Kawasaki disease: The Rheumatologist’s Perspective

By Angela Migowa, MBCHB-UON, MMed (Pediatrics and Child Health)-AKUHN
Pediatric Rheumatology Fellow
Montreal Children’s Hospital, McGill University
Mother Theresa of Calcutta

"...Few of us can do big things; but we can do small things in a big way...."
Objective

- To understand the pathophysiology of Kawasaki disease.
- To correlate the pathophysiology to the clinical manifestations and management of Kawasaki disease.
Introduction

Kawasaki disease (KD) is an acute, febrile, self-limiting vasculitis of unknown etiology which leads to the formation of ectasia, dilatation, or aneurysm of the coronary arteries in approximately 25% of untreated children.

Terai M et al. J Pediatr 1997;131:888e93
Introduction

Immune Complex Small Vessel Vasculitis
- Cryoglobulinemic Vasculitis
- IgA Vasculitis (Henoch-Schönlein)
- Hypocomplementemic Urticarial Vasculitis
  (Anti-C1q Vasculitis)

Medium Vessel Vasculitis
- Polyarteritis Nodosa
- Kawasaki Disease

Anti-GBM Disease

ANCA-Associated Small Vessel Vasculitis
- Microscopic Polyangiitis
- Granulomatosis with Polyangiitis
  (Wegener’s)
- Eosinophilic Granulomatosis with Polyangiitis
  (Churg-Strauss)

Large Vessel Vasculitis
- Takayasu Arteritis
- Giant Cell Arteritis
First case..... an unusual illness with rash and fever in a 4-year-old child at the Red Cross Hospital in Tokyo, Japan, in January, 1961.

“....I could make no diagnosis of this unusual sickness for which I could find no reference in the medical literature....”

Epidemiology

Second commonest vasculitis of childhood after Henoch Schönlein purpura.

The incidence in Japan is 138/100000 in children younger than 5 years, whereas in the USA, it is 17.1/100 000, and in the UK 8.1/100 000.

Approximately 85% of children with KD are younger than 5 years of age. Peak age incidence is 18–24 months.

Patients aged less than 3 months, or more than 8 years are encountered less commonly, but are at increased risk for coronary artery aneurysms (CAA).

## Epidemiology

<table>
<thead>
<tr>
<th>Author (Country, Year)</th>
<th>Publication</th>
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<tbody>
<tr>
<td>Elamin A et al (Sudan, 1993)</td>
<td>Case series (2 patients)</td>
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<td>Badoe et al (Ghana, 2007)</td>
<td>Case series (3 patients)</td>
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<td>Rakotovao D.N. et al (Madagascar, 2008)</td>
<td>Case series (5 patients)</td>
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Spectrum Of Musculoskeletal Inpatient Diagnoses At a Pediatric Center In East Africa In 2011

Angela Migowa¹, Ines Colmegna², Evelyne Ng’ang’a³, John Wachira⁴, Thomas Ngwiri⁵, Carol A. Hitchon⁶, Sasha Bernatsky⁷ and Rosie Scuccimarri⁸
¹Pediatrics, Aga Khan University Hospital, Nairobi, Kenya, ²Rheumatology, McGill University Health Centre, Montreal, QC, Canada, ³Pediatrics, University of Nairobi, Nairobi, Kenya, ⁴Pediatrics, Gertrude’s Children Hospital, Nairobi, Kenya, ⁵Pediatrics, Gertrude’s Children’s Hospital, Nairobi, Kenya, ⁶Rheumatology, University of Manitoba, Winnipeg, MB, Canada, ⁷Division of Clinical Epidemiology, McGill University Health Center, Montreal, QC, Canada, ⁸McGill University, Montreal, QC, Canada.

Results: The total number of admissions to Gertrude’s Hospital during 2011 was 8,011. Among those, 35 patients were identified as having an ‘M-code’ diagnosis at discharge. When the records were reviewed, non-MSK conditions accounted for 20% (7 cases) of all ‘M-code’ admissions. Minor surgical procedures made up 14.3% (5 cases). When both of these were excluded, diseases of the MSK system and CT represented 0.28% of the total admissions in 2011. Validated diagnoses were classified as inflammatory arthropathies (39.1% or 9 cases), septic arthritis (30.4% or 7 cases); soft tissue and muscle infections (17.4% or 4 cases) and Kawasaki disease (KD) (13.1 % or 3 cases).
Pathophysiology

- The aetiology of KD remains unknown.
- Various pathogens including retroviruses, Epstein–Barr virus, coronavirus, propionibacterium acnes, staphylococcal and streptococcal superantigens have been implicated as infectious triggers of KD.
- Various genetic predispositions have been implicated.

