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Kawasaki Disease: Understanding the Systemic Vasculitis of Childhood

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Abstract: *Kawasaki disease is an acute, self-limited systemic vasculitis of childhood that primarily affects medium-sized arteries, particularly the coronary arteries. It is one of the leading causes of acquired heart disease in children worldwide. The disease is characterized by prolonged fever, mucocutaneous inflammation, lymphadenopathy, and risk of coronary artery aneurysm formation. This narrative review summarizes current evidence on epidemiology, pathogenesis, clinical manifestations, diagnostic approaches, and management strategies, emphasizing early recognition and timely treatment to prevent cardiovascular complications and improve long-term outcomes.*

Keywords: Kawasaki Disease; Vasculitis; Child; Coronary Artery Aneurysm; Inflammation; Immunologic Response

I. INTRODUCTION

Kawasaki disease represents a major pediatric inflammatory condition with significant cardiovascular consequences if untreated. Since its first description, the incidence has increased worldwide, particularly in Asian populations. The disease is characterized by systemic vasculitis affecting small and medium-sized vessels, with a predilection for coronary arteries.

Early recognition is essential because timely treatment with intravenous immunoglobulin significantly reduces the risk of coronary complications. This review aims to provide a comprehensive overview of Kawasaki disease in children, focusing on epidemiology, pathophysiology, clinical spectrum, diagnostic approaches, and management strategies.

Epidemiology

Kawasaki disease occurs predominantly in children under five years of age, with the highest incidence reported in East Asian countries. Boys are affected more frequently than girls. Seasonal variation has been observed, with higher incidence in winter and early spring.

Genetic susceptibility, infectious triggers, and environmental factors are believed to contribute to disease occurrence. Despite extensive research, the exact cause remains unknown.

Pathophysiology

Kawasaki disease is characterized by immune-mediated inflammation of blood vessels. Activation of the immune system leads to endothelial damage, cytokine release, and infiltration of inflammatory cells into the vascular wall.

This inflammatory process results in vasculitis, which may cause dilation and aneurysm formation in coronary arteries. Elevated levels of interleukins, tumor necrosis factor, and other inflammatory mediators play an important role in disease progression.

Persistent inflammation without treatment can lead to structural damage of vessels and long-term cardiovascular complications.

Clinical Spectrum

The disease typically presents with prolonged fever and mucocutaneous signs. The classical clinical features include conjunctival injection, oral mucosal changes, rash, extremity changes, and cervical lymphadenopathy.

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Symptoms often appear in stages and may overlap, making early diagnosis challenging.

Fever

Persistent fever lasting more than five days is the hallmark of Kawasaki disease. The fever is usually high grade and poorly responsive to antipyretics.

Conjunctivitis

Bilateral non-purulent conjunctival congestion is commonly seen. It occurs early in the disease and helps differentiate from bacterial infection.

Oral and Mucosal Changes

Children may develop red cracked lips, strawberry tongue, and diffuse erythema of oral mucosa. These findings reflect mucosal inflammation

Skin Rash

Polymorphous rash is frequently present and may involve trunk and extremities. The rash is non-vesicular and varies in appearance.

Extremity Changes

Swelling and redness of hands and feet occur in the acute phase. In the subacute phase, peeling of skin around fingers and toes is characteristic.

Lymphadenopathy

Cervical lymph node enlargement, usually unilateral, is seen in many patients. Nodes are typically firm and non-suppurative.

Cardiovascular Involvement

Coronary artery inflammation is the most serious complication. Without treatment, aneurysm formation may occur, leading to long-term cardiac morbidity.

Diagnosis

Diagnosis is primarily clinical and based on the presence of fever for at least five days along with characteristic features. Laboratory findings may include elevated ESR, CRP, leukocytosis, thrombocytosis, and anemia.

Echocardiography is essential to evaluate coronary artery involvement and should be performed at diagnosis and during follow-up.

There is no single definitive test, therefore clinical judgment plays a key role.

Management

Early treatment is crucial to prevent coronary artery complications.

