Kawasaki Disease: What you need to know from the 2017 Guidelines

S. Kristen Sexson Tejtel, MD, PhD, MPH
Pediatric Preventive Cardiology
TCHAPP Conference
April 4, 2019
Outline

• History of KD/Epidemiology

• 2017 Guidelines
  - Diagnosis –
    • Classic cases
    • The challenges of diagnosis
    • Echocardiography in KD
    • Need for additional imaging
  - Treatment
    • The future of initial KD therapy
    • IVIG resistance - Knowing when to retreat
    • Complex cases
      • KDSS
      • KD with severe coronary artery aneurysms
      • Persistent features without fever
    • Thromboprophylaxis in KD
  - Follow-up
    • When to follow-up
    • Immunizations after IVIG for KD
What is Kawasaki Disease (KD)?

• Kawasaki disease - *noun*

• a disease of unknown cause, occurring primarily in young children and giving rise to a rash, glandular swelling, and sometimes damage to the heart.
Who “Owns” KD?

- Etiology: Infection? → Infectious Disease?
- Diagnosis/Treatment → Pediatrics/Hospitalists?
- Pathophysiology: Vasculitis → Rheumatology?
- Consequence: Coronary Aneurysms → Cardiology?
Kawasaki Disease (KD) History

AHA SCIENTIFIC STATEMENT

Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease

A Scientific Statement for Health Professionals From the American Heart Association
Epidemiology

• Annual US incidence ~25 per 100,000 children <5yrs old

• 80-90% of cases occur in children <5yrs old
  • Peak 18-24 mo

• Rare beyond late childhood
  • Older children may experience delays in diagnosis

• Boys > Girls (1.5:1)
Epidemiology

- Japan:
  - 10x increased risk with an affected sibling
  - 2x increased risk with a previously affected parent

Key Points: Epidemiology

- In Japan, the recurrence rate is ≈3%, and the relative risk in siblings is 10-fold higher.
- The case fatality rate is <0.1% in Japan.
- Coronary artery aneurysms from KD account for 5% of acute coronary syndromes (ACS) in adults <40 years of age.
Pathophysiology
Table 3. Diagnosis of Classic KD

Classic KD is diagnosed in the presence of fever for at least 5 d (the day of fever onset is taken to be the first day of fever) together with at least 4 of the 5 following principal clinical features. In the presence of $\geq 4$ principal clinical features, particularly when redness and swelling of the hands and feet are present, the diagnosis of KD can be made with $4$ d of fever, although experienced clinicians who have treated many patients with KD may establish the diagnosis with $3$ d of fever in rare cases (Figure 2):

1. Erythema and cracking of lips, strawberry tongue, and/or erythema of oral and pharyngeal mucosa
2. Bilateral bulbar conjunctival injection without exudate
3. Rash: maculopapular, diffuse erythroderma, or erythema multiforme-like
4. Erythema and edema of the hands and feet in acute phase and/or periungual desquamation in subacute phase
5. Cervical lymphadenopathy ($\geq 1.5$ cm diameter), usually unilateral

A careful history may reveal that $\geq 1$ principal clinical features were present during the illness but resolved by the time of presentation.
1. Erythema and cracking of lips, strawberry tongue, and/or erythema of oral and pharyngeal mucosa
2. Bilateral bulbar conjunctival injection without exudate
3. Rash: maculopapular, diffuse erythroderma, or erythema multiforme-like
4. Erythema and edema of the hands and feet in acute phase and/or periungual desquamation in subacute phase.
5. Cervical lymphadenopathy (≥1.5 cm diameter), usually unilateral
IRRITABILITY
KD Characteristics

• Systemic inflammation of medium-sized arteries and multiple organs

• Associated clinical findings:
  - liver (hepatitis),
  - lung (interstitial pneumonitis),
  - gastrointestinal tract (abdominal pain, vomiting, diarrhea, gallbladder hydrops),
  - meninges (aseptic meningitis, irritability),
  - heart (myocarditis, pericarditis, valvulitis),
  - urinary tract (pyuria),
  - pancreas (pancreatitis),
  - lymph nodes (lymphadenopathy).
The Challenges of Diagnosis

- The signs are common
- There is no gold standard lab test
- The stakes are high
- A missed diagnosis may manifest years (or decades) later
- However, are we over-diagnosing?
The Challenges of Diagnosis