Table 1  Genome-wide studies in disease

<table>
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<tr>
<th>Gene</th>
<th>Population</th>
<th>Kawa-Locus (KD)</th>
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Downloaded from http://adc.bmj.com on June 22, 2015 - Published by group.bmj.com
Figure 5: Proposed events in the evolution of Kawasaki disease. A: Inflammatoty stimulus sets in motion a cascade of events that in genetically predisposed individuals leads to inflammation, myointimal proliferation, thickening of the media, and angiogenesis at the vessel wall. A. Initially, activated circulating mononuclear cells and platelets interact with endothelial cells that express ICAM-1 and LFA-1 as well as adhesion molecule-1 (VLA-4) or P-selectin, leading to migration of activated endothelial cells that express MCP-1, which further attracts monocyte/macrophages, and vascular endothelial-cell growth factor. B: Later, platelets adhere to the vascular wall elements. Inflammation cells cross the endothelium, accumulate in the intima, and liberate proinflammatory cytokines (IL-1, 6, and 8, tumour necrosis factor a (TNFα), and matrix metalloproteinases (MMPs). Neutrophils release neutrophil elastase, which damages...
Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease

A Statement for Health Professionals From the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young American Heart Association

Endorsed by the American Academy of Pediatrics

Jane W. Newburger, MD, MPH; Masato Takahashi, MD; Michael A. Gerber, MD; Michael H. Gewitz, MD; Lloyd Y. Tani, MD; Jane C. Burns, MD; Stanford T. Shulman, MD; Mann F. Bolger, MD; Patricia FerriReal, MD; Robert S. MD; Walter R. Wilson, MD; Donald A. Palace, DMD; Kathryn A. Taubert, PhD

Background: Kawasaki disease is an acute self-limited vasculitis of childhood that is characterized by fever, nonexudative conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash, lymphadenopathy, and coronary artery aneurysms or ectasia and sudden death. It develops in 15% to 25% of untreated children and adults.

Methods and Results: A multidisciplinary committee of experts was convened to revise the American Heart Association recommendations for diagnosis, treatment, and long-term management of Kawasaki disease. The writing group developed an algorithm to aid clinicians in deciding which children with fever for >5 days and <4 classic criteria undergo echocardiography, receive intravenous gamma globulin (IVIG) treatment, or both for Kawasaki disease.
Complete Kawasaki Disease

### TABLE 1. Clinical and Laboratory Features of Kawasaki Disease

**Epidemiological case definition (classic clinical criteria)**

- Fever persisting at least 5 days
- Presence of at least 4 principal features:
  - Changes in extremities
    - Acute: Erythema of palms, soles; edema of hands, feet
    - Subacute: Periungual peeling of fingers, toes in weeks 2 and 3
  - Polymorphous exanthem
  - Bilateral bulbar conjunctival injection without exudate
  - Changes in lips and oral cavity: Erythema, lips cracking, strawberry tongue, diffuse injection of oral and pharyngeal mucosae
  - Cervical lymphadenopathy (>1.5-cm diameter), usually unilateral
- Exclusion of other diseases with similar findings

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*Newburger et al. Kawasaki Disease Diagnosis, Treatment, Management 2751*
Incomplete Kawasaki Disease