Intravenous immunoglobulin (IVIG) is the first-line therapy and should be given within the first 10 days of illness. High-dose aspirin is used during the acute phase to reduce inflammation and fever, followed by low-dose aspirin for antiplatelet effect.

In resistant cases, corticosteroids or biologic agents may be required.

Regular cardiac monitoring is necessary to detect coronary artery abnormalities.

Prevention

There is no specific prevention for Kawasaki disease because the exact cause is unknown. However, early recognition and treatment significantly reduce complications.

Regular follow-up and echocardiography help prevent long-term cardiac damage.

Awareness among clinicians and caregivers is essential for timely diagnosis.

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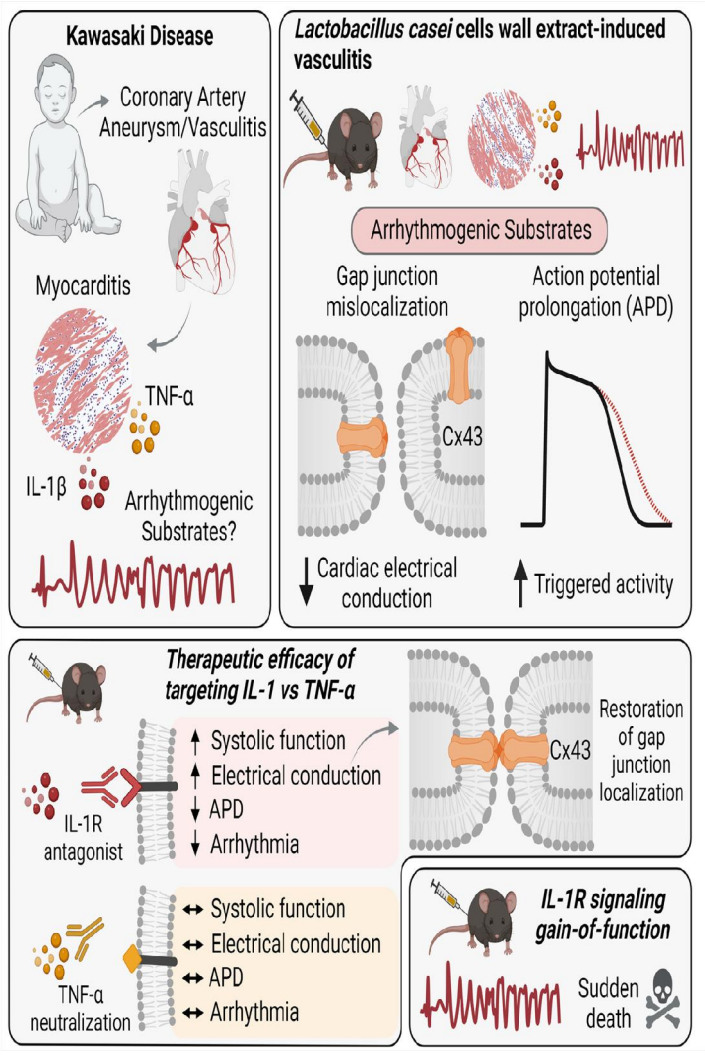
Discussion

Kawasaki disease remains one of the most important causes of acquired heart disease in children. The risk of coronary artery aneurysm highlights the need for early diagnosis and aggressive treatment.

Advances in immunology have improved understanding of the disease, but the exact trigger is still unclear. Ongoing research focuses on genetic factors, immune pathways, and targeted therapies that may improve outcomes in severe cases

One of the most important aspects of Kawasaki disease is the risk of coronary artery aneurysm formation. If treatment is not given within the first 10 days of illness, the risk of cardiovascular complications increases significantly. This makes early recognition extremely important in clinical practice. However, diagnosis may be challenging because the clinical features appear gradually and may mimic viral infections, scarlet fever, measles, or drug reactions. Incomplete or atypical Kawasaki disease is also common, especially in infants, and may lead to delayed treatment.



