

# Kawasaki Disease in the Prehospital & ED Setting

Adriana Tremoulet, MD, MAS

Associate Director, UCSD Kawasaki Disease Research Center

Professor, Department of Pediatrics

University of California, San Diego





## Vertical admitting of KD patients at RCHSD



Call RCHSD operator and ask for KD doc on call

# Kawasaki Disease (KD)

- Acute self-limited vasculitis of unknown etiology
- Most common cause of acquired pediatric heart disease in developed countries
- Coronary artery aneurysms develop in 25% of untreated children
- Early treatment with IVIG reduces risk of aneurysms



# First 50 Cases (1967)

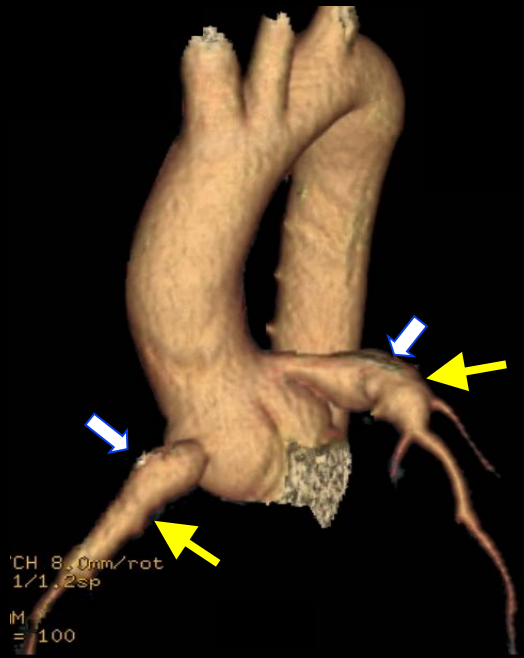


- Persistent fever
  1. Rash
  2. Cervical lymphadenopathy
  3. Conjunctival injection
  4. Red, cracked lips
  5. Extremity swelling
- Convalescent peeling