3. American Heart Association (AHA) Criteria 2004

Incomplete KD is more common in young infants than in older children, making accurate diagnosis and timely treatment especially important in these young patients who are at substantial risk of developing coronary abnormalities. The incidence of KD is actually higher than previously reported throughout the world, partly because earlier reports did not take incomplete forms into account. The AHA criteria (2004), which incorporate suggestions for laboratory tests and early echocardiography, are helpful for diagnosing incomplete KD. Consultation with an expert (cardiologist, immunologist, or rheumatologist) should be sought at any time when assistance in making a diagnosis is needed. Patients with fever for more than five days (with 2 or 3 principal clinical features for KD) without other explanation should undergo laboratory testing, and if there is evidence of systemic inflammation, an echocardiogram should be obtained even if the patient does not fully meet the clinical criteria for KD. Infants <6 months old with fever for >7 days without other explanation should undergo laboratory testing, and if evidence of systemic inflammation is found, it does not appear to lower the frequency of CAL formation. During the acute phase of the illness, aspirin is administered at 80 to 100 mg/kg per day (30–50 mg/kg in Japan) in four doses with IVIG. High-dose aspirin and IVIG appear to possess additive anti-inflammatory effects.

Table 2: Supplementary laboratory criteria for incomplete Kawasaki disease.

| Fever of >5 d associated with 2 or 3 clinical criteria, |
| C-reactive protein ≥3.0 mg/dL and/or erythrocyte sedimentation rate ≥40 mm/h with the following criteria |
| (1) albumin < 3.0 g/dL |
| (2) anemia for age |
| (3) elevation of alanine aminotransferase |
| (4) platelets after 7 d ≥ 450,000/mm³ |
| (5) white blood cell count ≥15,000/mm³ |
| (6) urine ≥10 white blood cells/high-power field |

Modified from Newburger et al. If ≥3 supplement criteria are met, intravenous immunoglobulin can be prescribed before performing echocardiography.

CRP = C-reactive protein; ESR = erythrocyte sedimentation rate; IVIG = Intravenous immunoglobulin.
Bilateral Bulbar Conjunctival Injection without Exudate
Changes in Lips and Oral Cavity (Erythema, Strawberry Tongue)
Polymorphous Exanthema
Cervical Lymphadenopathy
Peeling, Erythema, Edema of Hands and Feet
SKIN PEELING
SEEN IN HEALING
KAWASAKI

Courtesy of Dr C. Hlela
Kawasaki Shock Syndrome

ARTICLE

Recognition of a Kawasaki Disease Shock Syndrome

John T. Kan*, Magee, MD, 1, Matthew S. Wilder, MD, O. Jaram Melkara, MD, 3, J. Freré R. Frazer, MD, D.O., Joan Pancheri, RN, BSN, CCRN, Adriana H. Tambourie, MD, 2, Virginia E. Watson, MD, Brooke M. Bell, PharmD, 1, and Jane C. Burns, MD, 2

*Department of Pediatrics, University of California, San Diego, La Jolla, California; 1Department of Pharmacy, Skaggs School of Pharmacy and Pharmacal Sciences, University of California, San Diego, La Jolla, California; 2Department of Pediatrics, University of California, San Diego, La Jolla, California; and 3McGill University, Montreal, Quebec, Canada

The authors have indicated they have no potential conflicts of interest to disclose.

ABSTRACT

OBJECTIVE: We sought to define the characteristics that distinguish Kawasaki disease shock syndrome from hemodynamically normal Kawasaki disease.

METHODS: We collected data prospectively for all patients with Kawasaki disease who were treated at a single institution during a 4-year period. We defined Kawasaki disease shock syndrome on the basis of systolic hypotension for age, a sustained decrease in systolic blood pressure from baseline of >20%, or clinical signs of poor perfusion. We compared curatorial and laboratory features, coronary artery measurements, and responses to therapy and analyzed indices of ventricular diastolic and systolic function during acute and convalescent Kawasaki disease.

RESULTS: Of 187 patients with Kawasaki disease, 13 (7%) met the definition for Kawasaki disease shock syndrome. All received fluid resuscitation, and 7 (54%) required vasopressor infusions. Compared with patients without shock, patients with Kawasaki disease shock syndrome were more often febrile and had larger proportions of bands, higher C-reactive protein concentrations, and lower hemoglobin concentrations and platelet counts. Evidence of coronary artery activity was more common in the Kawasaki disease shock syndrome group. Patients with Kawasaki disease shock syndrome more often had impaired left ventricular systolic function (ejection fraction <55%); of 13 patients, 4 of 13 pa Lien ts [31%] vs 2 of 86 patients [2%]), mitral regurgitation (5 of 13 patterns [39%] vs 2 of 83 patterns [2%]), coronary aneurysmal abnormalities (8 of 13 patterns [62%] vs 20 of 86 patients [23%]), and myocardial function of 13 patients [46%] or 32 of 174 patients [18%]. Impairment of ventricular relaxation and compliance persisted with time.
Other Clinical Features

