

TROPONINI AND ITS RELATION TO DISEASE SEVERITY IN PAEDIATRIC HYPERTROPHIC CARDIOMYOPATHY

Sarah Watson

University College London, London, UK

Abstract

Troponin is associated with increased risk of adverse outcomes and correlates with multiple parameters of disease severity in adults with hypertrophic cardiomyopathy (HCM). However, prognostic and staging markers in adults are not always of value in children with HCM. This study assessed the ability of troponin I (TnI) to predict clinical variables in a paediatric cohort of HCM and compare this to well-established biomarker, NT-proBNP. TnI and NT-proBNP were measured in forty-nine patients with HCM [10.69±5.34 years old, 32 (65.31%) male] and elevated TnI is defined as ≥34ng/L (99th percentile reference limit). Evaluation included ECG, echocardiography, ambulatory ECG [19 (38.78%)], ICD interrogation [9 (18.37%)], exercise testing [19 (38.78%)], and cardiac magnetic resonance (CMR) imaging [16 (32.65%)]. TnI was detected in 19 (38.78%) and ≥34 ng/L in 14 (28.57%). There were significant differences in maximum wall thickness (MWT) z-score, E/E', mitral E-wave deceleration time, and CMR-assessed LV mass index between patients with TnI<34ng/L and TnI≥34ng/L. Continuous TnI, but not NT-proBNP, correlated with global longitudinal strain (rs=0.62, p<0.001), and there were significant differences in TnI levels in patients with ST-segment changes, and late gadolinium enhancement. Both biomarkers correlated with MWT z-score and E/E', although correlations were stronger for NT-proBNP. Multivariate analysis revealed TnI was an independent predictor of MWT and LV mass index. Troponin is a reliable biomarker to identify features of HCM (extreme hypertrophy and diastolic dysfunction) and may be an additive monitoring parameter in children. However, the utility beyond NT-proBNP, and the ability to identify subclinical ischaemia and fibrosis is uncertain.

Introduction

Cardiomyopathy is a disease of the heart muscle that makes it harder for your heart to pump blood to the rest of your body. Cardiomyopathy can lead to heart failure.

The main types of cardiomyopathy include dilated, hypertrophic and restrictive cardiomyopathy. Treatment — which might include medications, surgically implanted devices or, in severe cases, a heart transplant — depends on which type of cardiomyopathy you have and how serious it is.

Symptoms

There might be no signs or symptoms in the early stages of cardiomyopathy. But as the condition advances, signs and symptoms usually appear, including:

- Breathlessness with exertion or even at rest
- Swelling of the legs, ankles and feet
- Bloating of the abdomen due to fluid buildup
- Cough while lying down
- Fatigue
- Heartbeats that feels rapid, pounding or fluttering
- Chest discomfort or pressure
- Dizziness, lightheadedness and fainting

Signs and symptoms tend to get worse unless treated. In some people, the condition worsens quickly; in others, it might not worsen for a long time.

Types of cardiomyopathy include:

Dilated cardiomyopathy. In this type of cardiomyopathy, the pumping ability of your heart's main pumping chamber — the left ventricle — becomes enlarged (dilated) and can't effectively pump blood out of the heart.

Hypertrophic cardiomyopathy. This type involves abnormal thickening of your heart muscle, particularly affecting the muscle of your heart's main pumping chamber (left ventricle). The thickened heart muscle can make it harder for the heart to work properly.

Hypertrophic cardiomyopathy can develop at any age, but the condition tends to be more severe if it becomes apparent during childhood. Most affected people have a family history of the disease, and some genetic mutations have been linked to hypertrophic cardiomyopathy.

Arrhythmogenic right ventricular dysplasia. In this rare type of cardiomyopathy, the muscle in the lower right heart chamber (right ventricle) is replaced by scar tissue, which can lead to heart rhythm problems. It's often caused by genetic mutations.

Unclassified cardiomyopathy. Other types of cardiomyopathy fall into this category.