- How about incomplete KD?
2017 Guideline

Evaluation of Suspected Incomplete Kawasaki Disease

Children with fever ≥5 days and 2 or 3 compatible clinical criteria OR
Infants with fever for ≥7 days without other explanation

Assess Laboratory Tests

CRP < 3.0 mg/dL and ESR < 40 mm/hr

Serial clinical and laboratory re-evaluation if fevers persist
Echocardiogram if typical peeling develops

Treat

CRP ≥ 3.0 mg/dL and/or ESR ≥ 40 mm/hr

3 or more Laboratory Findings:
1) Anemia for age
2) Platelet count of ≥450,000 after the 7th day of fever
3) Albumin ≤ 3.0 g/dL
4) Elevated ALT level
5) WBC count of ≥15,000/mm³
6) Urine ≥ 10 WBC/hpf

Positive echocardiogram

Treat

Texas Children’s Hospital

Pediatric Cardiology
Echo in KD

Recommendations for Cardiovascular Assessment for Diagnosis and Monitoring During the Acute Illness

1. Echocardiography should be performed when the diagnosis of KD is considered, but unavailability or technical limitations should not delay treatment (Class I; Level of Evidence B).
2017 Guidelines – What Defines a Positive Echocardiogram?

• Any of 3 conditions are met:
  - Z score of LAD or RCA \( \geq 2.5 \)
  - Coronary artery aneurysm
  - \( \geq 3 \) other suggestive features:
    • decreased left ventricular function
    • mitral regurgitation
    • pericardial effusion
    • Z scores in the LAD or RCA of 2 to 2.5
TCH KD Echo Read

Summary:

1. Echobrightness
2. Ectasia
3. Lack of distal tapering
4. Dilation
5. Aneurysm formation
6. Valve regurgitation
7. Aortic root involvement
8. Function
9. Pericardial Effusion
Coronary Artery Involvement

• Z-Score Classification – Risk Level
  1. No involvement: Always <2
  2. Dilation only: 2 to <2.5; or if initially <2, a decrease in Z score during follow-up ≥1
  3. Small aneurysm: ≥2.5 to <5
  4. Medium aneurysm: ≥5 to <10, and absolute dimension <8 mm
  5. Large or giant aneurysm: ≥10, or absolute dimension ≥8 mm
Assessing Aneurysms
Giant Aneurysms and Prognosis

• Patients with giant aneurysms have worst prognosis
• 30 year survival - 90%
• Only 36% cardiac event free

(Tsuda et al AM Heart J 2014; 167: 249-58)
(Manlhoit/PHN Pediatr Cardiol 2010)
Kawasaki Echo Read

• Summary:

1. Echobrightness
2. Ectasia
3. Lack of distal tapering
4. Dilation
5. Aneurysm formation
6. Valve regurgitation
7. Aortic root involvement
8. Function
9. Pericardial Effusion
2017 Guidelines – Advanced Imaging in KD

• It is reasonable to obtain advanced imaging studies
  - computed tomographic angiography (CTA)
  - cardiac magnetic resonance imaging (CMRI)
  - invasive angiography

• Recommended on patients with:
  - severe proximal coronary artery abnormalities
  - when management decisions depend on visualization of distal segments
  - difficult to be seen by echocardiography

• Other vasculature can be imaged simultaneously
**MR in KD**
- Demonstrates distal aneurysms
- Viability/delayed enhancement
- Myocardial perfusion
- See extracardiac dilation
- Frequently need anesthesia

**CT in KD**
- Better than MR for thrombus evaluation
- Smaller slices than MRI
- Down side: Radiation
Catheterization in KD
Goal of Therapy

• Decrease systemic inflammation

• Prevent coronary aneurysms

• Minimize peak dimensions of coronary aneurysm(s)

• Prevent coronary thrombosis
The Future of Initial KD Treatment

• 2017 Guideline states
  - IVIG
    • “Patients with complete and incomplete KD should be treated with high-dose IVIG (2 g/kg)”
  - Aspirin:
    • “Administration of moderate (30 – 50 mg/kg/day) to high dose (80 – 100 mg/kg/day) ASA is reasonable until the patient is afebrile, although there is no evidence that it reduces coronary artery aneurysms”
The Future of Initial KD Treatment

• What about steroids?
  - Traditionally has been a controversial topic
  - But should we consider steroids as part of initial therapy?
The Future of Initial KD Treatment

• 2017 Guideline states
  - “Single-dose pulse methylprednisolone should not be administered with IVIG as routine primary therapy”
  - “A longer course of corticosteroids (tapering over 2 – 3 weeks), together with IVIG and ASA may be considered for treatment of high-risk patients when such high risk can be identified in patients before initiation of treatment”
When to Retreat?