- Irritability
- Arthritis
- Pneumonitis
- Uveitis
- Gastroenteritis, gastrointestinal ischaemia, hydrops of gallbladder
- Dysuria from sterile urethritis or meatitis
- Erythema and induration at BCG vaccination site
- Macrophage Activating Syndrome
Laboratory Investigations

- Elevated ESR, CRP
- Moderate to high WBC count with left shift
- Anemia
- Thrombocytosis (usually occurs later)
- Mild-moderate elevation in transaminases
- Low albumin
- Low sodium
- Sterile pyuria
## Differential Diagnosis of Kawasaki Disease:
### Diseases and Disorders With Similar Clinical Findings

- Viral infections (e.g., measles, adenovirus, enterovirus, Epstein-Barr virus)
- Scarlet fever
- Staphylococcal scalded skin syndrome
- Toxic shock syndrome
- Bacterial cervical lymphadenitis
- Drug hypersensitivity reactions
- Stevens-Johnson syndrome
- Juvenile rheumatoid arthritis
- Rocky Mountain spotted fever
- Leptospirosis
- Mercury hypersensitivity reaction (acrodynia)
Red eyes + ? Kawasaki syndrome, Recent MMR ...

Fever + ? Vaccine reaction

Travel, pets + ? Measles, leptospirosis
Rash accentuated + ? Viral exanthem
Erythrodema + ? Toxic shock syndrome

in groin with peeling + ? Kawasaki syndrome, scarlet fever
Pruritic rash + ? Scarlet fever, drug reaction, chronic course atopic dermatitis

Lesions coxsackie virus, measles
Maculopapular Discrete intracranial + ? Adenovirus simplevirus
Mucous membrane + ? Stevens-Johnson syndrome, bacterial

with Koplik spots

plaques measles

Extremity changes + ? Epstein-Barr virus, cytomegalovirus

Cervical

Cat scratch, tularaemia

Itchy eyes with tearing + ? Allergic conjunctivitis

Oedema and peeling + ? Kawasaki syndrome, scarlet fever, reactions, enterovirus

Resting

Rash

Arthritis and abdominal pain.

Uveitis + ? Kawasaki syndrome, conjunctivitis, adenovirus.

Respiratory symptoms + ? Adenovirus, measles enterovirus.

Fingers and toes + ? Adenovirus, measles enterovirus.

Toxic shock

Fever + ? Generalised lymphadenopathy, ? Kawasaki syndrome, splenomegaly, arthritis,
Cardiac Monitoring

- All patients with KD should undergo echocardiography at diagnosis at 10-14 days and at 6–8 weeks after the onset of the disease.

- Echocardiography should be performed at least weekly in those with aneurysms detected on initial echocardiography (ECHO) and those with ongoing active inflammation to monitor aneurysm size progression, or the development of thrombus formation.

- A negative echocardiogram does not exclude the diagnosis of KD.

Endocytosis

Pro-inflammatory cytokine production

NK cell activation

Expression of MHC class II and costimulatory molecules

Adaptive immunity

NK-cell Allot

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Expression of inhibitory FcRnB1

Blockade of activating FcRs

Production of proinflammatory cytokines

Expression of FcRγ2

Expulsion of eVilifying FCAs I.

Expression of inhibitory FcRnB1

Inhibitory Fc, FcRn

&cell polarization

ElonolC01

Diffusion of Fc, FcRn

&cell polarization

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Diffusion of Fc, FcRn

&cell polarization

Expulsion of eVilifying FCAs I.
Treatment

The Lancet · Saturday 10 November 1984

HIGH-DOSE INTRAVENOUS GAMMA GLOBULIN FOR KAWASAKI DISEASE

KENSHI FUKUSHO  TETSURO KAMITA
HIROYUKI KANAMU  NOBUYUKI KIYOSAWA
KAZUHIKO SHINOHAMA  TATSUHIKO HAYASHIDERA
JUNICHI TANIGAWA  OSAMU HIRIOSE
YUTAKA MAHARA  TATSUHIKO YOKOYAMA
MASAHARU KAWARAGI  KUNICHI BABA
KIYOSHI BABA  CHUZO MURI

