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Editorial

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Pediatric Acute Myocarditis: Current Approach to Diagnosis and Treatment

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▸ See the article "Acute Myocarditis in Children: a 10-year Nationwide Study (2007–2016) based on the Health Insurance Review and Assessment Service Database in Korea" in volume 50 on page 1013.

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Myocarditis is a common disease in the pediatric population with variable clinical presentations. It is usually categorized into four major types, such as acute, fulminant, chronic active, and chronic persistent.[1] The accurate diagnosis of pediatric acute myocarditis is difficult because the symptoms are frequently non-specific, especially in infants and children. Acute fulminant myocarditis is characterized by a sudden onset of severe congestive heart failure or cardiogenic shock after a flu-like illness. Aggressive treatment is warranted in these patients, including mechanical circulatory support and heart transplantation in patients with persistent myocardial dysfunction.

The actual incidence rate of pediatric myocarditis is difficult to ascertain because of its frequent subclinical presentations and lack of the diagnostic criteria. The incidence rate is probably underestimated because pediatric myocarditis may be asymptomatic in many patients and sudden death is a common presentation of undiagnosed myocarditis. Neonates have the highest incidence with a second peak occurring around mid-adolescence.[2] The prevalence is higher among boys. The age and sex-wise distributions are reported in a previous study.[3]

Mostly, myocarditis in children is attributable to viruses, such as myocarditis-enterovirus, herpes virus (Ebstein-Barr virus), and influenza A virus. Giant cell myocarditis and cardiac sarcoidosis are rarely detected in children, but these conditions should be kept in mind, especially in adolescent patients, because the treatment and outcomes differ from those of viral myocarditis.[4] While endomyocardial biopsy (EMB) is the gold standard of diagnosis of acute myocarditis, it is limited by high inter-observer variability, sampling error, and the risk of potential adverse events. Due to these limitations, the American Heart Association and the American College of Cardiology have established guidelines to perform EMB only in certain cases.[5] It is required to distinguish the specific histological types, such as giant cell myocarditis, eosinophilic myocarditis, and sarcoidosis, from lymphocytic myocarditis because, in the former conditions, early immunosuppressive therapy is recommended.

Cardiac magnetic resonance imaging (MRI) is currently considered to be the gold standard non-invasive diagnostic test of myocarditis and only secondary to EMB. Cardiac MRI is safe, offers clear understanding of the anatomy, and is consistent and accurate in interpretation. The MRI findings of acute myocarditis are relative edema, relative enhancement, and late

enhancement. The diagnosis of myocarditis by cardiac MRI is made by the Lake-Louise criteria, which were revised.[6] Following the established MRI diagnostic criteria, detection of patchy myocardial involvement is possible because of the excellent correlation between the findings of cardiac MRI and those of EMB.[7] A study has shown high sensitivity (78%) and specificity (91%) for the signs of myocarditis, especially if performed in the first 2 weeks of the disease onset.[8] However, less is currently known regarding the prognostic value of cardiac MRI in pediatric myocarditis.

The treatment of this disease is not standardized and studies in the pediatric population are lacking. Further randomized clinical trials are essential to determine the subset of patients who would benefit from immunoglobulin therapy, immunosuppressive therapy, or both. The use of intravenous immunoglobulin and steroids remains consistently high despite a lack of consensus in the literature on the benefits of immunosuppressive therapy. The ongoing use of both immunoglobulin and steroid, despite limited evidence on the improved outcome, highlights the need for a prospective randomized trial in the pediatric population. With the increasing use of advanced mechanical circulatory support,[9] the mortality rate of critically ill patients is expected to decline.

The prognosis of childhood myocarditis is variable, ranging from full recovery to death or cardiac transplantation. Although the mortality rate is high in patients with more severe illness at presentation and in those who need inotropic as well as mechanical support, those surviving from acute fulminant myocarditis have a better long term prognosis. The mortality is rate is, however, higher in infants (33–45%).

A recent study reported the largest description of the trends in the incidence and outcome of pediatric acute myocarditis in Korea.[3] In this large cohort of children with myocarditis, the authors have highlighted the use of extra-corporeal membrane oxygenation (ECMO), the frequency of use of EMB, and the use of intravenous immunoglobulin and steroids in Korea. In that study, myocarditis was found to cause significant morbidity and mortality, with a large percentage of patients requiring mechanical cardiovascular support. Although the use of ECMO and mechanical ventilatory support and vasoactive drugs were not associated with the outcome, 69.5% of the high-risk patients on ECMO or mechanical ventilation survived, suggesting that these are successful strategies for treating patients.

The wide spectrum of the presentation, lack of clear diagnostic criteria, and practice variations make it difficult to establish a consensus on the diagnosis and management of acute pediatric myocarditis. Nevertheless, viral myocarditis should be the differential diagnosis in all critically ill children. With such a wide variation in the clinical presentation, viral myocarditis is often misdiagnosed leading to a delay in treatment. It is encouraging that several studies on non-invasive imaging as a diagnostic tool for the accurate diagnosis of this condition are ongoing.

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