# The “Red Flags” of KD







# KD in San Diego County

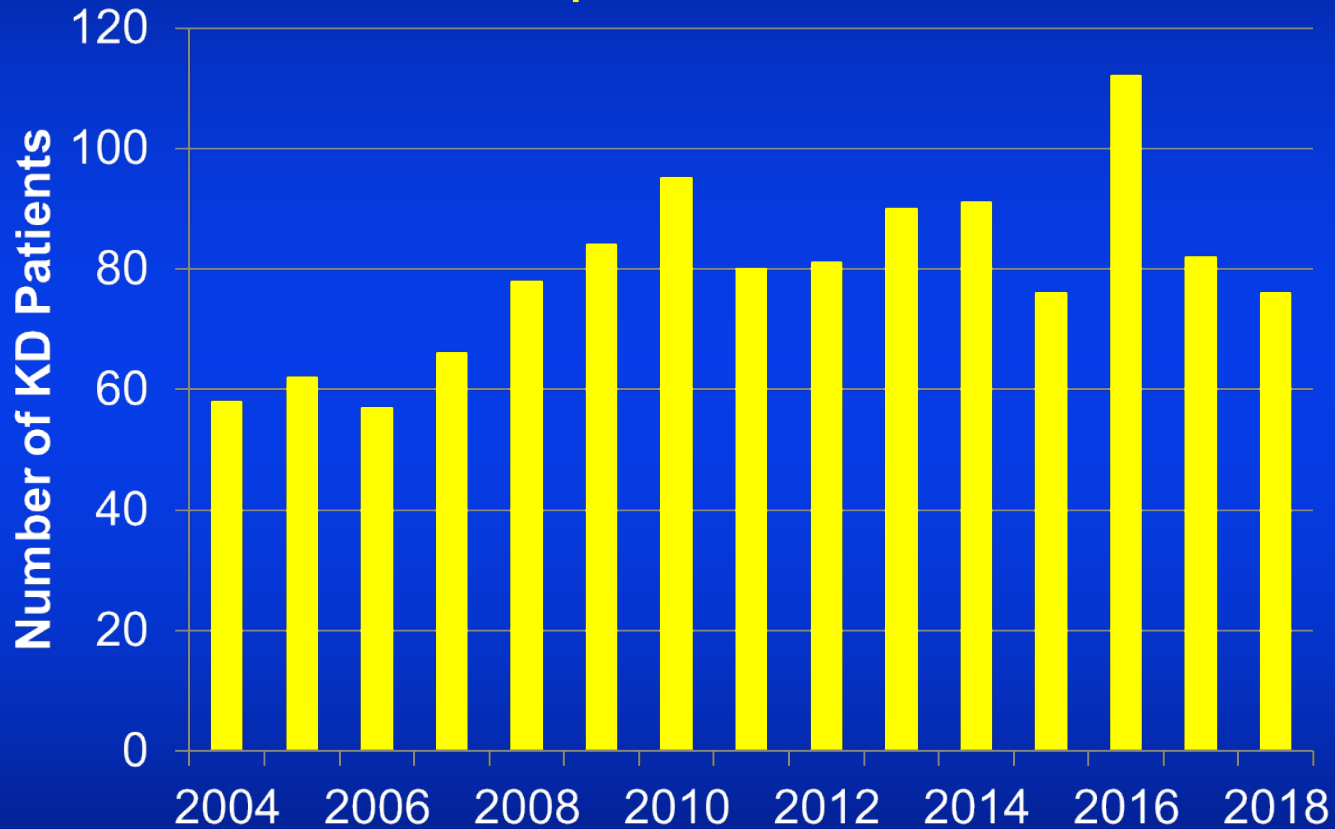


**Attack rate:  
≈48 cases/100,000  
children < 5yrs.**

**100 new cases/yr.**

**One in every 2,000  
children will develop  
KD**

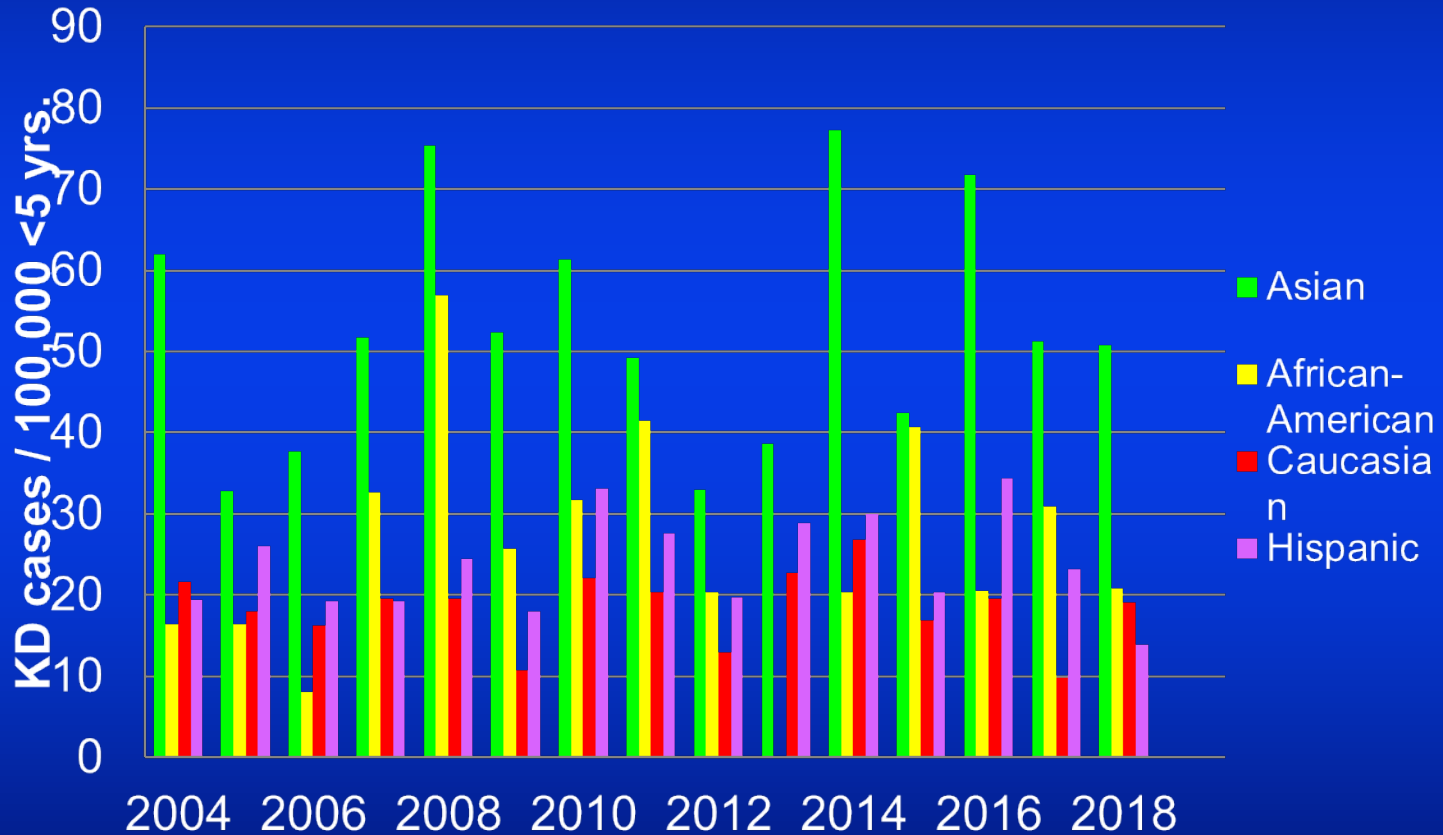
# Kawasaki Disease Cases at Rady Children's Hospital; DOO 2004-2018