Introduction

Kawasaki disease is self-limiting but a serious consequence is the possibility of coronary artery damage, and clinical research has concentrated on attempts at preventing such lesions. Aspirin therapy is usually advised, this being the recommendation of the Kawasaki Disease Research Committee, sponsored by the Japanese Ministry of Health and Welfare. However, there is little evidence of a declining incidence in post-Kawasaki coronary artery lesions. The cause of Kawasaki disease is unknown, though it is generally agreed that immunological mechanisms play a part. An immunological mechanism has similarly been proposed for idiopathic thrombocytopenic purpura (ITP), of which the cause is also unknown. High-dose intravenous gammaglobulin therapy (IVGG) rapidly raises the platelet count in ITP, and this treatment is now widely used even though it is still not certain why IVGG is effective. One possible mechanism is immunomodulation by the Fc-portion
A SINGLE INTRAVENOUS INFUSION OF GAMMA GLOBULIN AS COMPARED WITH FOUR INFUSIONS IN THE TREATMENT OF ACUTE KAWASAKI SYNDROME

JANE W. NEWBURGER, M.D., MASATO TAKAHASHI, M.D., ALEXA S. BEISER, PH.D., JANE C. BURNS, M.D., JOHN BASTIAN, M.D., KYUNJHA CHUNG, M.D., STEVEND, COLAN, M.D., C, ELISE DUFFY, M.D., DAVID R. FULTON, M.D., MARV P. GLODE, M.D., WILBERT H. MASON, M.D., H. COV MEISSNER, M.D., ANNE H. ROWLEY, M.D., STANFORD T. SHULMAN, M.D., VENU DHAR REDDY, M.D., ROBERT P. SUNDEL, M.D., JAMES W. WIGGINS, M.D., THEODORE COLTON, S.D., MARIAN E. MELISH, M.D., AND FRED S. ROSEN, M.D.

Abstract  Background. Treatment of acute Kawasaki syndrome with a four-day course of intravenous gamma globulin, together with aspirin, has been demonstrated to be both safe and effective in preventing coronary-artery lesions and reducing systemic inflammation. We hypothesized that therapy with a single, very high dose of gamma globulin would be at least as effective as the standard regimen.

Methods. We conducted a multicenter, randomized, controlled trial involving 549 children with acute Kawasaki syndrome. The children were assigned to receive gamma globulin either as a single infusion of 2 g per kilogram of body weight over 10 hours or as daily infusions of 400 mg per kilogram for four consecutive days. Both treatment groups received aspirin (100 mg per kilogram per day) through the fourth day of illness, then 3 to 5 mg per kilogram per day.

Results. The relative prevalence of coronary artery abnormalities, adjusted for age and sex, among patients treated with the four-day regimen, as compared with those treated with the single-infusion regimen, was 1.94 (95 percent confidence limits, 1.01 to 3.71) two weeks after enrollment and 1.84 (95 percent confidence limits, 0.89 and 3.82) seven weeks after enrollment. Children treated with the single-infusion regimen had lower mean temperatures while hospitalized (day 2, P < 0.001; day 3, P < 0.004), as well as a shorter mean duration of fever (P = 0.028). Furthermore, in the single-infusion group the laboratory indexes of acute inflammation moved more rapidly toward normal, including the adjusted serum albumin level (P = 0.004), alpha-antitrypsin level (P = 0.007), and C-reactive protein level (P = 0.017). Lower IgG levels on day 4 were associated with a higher prevalence of coronary lesions (P = 0.005) and with a greater degree of systemic inflammation. The two groups had a similar incidence of congestive heart failure, which occurred in 2.7 percent of the children overall. All the adverse effects were transient.

Conclusions. In children with acute Kawasaki disease, a single large dose of intravenous gamma globulin is more effective than the conventional regimen of four daily smaller doses and is equally safe. (N Engl J Med 1991; 324: 1633-9.)

