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The Etiology of Cardiac Hypertrophy in Infants

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Introduction
Cardiac hypertrophy in young childhood is a rare and heterogeneous disease. Unfortunately, often the etiology remains idiopathic. The pathophysiology and distribution of etiologies in infants are supposedly different from cardiac hypertrophy developing later in childhood.

Objectives
To investigate the distribution of etiologies, association with hyperinsulinism, distinctive (echocardiographic) variables and the prognosis of cardiac hypertrophy in infants.

Methods
This single center retrospective study included all patients born between 2005-2014 with cardiac hypertrophy measured by echocardiography (diastolic interventricular septum (IVSd) or left ventricular posterior wall (LVPWd) thickness with Z-score ≥2.0) below the age of 1 year. Children with cardiac hypertrophy due to congenital heart disease (CHD) or hypertension were excluded. Underlying diagnosis, echocardiographic data and clinical follow-up were extracted from patient files. Association with hyperinsulinism was reviewed for each underlying cause. □□

Results
Echocardiograms of 6941 infants were screened. Cardiac hypertrophy was reported in 205 cases. After exclusion of 141 children with Z-scores <2.0 (n=72), underlying CHD (n=56) or hypertension (n=13) 64 infants remained eligible for analysis. In two-thirds of these children (n=44;69%) an etiology was identified. Malformation syndromes (n=21;33%, including Noonan n=10;16%) and maternal diabetes mellitus (n=13;20%) were most common causes. Less common causes were sarcomeric disease (n=4;6%), metabolic disease (n=3;5%), congenital hyperinsulinism (n=2;3%) and neuromuscular disease (n=1;2%). In half of the identified causes the etiology was associated with hyperinsulinism (n=21;33%). Cardiac hypertrophy by hyperinsulinism was diagnosed significantly earlier, had lower LVPWd (Z-scores), a higher IVSd:LVPWd ratio and a more often and faster normalization of cardiac hypertrophy compared with cases without hyperinsulinism (all $P<0.05$). Metabolic disease, sarcomeric disease and malformation syndromes had higher mortality rates and worse survival than children with maternal diabetes mellitus, idiopathic cardiac hypertrophy, congenital hyperinsulinism or a neuromuscular disorder.

Conclusions
An etiology can be identified in most infants with cardiac hypertrophy. Hyperinsulinism is a causative factor in the development of cardiac hypertrophy in many infants. Echocardiographic variables may distinguish between the different causes of cardiac hypertrophy. Prognosis depends on the underlying cause.