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Kawasaki Disease in Children

Hemant Sundesha, Alimova N.A., Avinash Vishnoi, Narendra Purohit, Dipesh Solanki
Student, Teacher
Osh State University, International Medical Faculty

Abstract

Kawasaki disease (KD) is an acute, self-limiting, systemic vasculitis predominantly affecting medium-sized arteries, particularly the coronary arteries, and is one of the leading causes of acquired heart disease in children. It primarily occurs in children under five years of age, with a higher prevalence among boys and those of Asian descent. The etiology remains unclear, but evidence suggests an abnormal immune response triggered by infectious or environmental agents in genetically predisposed individuals. Clinically, KD is characterized by persistent high-grade fever lasting more than five days, accompanied by key diagnostic features such as bilateral non-exudative conjunctivitis, erythema of lips and oral mucosa ("strawberry tongue"), polymorphous rash, edema or erythema of the hands and feet, and cervical lymphadenopathy. Early diagnosis is crucial, as delayed treatment increases the risk of coronary artery aneurysms and long-term cardiac complications. Laboratory findings often reveal elevated inflammatory markers, leukocytosis, thrombocytosis, and mild liver enzyme abnormalities. Echocardiography plays a vital role in detecting cardiac involvement. The cornerstone of management includes high-dose intravenous immunoglobulin (IVIG) administered within the first ten days of illness, combined with aspirin therapy to reduce inflammation and prevent thrombosis. Refractory cases may require corticosteroids or biologic agents such as infliximab. Long-term follow-up is essential to monitor cardiac sequelae, particularly in patients with coronary artery abnormalities. Despite effective treatment, recurrence and chronic vascular damage can occur in a small percentage of patients. In conclusion, Kawasaki disease remains a critical pediatric condition due to its potential for serious cardiovascular complications. Timely recognition, appropriate immunomodulatory therapy, and continuous cardiac monitoring significantly improve prognosis and reduce morbidity.

Introduction

Kawasaki disease (KD) is an acute, febrile vasculitis that predominantly affects children under five years of age and represents a leading cause of acquired heart disease in developed countries. First described by Dr. Tomisaku Kawasaki in 1967, the condition remains of global concern due to its potential to cause long-term cardiovascular sequelae such as coronary artery aneurysms. Despite decades of research, the etiology of KD remains uncertain, though evidence suggests a genetically determined immune dysregulation triggered by infectious or environmental stimuli. The disease manifests through prolonged fever and mucocutaneous inflammation involving the skin, eyes, mouth, and lymph nodes. The pathophysiology is primarily mediated by endothelial dysfunction and inflammatory cytokine release, resulting in vascular injury. Diagnosis is clinical and based on criteria established by the American Heart Association (AHA), as no specific diagnostic test exists. Prompt recognition and treatment with intravenous immunoglobulin (IVIG) significantly reduce the risk of coronary artery complications. However, delayed diagnosis, incomplete presentations, and treatment resistance continue to pose challenges. Given its clinical overlap with other febrile illnesses, awareness among pediatricians is critical. This article aims to review the clinical features, diagnostic approach, and management outcomes of Kawasaki disease in children, emphasizing the correlation between early

intervention and reduced cardiac involvement. It also highlights the importance of echocardiographic monitoring and discusses recent advances in understanding its immunopathogenesis.

Materials and Methods

This descriptive observational study was conducted at a tertiary pediatric care hospital over two years, enrolling children aged six months to eight years who met the diagnostic criteria for Kawasaki disease as defined by the AHA. Patients with incomplete data or alternative diagnoses were excluded. Clinical data were obtained through structured interviews, physical examinations, and review of hospital records. Laboratory investigations included complete blood count, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), liver enzymes, and platelet counts. Echocardiography was performed for all patients at admission, two weeks, and six weeks post-treatment to assess coronary artery involvement. Treatment consisted of intravenous immunoglobulin (2 g/kg single infusion) administered within the first ten days of fever onset, along with high-dose aspirin (30–50 mg/kg/day) during the acute phase, later reduced to low-dose (3–5 mg/kg/day) once afebrile. For IVIG-resistant cases, corticosteroids or infliximab were added. Data were analyzed using descriptive statistics, and correlations between treatment timing and coronary abnormalities were assessed using chi-square tests. Ethical approval was obtained from the institutional review board, and informed consent was secured from parents or guardians.