KAWASAKI syndrome is an acute illness of childhood characterized by fever, rash, conjunctivitis, inflammation of the mucous membranes, swollen erythematous hands and feet, and lymphadenopathy. The histopathological features of vasculitis involving arterioles, capillaries, and venules appear in the earliest phase of the disease. Subsequently, the walls of the coronary arteries and other medium-sized muscular arteries may show evidence of focal segmental destruction, with coronary artery aneurysms or ectasia developing in approximately 15 to 25 percent of affected children. Studies in Japan suggested that the intravenous administration of gamma globulin during the acute phase of Kawasaki syndrome decreased the prevalence of coronary-artery lesions. On the basis of these observations, we conducted a multicenter, randomized trial in the United States. Gamma globulin administered in four consecutive daily doses, together with aspirin, resulted in a marked reduction in the prevalence of coronary-artery abnormalities as compared with aspirin alone. Furthermore, this treatment had a rapid and dramatic antinflammatory effect. Motivated by the potential economic and social benefits of shortening the hospital stay for patients with Kawasaki syndrome, we organized a second multicenter, randomized trial to ascertain whether the administration of intravenous gamma globulin in a single large dose would have similar or better efficacy and safety than the standard regimen of four daily doses.

IVIG dose: 2 g/kg given over 10 hours
IVIG

- If persistence of fever $\geq 36$ h after completion of the first infusion, retreat with IVIG (2g/kg).

Hemolytic anemia is a complication of IVIG (more often seen with IVIG retreatment). Therefore monitor hemoglobin.

- The presumed mechanism of hemolysis is that of direct antibody mediated attack.

- There is a greater risk for hemolysis in patients with non-O blood groups.

R. Berard, B. Whittemore and R. Scuccimarri. Pediatric Rheumatology 2012, 10:10
Steroids
Steroids

Intravenous methylprednisone therapy seems to benefit IVIG-resistant KD patients. If persistence of fever occurs after 2\textsuperscript{nd} IVIG, consider pulse methylprednisolone.

Aspirin

Effect of NSAID’s on Platelet-Endothelial Interactions
Aspirin

- ASA 80-100 mg/kg/day ÷ 4 doses until day 14 or until afebrile; then switch to 3 – 5 mg/kg/day until 6 – 8 weeks.

- If echo at 6 – 8 weeks is normal, then can stop ASA; if not continue.

- Some centers use 3 – 5 mg/kg/day throughout.

- Meta-analysis comparing anti-inflammatory doses of aspirin with high-dose aspirin combined with IVIG found no significant difference in the incidence of CAA between the groups.

Aspirin

In the convalescent phase of the condition, if aneurysms persist, antiplatelet therapy in the form of low-dose aspirin (2–5 mg/kg) should be continued long-term until the aneurysms resolve.

Clopidogrel is an alternative antiplatelet agent that could be considered.

In the presence of giant aneurysms (>8 mm) warfarin is recommended in addition to aspirin.

Other Treatment Options

- Infliximab (monoclonal antibody against tumor necrosis factor)
- Cyclophosphamide
- Cyclosporine A
- Methotrexate
- Abciximab
Prognosis

Consequential myocardial ischemia and/or infarction have been recorded not only shortly after KD, but also during later adult life in affected children.

MORTALITY AMONG CHILDREN WITH KAWASAKI DISEASE IN JAPAN

YoRiXX u NAKAMURA, M.D., HIrosHI YANAGAWA, M.D.7 AND ToMINaKo KAWASAKI, M.D.

Abstract Background and Methods: It is not certain whether patients with Kawasaki disease have a higher death rate than the age-matched healthy population. We therefore undertook a study to investigate this question. Between July 1982 and December 1988, 53 collaborating treatment centers collected data on all patients who had an unequivocal new diagnosis of Kawasaki disease. Patients who had recurrent disease or whose treatment center occurred more than 14 days after the onset of symptoms were excluded. Patients were followed from the time of the first visit to the treatment center until December 31, 1989, or until death, whichever occurred first. The expected number of deaths was calculated from Japanese vital-statistics data and compared with the number observed.

Results. Of 4676 patients who met the eligibility criteria, 4606 (98.5 percent) were followed through either the end of the study or the date of death. Thirteen patients (6 boys and 7 girls) died during the study period. The number of deaths expected was 7.61 (ratio of observed to expected).

