

The following vignette applies to the next 2 items.

A previously healthy 3-year-old boy comes to the office for evaluation of fever. For the past 5 days, he has had a fever of 39 C (102.2 F) to 40 C (104 F) that minimally subsides with acetaminophen. His eyes have been red without discharge. He also has a rash on his back. The child goes to a daycare center where several children have been sick. His temperature is 39.2 C (102.5 F), pulse is 120/min, and respirations are 20/min. The child is irritable during the examination. His neck is supple with full range of motion. There is no lymphadenopathy. Both hands and feet are slightly erythematous and edematous, and a maculopapular rash is present on the trunk. Nikolsky's sign is negative. The patient's eyes and lips are shown in the [exhibits](#).

Item 1 of 2

Which of the following is the most likely diagnosis?

- ☐ A. Adenovirus infection
- ☐ B. Hand, foot, and mouth syndrome
- ☐ C. Kawasaki disease
- ☐ D. Scarlet fever
- ☐ E. Staphylococcal scalded skin syndrome

Submit

Media Exhibit

1 of 2 Media 2 of 2



Media Exhibit

1 of 2 Media 2 of 2



Item 2 of 2

This patient is at greatest risk for which of the following complications?

- ☐ A. Bacteremia
- ☐ B. Coronary artery aneurysm
- ☐ C. Intussusception
- ☐ D. Keratitis
- ☐ E. Rheumatic fever

Submit

The following vignette applies to the next 2 items.

A previously healthy 3-year-old boy comes to the office for evaluation of fever. For the past 5 days, he has had a fever of 39 C (102.2 F) to 40 C (104 F) that minimally subsides with acetaminophen. His eyes have been red without discharge. He also has a rash on his back. The child goes to a daycare center where several children have been sick. His temperature is 39.2 C (102.5 F), pulse is 120/min, and respirations are 20/min. The child is irritable during the examination. His neck is supple with full range of motion. There is no lymphadenopathy. Both hands and feet are slightly erythematous and edematous, and a maculopapular rash is present on the trunk. Nikolsky's sign is negative. The patient's eyes and lips are shown in the [exhibits](#).

Item 1 of 2

Which of the following is the most likely diagnosis?

- ☐ A. Adenovirus infection [8%]
- ☐ B. Hand, foot, and mouth syndrome [12%]
- ☒ C. **Kawasaki disease** [73%]
- ☐ D. Scarlet fever [5%]
- ☐ E. Staphylococcal scalded skin syndrome [1%]

Proceed to Next Item

Explanation:

User Id: 

This patient has **Kawasaki disease (KD)**, an acute vasculitis of small and medium arteries. The peak age at presentation is 18-24 months. Almost all cases occur in **children age <5 years** and rarely in adults. The persistent release of proinflammatory cytokines causes a prolonged high fever (minimally responsive to antipyretics), **irritability**, and systemic inflammation. The incidence of KD is greatest among children of **East Asian** ethnicity.

Diagnosis is based on clinical presentation. The patient should have **fever for ≥5 consecutive days** as well as 4 of the following 5 findings:

1. Conjunctivitis: bilateral, nonexudative, spares limbus
2. Oral mucosal changes: erythema, fissured lips, "strawberry tongue"
3. Rash

[Proceed to Next Item](#)**Explanation:**

User Id:

This patient has **Kawasaki disease (KD)**, an acute vasculitis of small and medium arteries. The peak age at presentation is 18-24 months. Almost all cases occur in **children age <5 years** and rarely in adults. The persistent release of proinflammatory cytokines causes a prolonged high fever (minimally responsive to antipyretics), **irritability**, and systemic inflammation. The incidence of KD is greatest among children of **East Asian** ethnicity.

Diagnosis is based on clinical presentation. The patient should have **fever for ≥ 5 consecutive days** as well as 4 of the following 5 findings:

1. Conjunctivitis: bilateral, nonexudative, spares limbus
2. Oral mucosal changes: erythema, fissured lips, "strawberry tongue"
3. Rash
4. Extremity changes: erythema, edema, desquamation of the hands and feet, usually the last manifestation
5. Cervical lymphadenopathy: >1.5 cm, usually unilateral, least consistent finding (present in $<25\%$ - 50% of patients)

In patients with atypical presentation, supporting laboratory evidence can include the following:

- Elevated C-reactive protein and erythrocyte sedimentation rate
- Leukocytosis with neutrophilia (as opposed to lymphocytosis in viral infections)
- Reactive thrombocytosis
- Sterile pyuria on urinalysis

(Choice A) In children, adenovirus infection can present with fever and conjunctivitis. However, patients typically have pharyngitis, rather than mucositis of the lips, and lack the extremity changes seen in KD.