• Includes Missed KD cases diagnosed at RCHSD (data from REDCap)



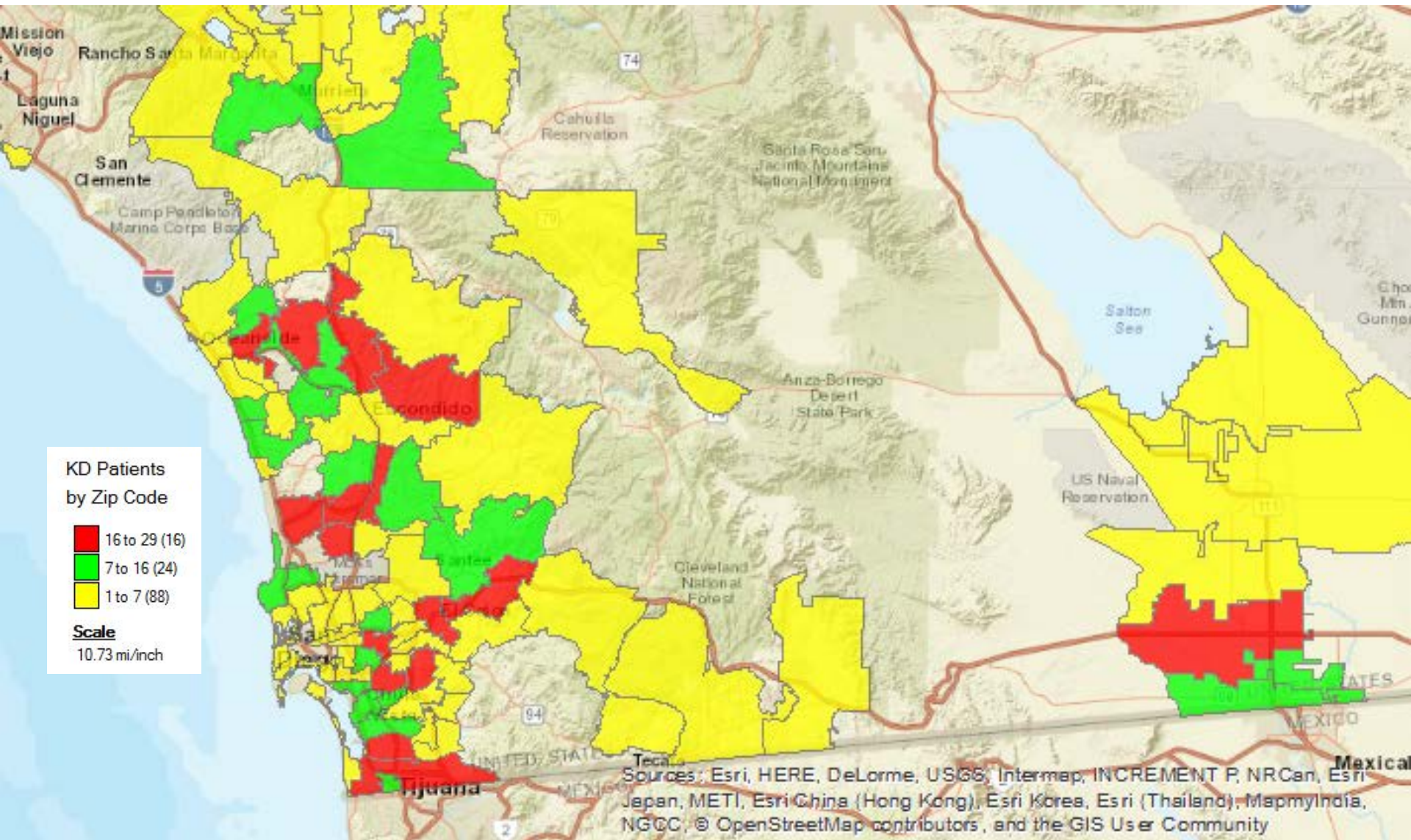
## Race/ethnicity-specific rates of KD at Rady Children's Hospital; onset 2004-2018



\* Excludes Mixed Ethnicity. Includes Missed KD cases diagnosed at RCHSD

# Kawasaki Disease Patients by Zip Code

## June 2015 – December 2016



# Proposed KD Paradigm 2019

Environmental  
trigger



Genetically susceptible host

Genetically resistant host



Manifests immunologic reaction  
(clinical KD)

