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Clinical manifestation and outcomes of children with hypertrophic cardiomyopathy in Kosovo

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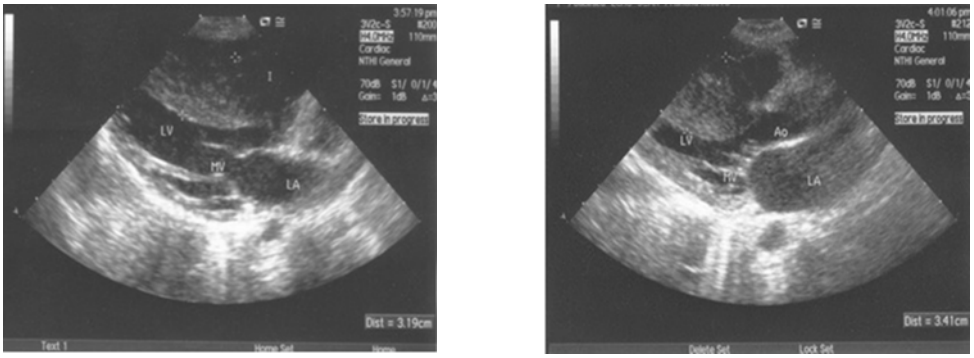
Introduction Identification of the manifestations, assessment and fellow up of children with hypertrophic cardiomyopathy (HCM) by transthoracic echocardiography may be important for clinical management and our understanding of the pathogenesis.

Aim of presentation is to present all children diagnosed with HCM in Kosovo, during the period 2005 -2017, diagnosed by transthoracic echocardiography, treatment and outcomes.

Result Here we present a comprehensive analysis of 43 patients seen in Kosovo, of whom 23 were male, aged between 4 months and 9 years at first presentation, (median of 2 years and 3 months). Cardiac failure, seen in almost half of them, was the most frequent presenting feature. In admission, on the chest x-ray, the cardiothoracic ratio was increased, to a mean of 72 % in 5 infants and to 65 % in 37 older children. Measured by transthoracic echocardiography, in 28 patients hypertrophy of left ventricle was asymmetric while 15 had concentric hypertrophy. Left ventricular ejection fraction was depressed in the 21 patients. Patients in cardiac failure received various combinations of diuretics, B-blockers, ACE inhibitors and aspirin. Death occurred in 8 patients, in 4 of them shortly after admission, 3 left Kosovo and continued examination abroad, and the remaining 32 were followed- up for a mean 42 months, with a range from 5 to 115 months. Surgical intervention was not performed to none of them, despite of clinical and echocardiographic indications. Recovery was noted in 14 patients but still requiring anti-failure medications. Slightly over two-fifths died. Of those with asymmetric form, 45 % died, in half of those presenting in infancy, and 89 % of those who presented at admission with signs of cardiac failure.

Key words: hypertrophic cardiomyopathy, left ventricular hypertrophy, heart failure, myectomy transthoracic echocardiography



A B

Figure 1. Parasternal left ventricular long-axis echocardiografic section obtained a patient in diastole [A] and systole [B] with hypertrophic cardiomyopathy. LV- left ventricle, LA- left atrium, Ao- aorta, MV- mitral valve