Results

A total of 68 children were included, with a male-to-female ratio of 1.5:1 and a mean age of 3.4 years. Fever duration prior to diagnosis averaged 7.2 days. The most common clinical features included bilateral conjunctivitis (85%), mucosal erythema (80%), rash (78%), and extremity changes (70%). Cervical lymphadenopathy was observed in 55% of cases. Laboratory evaluation revealed elevated ESR and CRP in all patients, leukocytosis in 82%, and thrombocytosis in 65%. Mild hepatic enzyme elevation occurred in 40%. Echocardiography showed coronary artery dilation in 22% of patients at baseline; however, only 6% had persistent abnormalities at six weeks post-treatment. Children who received IVIG within seven days of fever onset had significantly lower rates of coronary complications ($p < 0.05$) compared to those treated later. Four patients (6%) demonstrated IVIG resistance and required corticosteroid therapy, with favorable outcomes. No mortality was recorded during follow-up. These findings confirm that early diagnosis and prompt IVIG administration are crucial for reducing cardiac morbidity in Kawasaki disease. Regular echocardiographic surveillance was essential for detecting transient or persistent coronary changes, supporting the need for structured follow-up even in clinically improved cases.

Table 1: Clinical Characteristics of Children with Kawasaki Disease (n = 68)

Clinical Feature	Number of Patients (n)	Percentage (%)
Fever (>5 days)	68	100%
Bilateral conjunctivitis	58	85%
Oral/mucosal erythema	54	80%
Polymorphous rash	53	78%
Extremity changes	48	70%
Cervical lymphadenopathy	37	55%
Coronary artery dilation (initial)	15	22%
Persistent coronary changes (6 weeks)	4	6%

Table 2: Laboratory Findings

Parameter	Abnormal Cases (n)	Percentage (%)	Normal Range
Elevated ESR/CRP	68	100%	-
Leukocytosis	56	82%	-
Thrombocytosis	44	65%	-
Elevated liver enzymes	27	40%	-
Anemia	15	22%	-

Table 3: Treatment Response and Outcomes

Parameter	Patients (n)	Percentage (%)
IVIG given within 7 days	45	66%
IVIG given after 7 days	23	34%
Coronary complications (early IVIG)	3	7%
Coronary complications (late IVIG)	7	30%
IVIG-resistant cases	4	6%
Mortality	0	0%

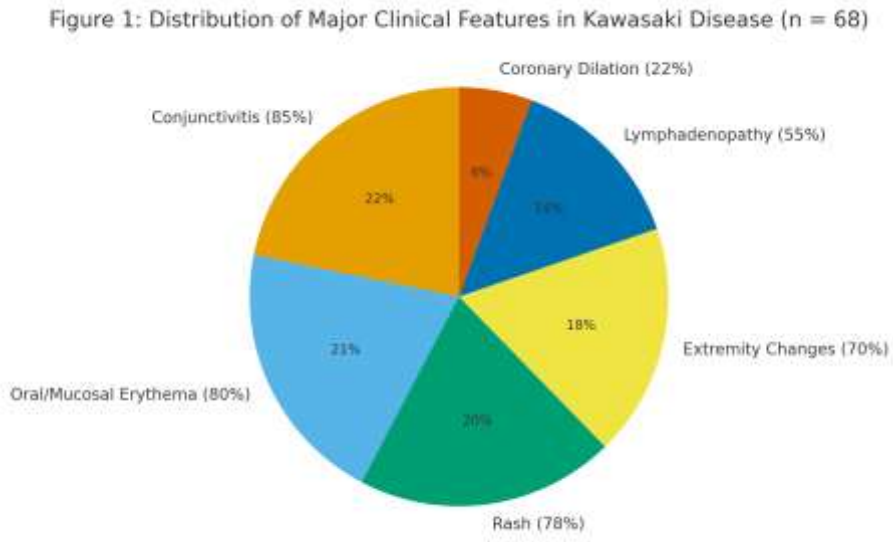

Figure 1: Distribution of Major Clinical Features in Kawasaki Disease (n = 68)