Asymptomatic



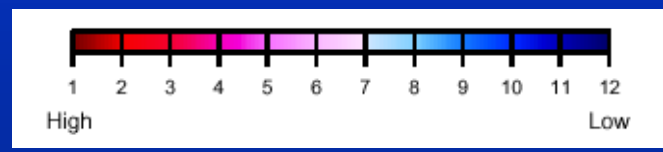
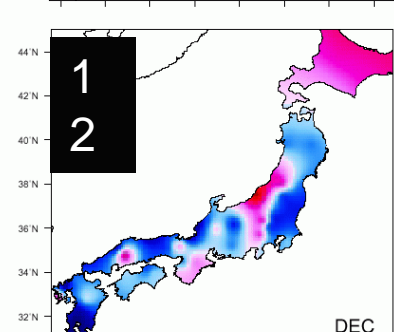
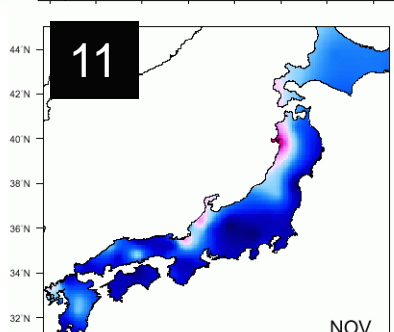
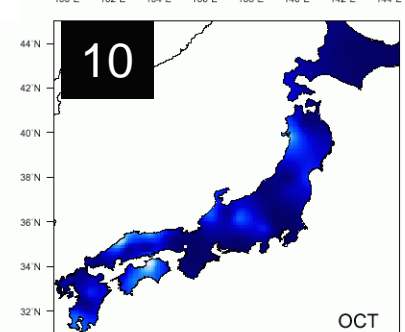
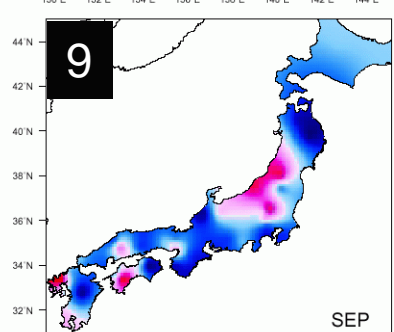
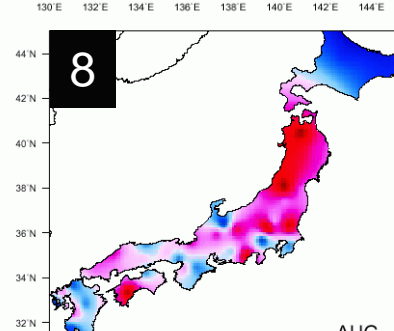
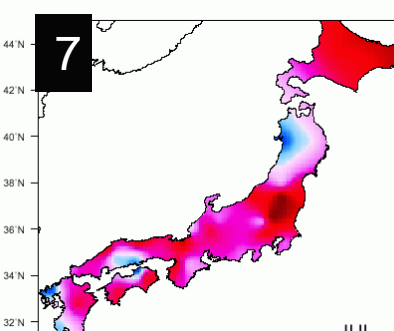
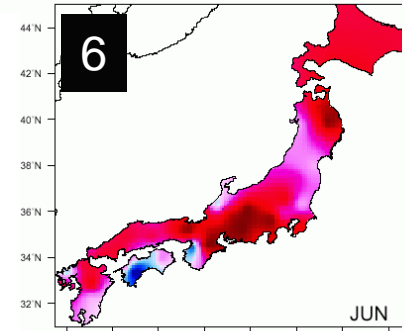
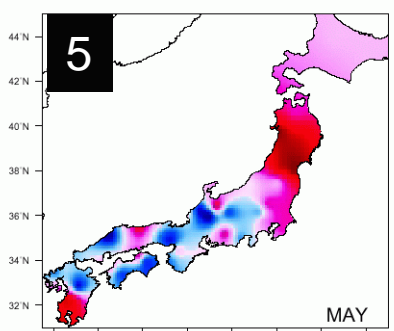
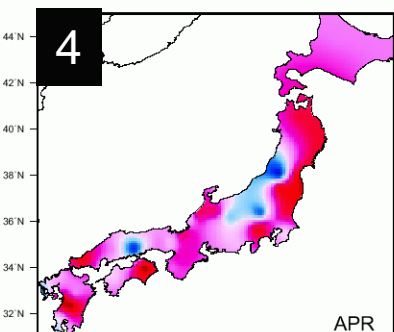
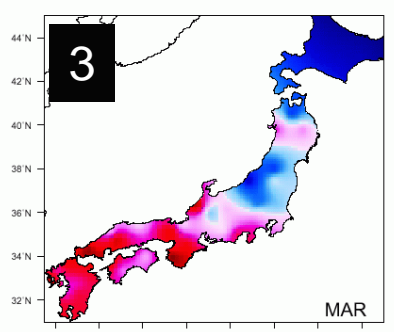
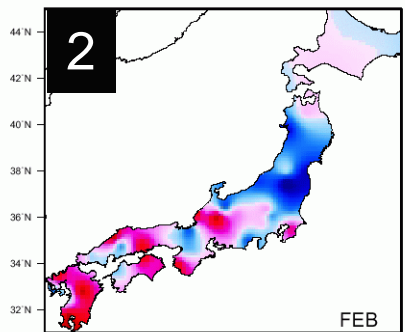
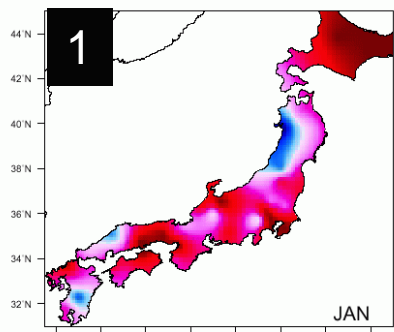
Genetically determined outcome  
(can be modified by treatment)