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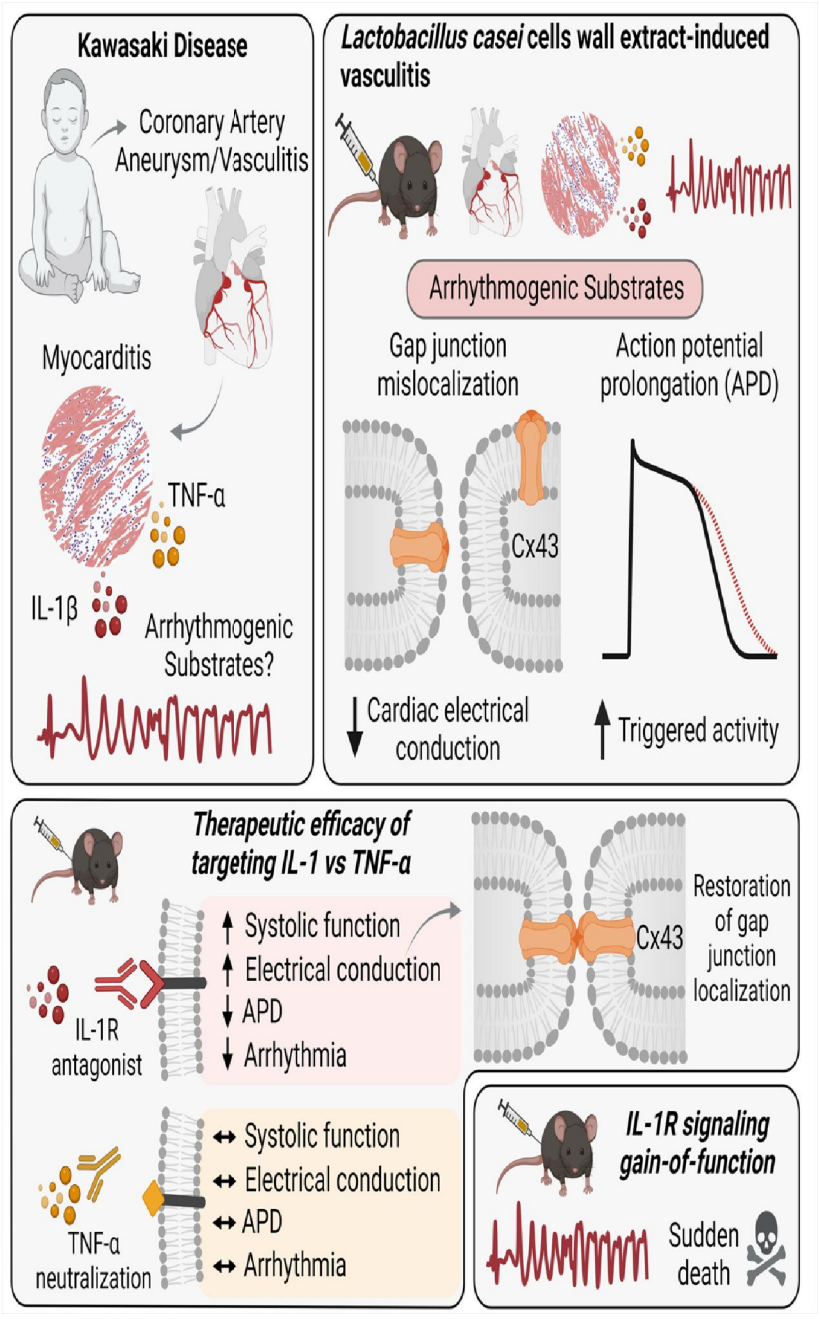
II. CONCLUSION

Kawasaki disease is an acute systemic vasculitis with potential for serious cardiovascular complications. Early recognition and prompt treatment with intravenous immunoglobulin are essential to reduce morbidity and mortality.

A multidisciplinary approach involving pediatricians, cardiologists, and caregivers is necessary for optimal management and long-term follow-up.



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Authors Perspective

In my opinion, Kawasaki disease remains under-recognized in many clinical settings despite its risk of severe cardiac complications. Greater emphasis should be placed on early clinical suspicion, prompt referral, and timely treatment. Improving awareness among healthcare professionals and parents can significantly reduce the burden of coronary artery disease in children. Integration of modern diagnostic tools with careful clinical evaluation will help achieve better long-term outcomes.

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