Since the first description of Kawasaki disease by Dr. Kawasaki of Kitasato University in 1967, 11 case reports from more than 50 countries have appeared. This febrile disease, the cause of which is unknown, primarily affects infants and toddlers, in whom it causes widespread vasculitis. Cardiac sequelae, notably coronary aneurysms and cardiitis, occur in 10 to 20 percent of patients. During the two-year period from 1985 through 1986, 28 deaths from Kawasaki disease (a case fatality rate of 0.1 percent) were reported in Japan.

Many investigators have conducted long-term follow-up studies that have focused on survival and cardiac sequelae among patients with Kawasaki disease. These studies, however, included only patients with cardiac lesions, and follow-up was conducted only to assess the efficacy of treatment. Although such studies yield valuable information about the outcomes of severely affected patients, the results may be biased regarding the prognosis for the total patient group with Kawasaki disease.

To clarify the overall prognosis, we conducted a large multicenter study in which virtually all patients who received a diagnosis of Kawasaki disease were followed, including those who did not return to the treatment center. Our goal was to determine whether patients with Kawasaki disease were more likely to die than age-matched healthy children.

METHODS

Entrance Criteria

To date, 11 nationwide surveys of Kawasaki disease have been conducted by the Japanese Kawasaki Disease Research Committee. All patients in whom Kawasaki disease was diagnosed during a 21.1-year period from July 1982 through December 1984 were included in the eighth survey. Patients included in the ninth survey were given the diagnosis during a two-year period from January 1985 through December 1986, and those included in the 10th survey were given the diagnosis during a two-year period from January 1987 through December 1988.

In 1990, a collaborative research group was formed to conduct a follow-up study of patients with Kawasaki disease. The research group included members of the Japanese Kawasaki Disease Research Committee and pediatricians from 55 treatment centers (see the Appendix). All patients in whom Kawasaki disease was diagnosed who were seen at the 55 treatment centers were included in the 8th, 9th, and 10th nationwide surveys and served as the patient population for this study. We included only patients who met the following criteria: the diagnosis of Kawasaki disease had to be unequivocal; treatments were given according to diagnostic guidelines developed by the committee; the disease had to be in its initial episode, and the patient had to have presented at the treatment center less than 15 days after the onset of symptoms. Thus, patients were excluded from the study if the disease was only probable, if they had recurrent disease, or if they were not seen after the 14th day of symptoms. These limitations were imposed to avoid bias due to the large numbers of patients with late cardiac sequelae seen at referral hospitals.

Protocol

The patients were followed from the time they first came to the treatment center until December 31, 1989, or the time of death, if it occurred before that date. The status of the patients on January 1, 1990, was confirmed by review of records of patient visits to the treatment center after January 1, 1990, by requests for information from the patients’ parents if there was no visit between January and September 1990, and by checks of resident registration records in municipal offices when no reply was received. Deaths were confirmed by letters from the parents or by residential registration.
Long Term Follow Up

• MI caused by thrombosis occlusion of abnormal coronary artery is principle cause of death.
  
  Usually occurs within first year.

  Small solitary aneurysms require long term aspirin therapy.

  Giant aneurysms or multiple complex aneurysms require long term anti-platelet therapy and anticoagulation.

  Primary surgical management is coronary artery bypass graft.
Miriam Makeba

“Age is...wisdom, if one has lived one's life properly.”
Summary

- Kawasaki disease (KD) is an acute, febrile, self-limiting vasculitis that leads to aneurysms of the coronaries in approximately 25% of untreated children.

- Early recognition and treatment is imperative to decrease risk of coronary aneurysms.

- IVIG and aspirin form the cornerstone of treatment in KD.

- Cardiac monitoring is key in detection and management of coronary aneurysms.
• Thanks to our patients who inspire us to improve and be better healthcare providers.
• Thanks to all my mentors and teachers in my professional journey.
• Thanks to Dr. R. Scuccimarri for her guidance and support in preparation for the scientific talks and lectures.
• Thanks to Dr. C Hlela for providing pictures in this presentation.
Asante! Thank you! Merci!