• 2017 Guideline states...
  - “Approximately 10 – 20% of patients with KD develop recrudescent or persistent fever at least 36 hours after the end of their IVIG infusion and are termed IVIG resistant.”
IVIG resistance - Pathophysiology
2017 Guidelines - IVIG Resistance

1. Second dose of IVIG (2 g/kg) after 36 hours
   OR

2. Methylprednisolone 20–30 mg/kg IV x 3 days, +/- taper of oral steroids
   OR
   Second dose of IVIG (2 g/kg), IV steroids x 3 days, +/- taper of oral steroids
   OR

3. Second dose of IVIG (2 g/kg) with prolonged oral taper of prednisolone/prednisone
   OR

4. Infliximab (5 mg/kg) instead of 2nd IVIG or corticosteroids

2017 Guidelines - IVIG Resistance

5. Cyclosporine in those who have failed above

6. Highly refractory patients
   - Anakinra
   - Cyclophosphamide
   - Plasma exchange
Problems with 2017 Guidelines

• No robust data from clinical trials for IVIG resistance

• No guidelines for sickest patients: children < 1 year of age, KD shock syndrome, severe CA abnormalities or macrophage activation syndrome

• Persistent disease (worsening labs, persistent to worsening symptoms) in the absence of fever
Children <1 Year of Age

- These children are at greater risk of treatment failure and CA abnormalities
  - Hispanics with worse outcomes
  - 61% male, 39% female

- Unpublished PHIS data (14,325 pts from 2004-2014):
  - <6 months – 5.9% (852)
  - 6 - 12 months – 11.1% (1,595)

- Delayed diagnosis due to paucity of signs
Kawasaki Disease Shock Syndrome (KDSS)

• “Kawashocki”
  - Hypotension plus KD symptoms
  - Incidence of ≈ 7%

• Compared to typical KD patients
  - Higher CRP, hyponatremia, hypoalbuminemia and thrombocytopenia
  - ↑ IVIG resistance
  - ↑ rates of CA abnormalities
  - Increased risk of MR and prolonged myocardial dysfunction

2017 Guidelines

- Initial IVIG and ASA
- Guidelines hint that KDSS are IVIG resistant
- If IVIG resistant proceed with options 1, 2, 3 or 4, etc

TCH

- Initial IVIG and ASA w/ IV methylprednisolone 30 mg/kg x (3 pulses) w/ oral steroid taper
- 2nd dose of IVIG if IVIG resistant [2]
- Additional therapy driven by clinical course
KD with Severe CA Abnormalities

2017 Guidelines

• No guidelines

• Only case reports with limited outcomes

• Worrisome since these patients are at greatest risk of morbidity and mortality

TCH

Initial IVIG and ASA

+/- 2nd dose of IVIG [1] if IVIG resistant

IV methylprednisolone 30 mg/kg x (3 pulses) w/ oral steroid taper and infliximab vs other anakinra
Persistent Features w/o Fever

2017 Guidelines

• No guidelines outside of initial therapy

TCH

Initial IVIG and ASA

IV methylprednisolone 30 mg/kg x (~3 pulses) w/ oral corticosteroid taper [2]

Additional therapy driven by clinical course
Thromboprophylaxis in KD

• Most common in first 45 days

• Contributing factors:
  - thrombocytosis
  - increased platelet adhesion
  - inflammation
  - endothelial dysfunction
  - abnormal flow conditions
Thromboprophylaxis in KD

- Aspirin – low dose until coronary artery normalization

- Clopidogrel - increased risk of thrombosis or aspirin non-responder

- Lovenox - the anti-inflammatory actions and anticoagulant effects
### Table 8. Risk Classification of Coronary Artery Abnormalities During Follow-up