**Immunity**

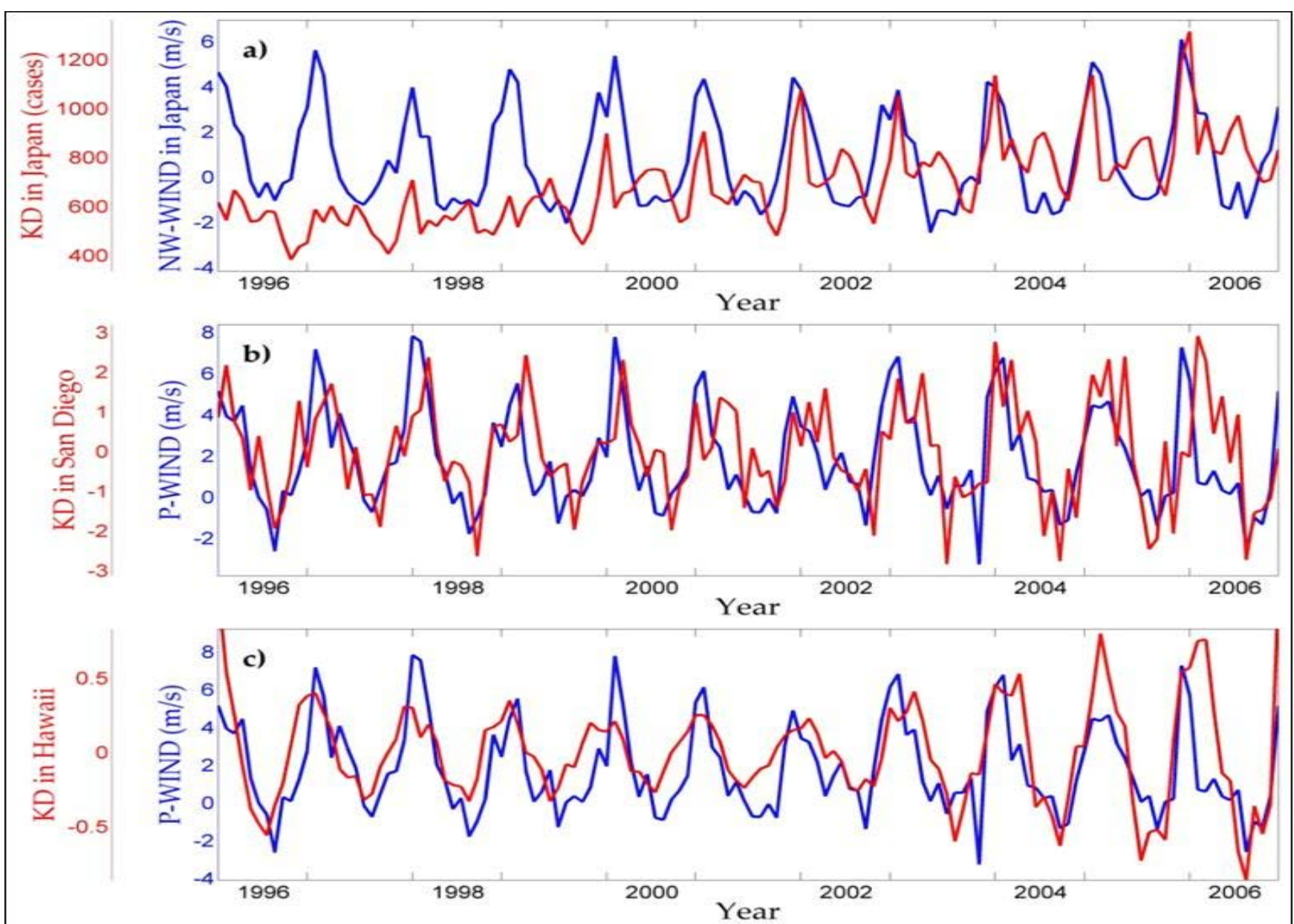
A dramatic sky scene with a bright sun breaking through clouds, casting a strong light over a vast, textured landscape. The sun is positioned in the upper center, creating a lens flare and illuminating the scene. The clouds are dark and heavy on the left, while the right side is dominated by a bright, hazy glow. The foreground shows a textured, possibly snowy or sandy, surface with a path leading towards the horizon.