(Choice B) Hand, foot, and mouth syndrome is a contagious pediatric infection caused by coxsackieviruses. Children have fever; oral vesicles on the **buccal mucosa** and tongue; and small, tender cutaneous lesions on the **palms** and **soles**. This self-limiting illness usually resolves in 2-3 days without complications, making the diagnosis unlikely.

(Choice D) Scarlet fever shares some features of KD (eg, fever, "strawberry tongue," rash, cervical lymphadenopathy). However, in scarlet fever ocular symptoms are uncommon, exudative pharyngitis is usually present, and the rash typically has a "sandpaper" texture with sparing of the palms and soles.

In patients with atypical presentation, supporting laboratory evidence can include the following:

- Elevated C-reactive protein and erythrocyte sedimentation rate
- Leukocytosis with neutrophilia (as opposed to lymphocytosis in viral infections)
- Reactive thrombocytosis
- Sterile pyuria on urinalysis

(Choice A) In children, adenovirus infection can present with fever and conjunctivitis. However, patients typically have pharyngitis, rather than mucositis of the lips, and lack the extremity changes seen in KD.

(Choice B) Hand, foot, and mouth syndrome is a contagious pediatric infection caused by coxsackieviruses. Children have fever; oral vesicles on the **buccal mucosa** and tongue; and small, tender cutaneous lesions on the **palms** and **soles**. This self-limiting illness usually resolves in 2-3 days without complications, making the diagnosis unlikely.

(Choice D) Scarlet fever shares some features of KD (eg, fever, "strawberry tongue," rash, cervical lymphadenopathy). However, in scarlet fever ocular symptoms are uncommon, exudative pharyngitis is usually present, and the rash typically has a "sandpaper" texture with sparing of the palms and soles.

(Choice E) Staphylococcal scalded skin syndrome presents with fever, irritability, and rash. Newborns are particularly susceptible to this disease. Diffuse erythema typically begins around the mouth and spreads rapidly throughout the body. Flaccid blisters appear 1-2 days later in flexural areas, buttocks, hands, and feet with positive Nikolsky's sign. This patient's age and rash with negative Nikolsky's sign make this diagnosis unlikely.

Educational objective:

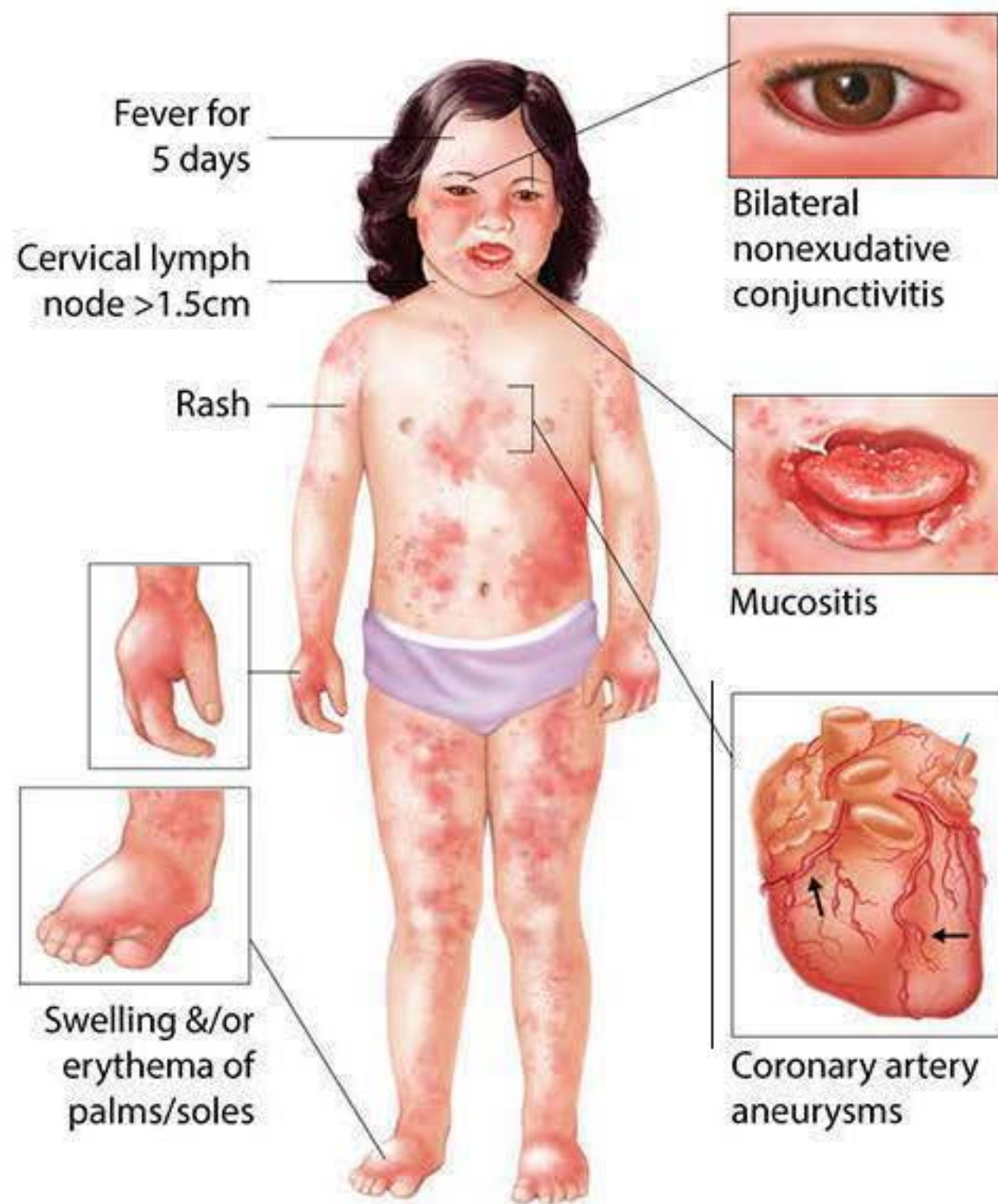
Kawasaki disease is a vasculitis characterized by fever for ≥ 5 days and 4 of the following 5 findings: nonexudative conjunctivitis, extremity changes, cervical lymphadenopathy, oral mucosal changes, and polymorphous rash.

