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Perspective

Emerging Techniques and Clinical Implications of Myocardial Fibrosis in Children with Hypertrophic Cardiomyopathy (HCM)

Harald Karlsen*

Department of Medical Biochemistry, University of Oslo, Oslo, Norway

DESCRIPTION

Hypertrophic Cardiomyopathy (HCM) is a complex and heterogeneous genetic cardiovascular disorder characterized by hypertrophy of the myocardium, often accompanied by abnormal diastolic function and myocardial fibrosis. While HCM affects individuals of all ages, including children, the assessment of myocardial fibrosis in pediatric patients presents unique challenges due to differences in disease progression, limited diagnostic techniques, and concerns about ionizing radiation exposure.

HCM in children can lead to various clinical manifestations, including chest pain, syncope, heart failure, arrhythmias, and sudden cardiac death. While the majority of pediatric patients with HCM do not experience severe symptoms, accurate risk stratification and early intervention are significant to prevent adverse outcomes. Myocardial fibrosis, characterized by the excessive deposition of collagen and other extracellular matrix components, is a central pathological feature of HCM and contributes to ventricular dysfunction and arrhythmogenesis. Accurate assessment of myocardial fibrosis is essential for risk stratification, treatment selection, and disease monitoring in children with HCM.

Cardiac imaging modalities play a pivotal role in visualizing and quantifying myocardial fibrosis. Several advanced imaging techniques have emerged as valuable tools for assessing myocardial fibrosis in pediatric HCM patients such as Cardiac Magnetic Resonance Imaging (CMR), Echocardiography, Computed Tomography (CT). Cardiac Magnetic Resonance Imaging (CMR) is considered the gold standard for non-invasive assessment of myocardial fibrosis.

Late Gadolinium Enhancement (LGE) imaging, a CMR technique, visualizes areas of fibrosis as bright regions following gadolinium contrast administration. LGE-CMR provides detailed information about fibrosis location, extent, and distribution. In pediatric HCM patients, LGE-CMR offers insights into disease severity, risk stratification, and treatment response. Echocardiography is widely used for the initial evaluation and serial monitoring of pediatric HCM patients. While it may not directly visualize myocardial fibrosis, echocardiography-derived parameters, such as strain imaging and tissue Doppler imaging, provide information about myocardial function and deformation.

Combining strain imaging with other clinical markers allows for indirect assessment of fibrosis-related alterations in myocardial mechanics. Cardiac Computed Tomography (CT) can offer valuable insights into myocardial fibrosis by visualizing structural abnormalities and evaluating myocardial perfusion. Emerging CT techniques, such as CT-derived myocardial native T1 mapping, has the potential to provide non-invasive fibrosis assessment in pediatric HCM patients. In addition to cardiac imaging modalities, biomarkers have gained attention as potential tools for assessing myocardial fibrosis in pediatric HCM patients.

Biomarkers are measurable indicators that reflect underlying pathological processes. Several biomarkers has the potential for evaluating myocardial fibrosis in children with HCM such as Serum Biomarkers, Natriuretic Peptides, MicroRNAs (miRNAs). Biomarkers such as serum collagen peptides (procollagen type I and III), soluble ST2, and galectin-3 have been investigated for their association with myocardial fibrosis. These biomarkers may provide valuable insights into fibrotic processes and disease progression. However, their utility in pediatric populations requires further validation. Natriuretic peptides, including B-type natriuretic peptide (BNP) and N-terminal pro-B-type natriuretic peptide (NT-proBNP), are established biomarkers for heart failure.

Elevated natriuretic peptide levels in pediatric HCM patients may indicate myocardial dysfunction and fibrosis-related ventricular remodeling. miRNAs are small non-coding RNAs that regulate gene expression. Altered miRNA profiles have been associated with myocardial fibrosis in various cardiovascular

Correspondence to: Harald Karlsen, Department of Medical Biochemistry, University of Oslo, Oslo, Norway, E-mail: hardlsn@gmail.com

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diseases. Exploring miRNA signatures in pediatric HCM patients could potentially provide insights into fibrotic processes and disease severity. The accurate assessment of myocardial fibrosis in children with hypertrophic cardiomyopathy holds significant clinical implications.

Identification of fibrosis patterns and disease severity informs risk stratification, guides therapeutic decisions, and facilitates monitoring of treatment efficacy. Integrating information from various cardiac imaging modalities and biomarkers can enhance diagnostic accuracy and provide a comprehensive understanding of myocardial fibrosis in pediatric HCM patients. Standardized protocols for imaging acquisition, analysis, and interpretation are essential to ensure consistent and reproducible results across different centers. The application of machine learning and artificial intelligence algorithms to multi-modal imaging and biomarker data may enhance the accuracy of fibrosis assessment and risk prediction.

CONCLUSION

Accurate assessment of myocardial fibrosis is a critical component of managing pediatric hypertrophic cardiomyopathy. Cardiac imaging modalities, such as cardiac magnetic resonance imaging and echocardiography, provide valuable insights into fibrosis location and extent. Biomarkers offer a complementary approach to evaluate fibrosis-related processes and disease severity. Integrating data from multiple sources, including imaging and biomarkers, holds the potential to improve risk stratification, guide therapeutic decisions, and enhance the care of children with hypertrophic cardiomyopathy. As technology advances and our understanding of fibrosis processes deepens, the field has the potential to make substantial advancements in both the diagnosis and treatment of myocardial fibrosis in paediatric HCM patients.