**“Blowin’ in the Wind”**  
**Nature, April 5, 2012**









**KD and surface winds in Japan (a), San Diego (b) and Hawaii (c).**

# AIRCRAFT DUST SAMPLING MARCH 4, 2011

MAXIMUM ALTITUDE 3,000 M  
SAMPLING TIME 2H 20 MIN

Tokyo

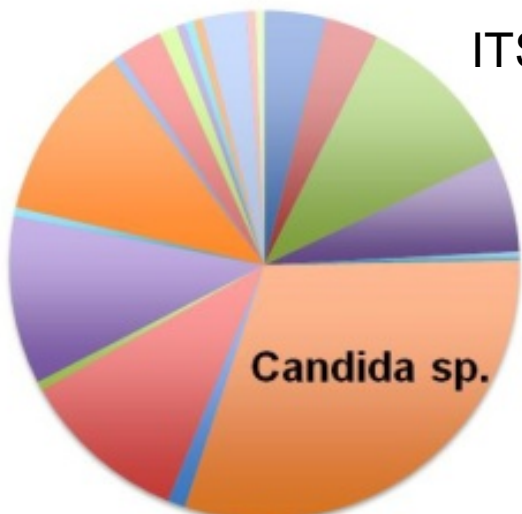






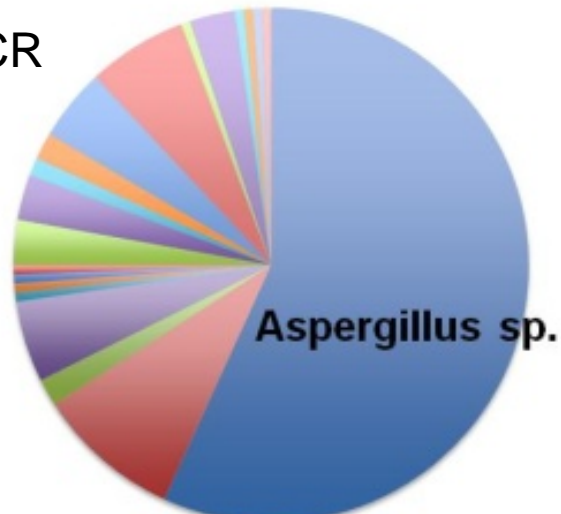
A.

Flight



B.

Surface



C.

Fungi: ITS1-ITS4 region PCR

Flight Filter

Surface Filter

Blank Filter

Reagent Cntrl

PCR Cntrl

Soil + control



D.

Fungi: 18S (817-1536) PCR

Flight Filter

Surface Filter

Blank Filter

Reagent Cntrl

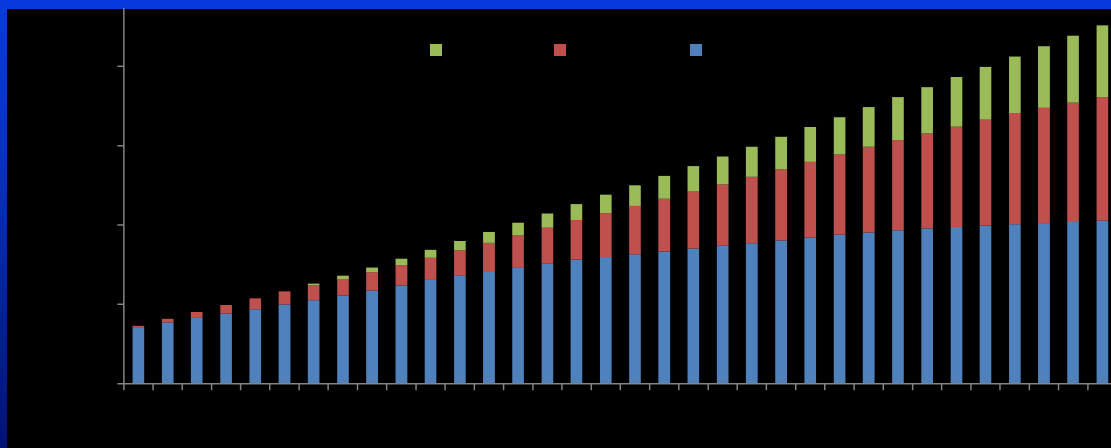
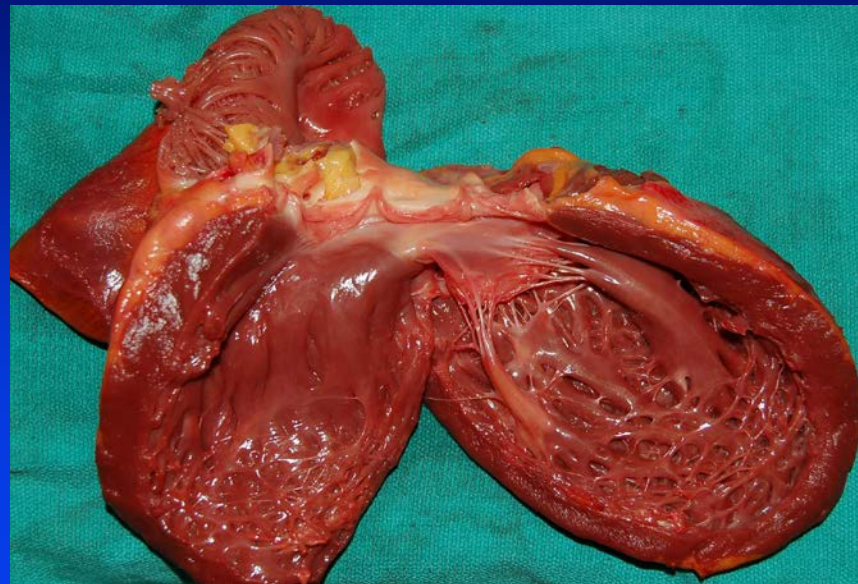
PCR Cntrl