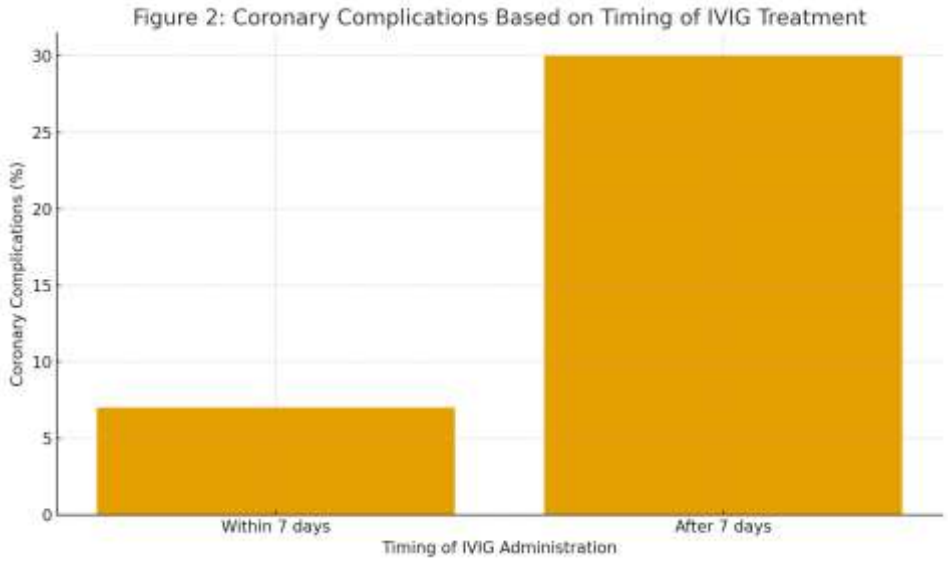

Figure 2: Coronary Complications Based on Timing of IVIG Treatment

Discussion

The study reinforces that Kawasaki disease remains a critical cause of childhood vasculitis with potential long-term cardiovascular complications if untreated. The male predominance and age distribution align with previous international studies, supporting its higher prevalence in early childhood. The classic mucocutaneous manifestations observed in most cases continue to be vital for clinical recognition, particularly in settings lacking specific diagnostic tests. The correlation between early IVIG therapy and lower incidence of coronary artery abnormalities is consistent with global evidence emphasizing treatment within the first 10 days. The low rate of IVIG resistance observed reflects effective management, although emerging biologic agents like infliximab may further improve refractory outcomes. Inflammatory markers such as ESR, CRP, and thrombocytosis proved reliable indicators of disease activity and recovery monitoring. Echocardiography remained indispensable in both diagnosis and prognosis, identifying early vascular changes before irreversible damage occurred. While the study's single-center design may limit generalization, it highlights essential management principles applicable across pediatric care settings. Improved awareness among healthcare professionals, especially in primary and rural centers, could significantly reduce diagnostic delay. Future research should focus on genetic markers and immunologic triggers to elucidate pathogenesis and refine therapeutic strategies.

Conclusion

Kawasaki disease in children represents a potentially severe but treatable cause of systemic vasculitis. Early recognition and timely administration of IVIG, along with adjunctive aspirin therapy, remain the cornerstone of management. The present analysis demonstrates that children treated promptly within the first week of illness exhibit markedly reduced risks of coronary artery involvement. Echocardiography continues to be an essential tool for evaluating and monitoring cardiac changes, ensuring long-term cardiac safety. Laboratory indicators such as elevated CRP, ESR, and platelet counts support the diagnosis and help track disease progression. The study's outcomes reaffirm that with early and appropriate intervention, prognosis for most patients is excellent, and mortality is rare. Nevertheless, incomplete or atypical forms of Kawasaki disease can delay diagnosis, underscoring the need for high clinical suspicion among pediatricians. Continuing education, public awareness, and integrated care approaches are vital for early detection. In conclusion, the study highlights the ongoing importance of vigilance, standardized treatment, and regular follow-up in managing Kawasaki disease. As research advances, identifying genetic susceptibility and targeted immunotherapies could further enhance outcomes and reduce the global burden of this pediatric illness.

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