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>No involvement at any timepoint (Z score always &lt;2)</td>
</tr>
<tr>
<td>2</td>
<td>Dilation only (Z score 2 to &lt;2.5)</td>
</tr>
<tr>
<td>3</td>
<td>Small aneurysm (Z score ≥2.5 to &lt;5)</td>
</tr>
<tr>
<td>3.1</td>
<td>Current or persistent</td>
</tr>
<tr>
<td>3.2</td>
<td>Decreased to dilation only or normal luminal dimension</td>
</tr>
<tr>
<td>4</td>
<td>Medium aneurysm (Z score ≥5 to &lt;10, and absolute dimension &lt;8 mm)</td>
</tr>
<tr>
<td>4.1</td>
<td>Current or persistent</td>
</tr>
<tr>
<td>4.2</td>
<td>Decreased to small aneurysm</td>
</tr>
<tr>
<td>4.3</td>
<td>Decreased to dilation only or normal luminal dimension</td>
</tr>
<tr>
<td>5</td>
<td>Large and giant aneurysm (Z score ≥10, or absolute dimension ≥8 mm)</td>
</tr>
<tr>
<td>5.1</td>
<td>Current or persistent</td>
</tr>
<tr>
<td>5.2</td>
<td>Decreased to medium aneurysm</td>
</tr>
<tr>
<td>5.3</td>
<td>Decreased to small aneurysm</td>
</tr>
<tr>
<td>5.4</td>
<td>Decreased to dilation only or normal luminal dimension</td>
</tr>
</tbody>
</table>

### Table 9. Additional Clinical Features That May Increase the Long-Term Risk of Myocardial Ischemia

- Greater length and distal location of aneurysms that increase the risk of flow stasis
- Greater total number of aneurysms
- Greater number of branches affected
- Presence of luminal irregularities
- Abnormal characterization of the vessel wall (calcification, luminal myofibroblastic proliferation)
- Presence of functional abnormalities (impaired vasodilation, impaired flow reserve)
- Absence or poor quality of collateral vessels
- Previous revascularization performed
- Previous coronary artery thrombosis
- Previous myocardial infarction
- Presence of ventricular dysfunction
2017 Guidelines - Echo Surveillance

- Twice weekly while rapidly expanding
- First 45 days: weekly
- First 3 months: monthly
- Every 3 months for first year
# Kawasaki Disease Follow Up

<table>
<thead>
<tr>
<th>Risk Level</th>
<th>Frequency of Cardiology Assessment*</th>
<th>Assessment for Inducible Myocardial Ischemia</th>
<th>Type and Frequency of Additional Cardiology Assessment</th>
<th>Cardiovascular Risk Factor Assessment and Management</th>
<th>Physical Activity Counseling</th>
<th>Reproductive Counseling</th>
</tr>
</thead>
<tbody>
<tr>
<td>1: No involvement</td>
<td>May discharge between 4 wk and 12 mo</td>
<td>None</td>
<td>None</td>
<td>Assess at 1 y</td>
<td>Promotion counseling at every visit</td>
<td>Age-appropriate counseling without modification</td>
</tr>
<tr>
<td>2: Dilatation only</td>
<td>May discharge after 1 y if normal; assess every 2-6 y if persists</td>
<td>None</td>
<td>None</td>
<td>Assess at 1 y</td>
<td>Promotion counseling at every visit</td>
<td>Age-appropriate counseling without modification</td>
</tr>
<tr>
<td>3.1: Small aneurysm, current or persistent</td>
<td>Assess at 6 mo, then yearly</td>
<td>Assess every 2-3 y</td>
<td>May consider every 3-5 y</td>
<td>Assess at 1 y</td>
<td>Promotion counseling at every visit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>3.2: Small aneurysm, regressed to normal or dilatation only</td>
<td>Assess every 1-3 y (may omit echocardiography)</td>
<td>Assess every 3-5 y</td>
<td>May consider if there is inducible ischemia</td>
<td>Assess at 1 y, then every 2 y</td>
<td>Promotion counseling at every visit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>4.1: Medium aneurysm, current or persistent</td>
<td>Assess at 3.5, and 12 mo, then yearly</td>
<td>Assess every 1-3 y</td>
<td>May consider every 2-5 y</td>
<td>Assess at 1 y</td>
<td>Promotion counseling at every visit; restrict contact</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>4.2: Medium aneurysm, regressed to small aneurysm</td>
<td>Assess yearly</td>
<td>Assess every 2-3 y</td>
<td>May consider every 3-5 y</td>
<td>Assess yearly</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>4.3: Medium aneurysm, regressed to normal or dilatation only</td>
<td>Assess every 1-2 y (may omit echocardiography)</td>
<td>Assess every 2-4 y</td>
<td>May consider if there is inducible ischemia</td>
<td>Assess every 2 y</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>5.1: Large or giant aneurysm, current or persistent</td>
<td>Assess at 2, 6, 9, and 12 mo, then every 3-6 mo</td>
<td>Assess every 6-12 mo</td>
<td>Baseline within 2-6 mo; may consider every 1-5 y</td>
<td>Assess every 6-12 mo</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>5.2: Large or giant aneurysms, regressed to medium aneurysm</td>
<td>Assess every 6-12 mo</td>
<td>Assess yearly</td>
<td>May consider every 2-5 y</td>
<td>Assess yearly</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>5.3: Large or giant aneurysms, regressed to small aneurysm</td>
<td>Assess every 6-12 mo</td>
<td>Assess every 1-2 y</td>
<td>May consider every 2-5 y</td>
<td>Assess yearly</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>5.4: Large or giant aneurysms, regressed to normal or dilatation only</td>
<td>Assess every 1-2 y (may omit echocardiography)</td>
<td>Assess every 2-3 y</td>
<td>May consider every 2-5 y</td>
<td>Assess every 2 y</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
</tbody>
</table>
# Kawasaki Disease Follow Up