Soil + control



# Lifelong Morbidity

- Aneurysms in 5% of adults <40 years of age with suspected MI
- By 2030: 175,000 US adults with KD and 5500-12,000 with aneurysms



Daniels LB. Circulation 2012;125:2447-53  
Huang SK. J Pediatr. 2013 Jul;163(1):126-31  
Gordon JB. JACC Cardiovasc Interv. 2016;9(7):687-96



Point-of-Care Differentiation of Kawasaki Disease from  
Other Febrile Illnesses

# Clinical + Lab Algorithm

- 2 cohorts from UCSD database
  - Derivation: 276 KD, 243 FC
  - Validation: 136 KD, 121 FC
- Linear discriminant analysis (LDA)
  - Probability of KD

# Multicenter Validation

- 5 KD referral centers (3 PECARN sites\*)
  - Boston,\* Colorado,\* CHOC, Columbus,\* SD
- KD databases
  - 1059 KD, 282 FC



Hao S et al. *Arch Dis Child* 2019  
(accepted pending revision)



Pediatric Emergency Care  
Applied Research Network

Most children with KD arrive  
to RCHSD by private car

So when would  
CHET be involved?

# Scenarios for CHET Transfer

1. Ill-appearing child with fever and rash
2. Shock from KD
3. Acute myocardial infarction



# You've been called to transport a sick child with fever and rash...

## □ Differential Diagnosis

- » Meningococemia
- » Toxic shock
- » Measles
- » Stevens Johnson/TEN
- » Kawasaki disease



# KD Shock Syndrome (KDSS) aka “Kawashocki”

- 2006-2007: Increased # of KD patients required hemodynamic support in PICU
- Evaluated KD patients from 2003-2007
- KDSS if volume expansion or vasoactive support for:
  1. Systolic hypotension for age
  2.  $\geq 20\%$  decrease in SBP from baseline
  3. Clinical signs of poor perfusion

# KD Shock Syndrome (KDSS)

- 7% of KD patients treated between 2003-2007 met the definition of KDSS
- More likely to be female & more inflammation on initial labs
- More likely to be resistant to primary therapy with IVIG
- More coronary artery abnormalities and diastolic dysfunction
- Lower ejection fraction
- Signs of consumptive coagulopathy (+ D dimer, low platelets, high PTT)

# Vasoactive Medications in KDSS patients

- 7/13 KDSS pts (54%) required pressors
- 4/13 KDSS pts (31%) required multiple pressors
- Included dobutamine, dopamine, epinephrine, milrinone, norepinephrine





