generally been advocated as a means to relieve the hemodynamic ball-valve occlusion. Although preservation of the aortic valve has been generally achieved, the Ross procedure has been reported given the high risk for irreversible damage to the semilunar valve after tumor resection [3]. Furthermore, surgical resection requires cardiopulmonary bypass and often some degree of aortic cross-clamp.

There are limited reports documenting single ventricle palliation for cases of left ventricular tumor. Initially our patient had significant LVOT obstruction combined with a hypoplastic LV with poor function. The hybrid procedure was felt to be advantageous over tumor resection as there was concern that the LV would not be able to provide adequate cardiac output. The hybrid procedure also afforded the benefit of allowing for natural tumor regression and sparing any operative complications of aortic valve injury or conduction system damage.

The etiology of the patient’s transient right ventricular dysfunction post hybrid procedure was never clearly identified. He had neither ischemic changes on electrocardiogram nor obstruction to coronary artery blood flow visualized by echo. Although there was no evidence of tumor encroachment into the right ventricle, the tumor burden within the septum may have resulted in further dyssynchrony or an unfavorable ventricular-ventricular interaction causing right ventricular systolic dysfunction. Similar cases of development of right ventricular dysfunction after a hybrid procedure in this clinical setting have not been documented.

Both tumor resection and the hybrid procedure have advantages and disadvantages. Tumor resection can be successfully completed in 1 operation, but for masses with extensive valvular tissue involvement, technical success can be quite difficult. The hybrid procedure does not require cardiopulmonary bypass, which theoretically diminishes the risk of neurologic insult in this at-risk population. The hybrid procedure also offers the potential for spontaneous tumor regression to support eventual biventricular circulation. As seen in our patient, sustained tumor burden may have deleterious effects on biventricular function, which may preclude further single ventricular palliative operations. However, the right ventricular function improved over time with medical therapy, resulting in a successful superior cavopulmonary anastomosis palliation in this patient with a large, obstructive cardiac rhabdomyoma.
References