## Table 10. Long-Term Assessment and Counseling Algorithm

<table>
<thead>
<tr>
<th>Risk Level</th>
<th>Frequency of Cardiology Assessment</th>
<th>Assessment for Inducible Myocardial Ischemia</th>
<th>Type and Frequency of Additional Cardiology Assessment</th>
<th>Cardiovascular Risk Factor Assessment and Management</th>
<th>Physical Activity Counseling</th>
<th>Reproductive Counseling</th>
</tr>
</thead>
<tbody>
<tr>
<td>1: No involvement</td>
<td>May discharge between 4 wk and 12 mo</td>
<td>None</td>
<td>None</td>
<td>Assess at 1 y</td>
<td>Promotion counseling at every visit</td>
<td>Age-appropriate counseling without modification</td>
</tr>
<tr>
<td>2: Dilation only</td>
<td>May discharge after 1 y if normal; assess every 2–5 y if persists</td>
<td>None</td>
<td>None</td>
<td>Assess at 1 y</td>
<td>Promotion counseling at every visit</td>
<td>Age-appropriate counseling without modification</td>
</tr>
</tbody>
</table>
### Kawasaki Disease Follow Up

#### Table 10. Long-Term Assessment and Counseling Algorithm

<table>
<thead>
<tr>
<th>Risk Level</th>
<th>Frequency of Cardiology Assessment*</th>
<th>Assessment for Inducible Myocardial Ischemia†</th>
<th>Type and Frequency of Additional Cardiology Assessment</th>
<th>Cardiovascular Risk Factor Assessment and Management‡</th>
<th>Physical Activity Counseling§</th>
<th>Reproductive Counseling</th>
</tr>
</thead>
<tbody>
<tr>
<td>1: No involvement</td>
<td>May discharge between 4 wk and 12 mo</td>
<td>None</td>
<td>None</td>
<td>Assess at 1 y</td>
<td>Promotion counseling at every visit</td>
<td>Age-appropriate counseling without modification</td>
</tr>
<tr>
<td>2: Dilation only</td>
<td>May discharge after 1 y if normal; assess every 2–5 y if persists</td>
<td>None</td>
<td>None</td>
<td>Assess at 1 y</td>
<td>Promotion counseling at every visit</td>
<td>Age-appropriate counseling without modification</td>
</tr>
</tbody>
</table>
## Kawasaki Disease Follow Up