References:

1. **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.**

Kawasaki disease

Kawasaki disease



©UWorld

Media Exhibit

oot and mouth disease



Media Exhibit

oot and mouth disease



Media Exhibit

oot and mouth



Item 2 of 2

This patient is at greatest risk for which of the following complications?

- ☐ A. Bacteremia [2%]
- ☒ B. **Coronary artery aneurysm** [89%]
- ☐ C. Intussusception [3%]
- ☐ D. Keratitis [4%]
- ☐ E. Rheumatic fever [3%]

Proceed to Next Item

Explanation:

User Id: [REDACTED]

Kawasaki disease	
Diagnostic criteria	<p>Fever ≥ 5 days plus ≥ 4 of the following findings:</p> <ul style="list-style-type: none">• Bilateral nonexudative conjunctivitis• Mucositis (injected or fissured lips, injected pharynx, or strawberry tongue)• Cervical lymphadenopathy with at least one lymph node being >1.5 cm in diameter• Erythematous polymorphous rash• Extremity changes (edema & erythema)
Treatment	Aspirin plus intravenous immunoglobulin
Complications	<ul style="list-style-type: none">• Coronary artery aneurysms• Myocardial infarction & ischemia

©UWorld

Although the systemic inflammation in Kawasaki disease (KD) typically self-resolves in about 12 days without intervention, untreated patients are at risk for life-threatening cardiovascular sequelae, especially **coronary artery aneurysms**. The dilated arteries are prone to thrombotic occlusion and consequent myocardial ischemia and death.

Although the systemic inflammation in Kawasaki disease (KD) typically self-resolves in about 12 days without intervention, untreated patients are at risk for life-threatening cardiovascular sequelae, especially **coronary artery aneurysms**. The dilated arteries are prone to thrombotic occlusion and consequent myocardial ischemia and death. **Echocardiography** should be performed at the time of diagnosis and repeated 6-8 weeks later to look for changes that may require closer monitoring and prolonged therapy.

Treatment with **aspirin** and **intravenous immunoglobulin** should be started within 10 days of fever onset to decrease the risk of complications. Both aspirin and intravenous immunoglobulin serve to reduce systemic inflammation, and aspirin also has antithrombotic properties. Aspirin is typically avoided in children due to the risk of Reye syndrome. Reye syndrome is a rare but life-threatening hepatic encephalopathy that can develop in children using aspirin during influenza or varicella infections. However, aspirin is the mainstay of therapy for preventing coronary thrombosis, and caregivers should be warned about this possibility.

(Choice A) Bacteremia is a potential complication of staphylococcal scalded skin syndrome. Patients with KD are not at increased risk of serious bacterial infection.

(Choice C) Intussusception is not associated with KD but can be a complication of such other conditions as Henoch-Schönlein purpura and Meckel's diverticulum.

(Choice D) Keratitis and vision loss are potential complications of adenovirus conjunctivitis. In contrast, the conjunctivitis in KD self-resolves without complications.

(Choice E) Rheumatic fever can occur after an untreated streptococcal infection such as scarlet fever and is not associated with KD.

Educational objective:

Patients with Kawasaki disease are at risk for developing coronary artery aneurysms and thrombosis. Treatment with aspirin and intravenous immunoglobulin is critical in decreasing cardiovascular morbidity and mortality.

References:

1. **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.**