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Etiology of Hypertrophic Cardiomyopathy in Young Childhood

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Introduction:
Hypertrophic cardiomyopathy (HCM) in young childhood is a rare and heterogeneous disease. The prognosis is variable and depends upon the underlying disorder. Unfortunately the etiology of HCM remains unclear in the majority of reported cases. This study aims to describe the diversity of etiologies and the prognosis of HCM in infants under the age of one year.

Methods:
In this retrospective single center study all patients under the age of one year born from 2007-2012 with cardiac hypertrophy measured by echocardiography (defined as interventricular septum thickness with Z-score >1.64) were included. Patients with underlying congenital heart disease or hypertension were excluded. Diagnosis and clinical follow up were extracted from patient files.

Results:
During the studied period 32 children with HCM matching the inclusion criteria were identified. Malformation syndromes (including Beckwith-Wiedemann syndrome, Costello syndrome, Noonan syndrome, Cantu syndrome, Leprechaunism, Bernadelli-Seip congenital lipodystrophy and Trisomy 18) were most frequently diagnosed (31%). Other underlying causes were maternal diabetes mellitus (19%), metabolic disease (6%), sacromeric disease (3%), toxic disease (3%) and congenital hyperinsulinemia (3%). In 34% of the patients no underlying cause of the cardiac hypertrophy was identified. Overall, the underlying etiology of HCM was related to hyperinsulinemia in more than half of the HCM patients. In this studied group 25% of the patients died, all during their first year of life (mean age of death: 4 months). During follow-up HCM normalized in 80% of the surviving patients. Patients diagnosed with malformation syndromes had the highest chance of dying and persistent HCM.

Conclusion:
An underlying etiology can be found in 2/3 of infants with HCM under the age of 1 year. Hyperinsulinemia is a likely causative factor in more than half of these patients and may have direct consequences for prognosis and treatment. Infants with HCM surviving their first year of life have a good prognosis as the HCM resolves in most of the cases.