

Treatment of Pediatric Hypertrophic Cardiomyopathy without Significant LVOTO on Echo

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Hypertrophic cardiomyopathy (HCM) is defined as left ventricular hypertrophy in the absence of abnormal loading condition (valve disease, congenital heart disease, hypertension)¹. The diagnosis of HCM is made by clinical symptoms, electrocardiogram, and echocardiogram. Echocardiography is the gold standard for the diagnosis, with the presence of LV wall thickness > 2 standard deviations above the body surface area corrected mean. LV outflow tract obstruction is present at rest in 25-40% of children with HCM^{2,3}.

Beta blockers are the initial drug chosen for medical therapy of symptomatic LV outflow tract obstruction. The oral β -blocker agents most commonly employed in children include propranolol, metoprolol, bisoprolol or atenolol, in a propranolol equivalent dosage of 2-5mg/kg/day. The addition of disopyramide (6-20mg/kg/day) may decrease LV outflow obstruction, and result in symptomatic improvement⁴. The calcium channel blocker verapamil (at a dosage of 3-7 mg/kg/day, administered 6-8 hourly) results in improved diastolic relaxation and can also improve symptoms in obstructive and non-obstructive HCM in children⁵.

In children with obstructive symptoms refractory to pharmacological therapy, surgical septal myotomy-myectomy results in significant symptomatic improvement⁶. Another surgical option is direct injection of alcohol into septal perforators which can result in relief of LV obstruction and symptomatic improvement in adults⁷, but there is no role for alcohol septal ablation in children outside the research setting. Also, radiofrequency ablation of the ventricular septum to relieve LV outflow obstruction has been reported in children, but there was significant morbidity and mortality⁸. The use of dual chamber pacemaker therapy has limitations, too. At present, pacemaker treatment of LV outflow tract obstruction in children with HCM is limited to individual in whom surgical myectomy is contraindicated or not possible, for example due to significant comorbidities, or in those in whom implantation of an implantable cardioverter defibrillator is being considered.

In symptomatic children without LV outflow tract obstruction, chest pain and dyspnea are often caused by impaired LV diastolic function or myocardial ischemia. β -blockers and calcium channel antagonists such as verapamil and diltiazem can be used to improve LV filling, reduce contractility and reduce myocardial ischemia. For patients with heart failure symptoms should be treated with conventional heart failure medication.