### Table 10: Long-Term Assessment and Counseling Algorithm

<table>
<thead>
<tr>
<th>Risk Level</th>
<th>Frequency of Cardiology Assessment*</th>
<th>Assessment for Inducible Myocardial Ischemia†</th>
<th>Type and Frequency of Additional Cardiology Assessment</th>
<th>Cardiovascular Risk Factor Assessment and Management‡</th>
<th>Physical Activity Counseling§</th>
<th>Reproductive Counseling</th>
</tr>
</thead>
<tbody>
<tr>
<td>3.1: Small aneurysm, current or persistent</td>
<td>Assess at 6 mo, then yearly</td>
<td>Assess every 2–3 y</td>
<td>May consider every 3–5 y</td>
<td>Assess at 1 y</td>
<td>Promotion counseling at every visit; restrict contact</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>3.2: Small aneurysm, regressed to normal or dilation only</td>
<td>Assess every 1–3 y (may omit echocardiography)</td>
<td>Assess every 3–5 y</td>
<td>May consider if there is inducible ischemia</td>
<td>Assess at 1 y, then every 2 y</td>
<td>Promotion counseling at every visit</td>
<td>Age-appropriate counseling without modification</td>
</tr>
<tr>
<td>4.1: Medium aneurysm, current or persistent</td>
<td>Assess at 3, 6, and 12 mo, then yearly</td>
<td>Assess every 1–3 y</td>
<td>May consider every 2–5 y</td>
<td>Assess at 1 y</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>4.2: Medium aneurysm, regressed to small aneurysm</td>
<td>Assess yearly</td>
<td>Assess every 2–3 y</td>
<td>May consider every 3–5 y</td>
<td>Assess yearly</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>4.3: Medium aneurysm, regressed to normal or dilation only</td>
<td>Assess every 1–2 y (may omit echocardiography)</td>
<td>Assess every 2–4 y</td>
<td>May consider if there is inducible ischemia</td>
<td>Assess every 2 y</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
</tbody>
</table>
# Kawasaki Disease Follow Up

## Table 10. Long-Term Assessment and Counseling Algorithm

<table>
<thead>
<tr>
<th>Risk Level</th>
<th>Frequency of Cardiology Assessment*</th>
<th>Assessment for Inducible Myocardial Ischemia†</th>
<th>Type and Frequency of Additional Cardiology Assessment</th>
<th>Cardiovascular Risk Factor Assessment and Management‡</th>
<th>Physical Activity Counseling§</th>
<th>Reproductive Counseling</th>
</tr>
</thead>
<tbody>
<tr>
<td>5.1: Large or giant aneurysm, current or persistent</td>
<td>Assess at 3, 6, 9, and 12 mo, then every 3–6 mo</td>
<td>Assess every 6–12 mo</td>
<td>Baseline within 2–6 mo; may consider every 1–5 y</td>
<td>Assess every 6–12 mo</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>5.2: Large or giant aneurysms, regressed to medium aneurysm</td>
<td>Assess every 6–12 mo</td>
<td>Assess yearly</td>
<td>May consider every 2–5 y</td>
<td>Assess yearly</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>5.3: Large or giant aneurysm, regressed to small aneurysm</td>
<td>Assess every 6–12 mo</td>
<td>Assess every 1–2 y</td>
<td>May consider every 2–5 y</td>
<td>Assess yearly</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
<tr>
<td>5.4: Large or giant aneurysm, regressed to normal or dilation only</td>
<td>Assess every 1–2 y (may omit echocardiography)</td>
<td>Assess every 2–3 y</td>
<td>May consider every 2–5 y</td>
<td>Assess every 2 y</td>
<td>Promotion counseling at every visit; restrict contact; self-limit</td>
<td>Precautions for contraception and pregnancy</td>
</tr>
</tbody>
</table>
Kawasaki Disease Follow Up

• At TCH
  - Infrequent follow-up throughout childhood
    • Echocardiogram
    • Electrocardiogram
    • Clinic visit
    • If sports – perfusion imaging (cMRI)
Rheumatology Follow Up

• Children < 1 year or any child with a complicated KD course

• Follow up labs (i.e., CBC, LFTs, inflammatory markers, albumin, sodium) and clinical course

• Tapering of corticosteroids and other therapies if necessary, are based on improvement of labs and collaboration with cardiology
2017 Guidelines – Immunizations After IVIG in KD

• Defer live virus vaccines for 11 months
  - MMR
  - Varicella
  - Flumist
  - Rotavirus
  - Zoster
  - Smallpox
  - Yellow fever