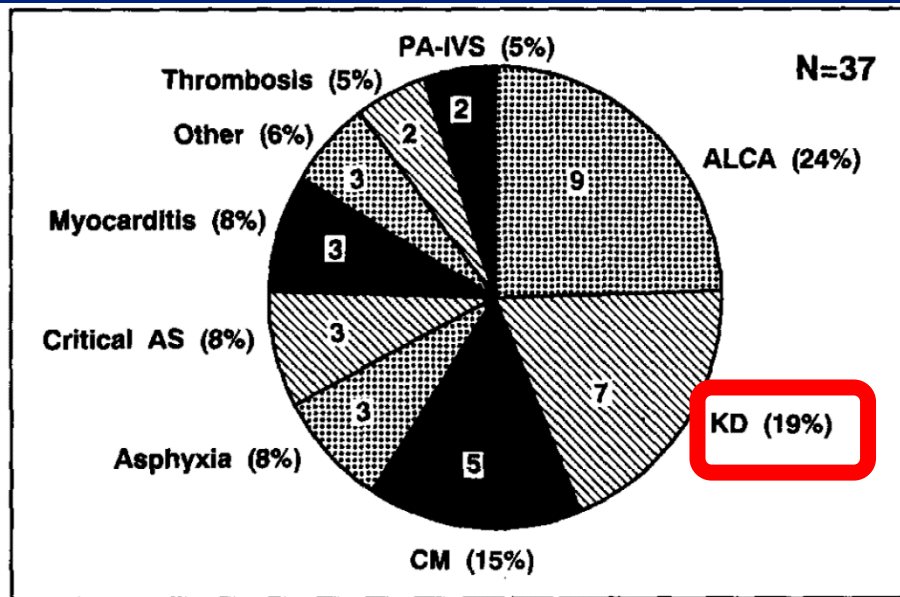
# Support Pre-Hospital of KDSS

- Pump dysfunction (Myocarditis/Cardiogenic shock)
  - » Dopamine or dobutamine
  - » Children <12 months may not respond to dopamine/dobutamine and require epinephrine
  
- Lack of appropriate vasoconstriction (Vasodilatory/Warm shock)
  - » Dopamine
  - » Dopamine-resistant shock commonly responds to norepinephrine

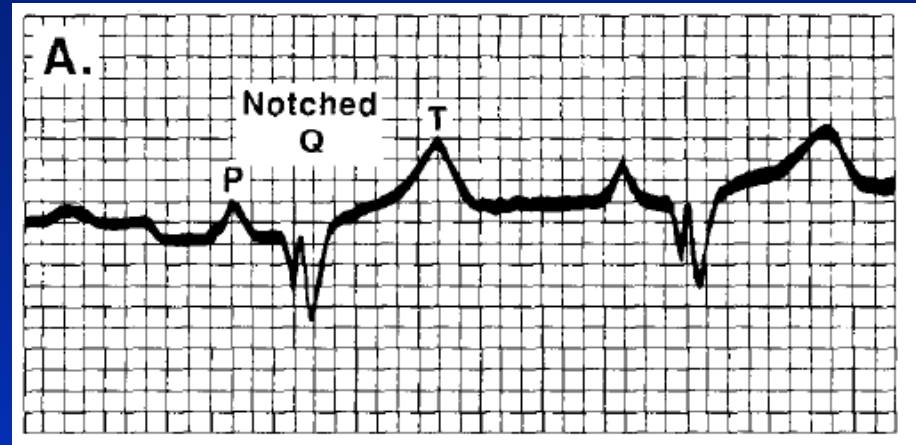
# Who has KD?



# Acute MI in Pediatrics



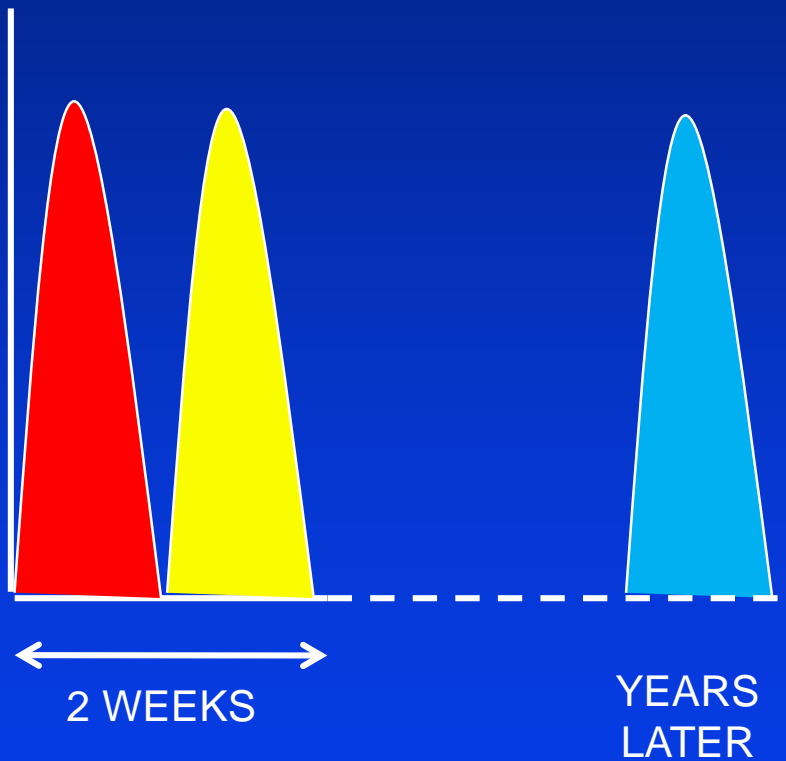
**FIGURE 1. Myocardial infarction in childhood. The distribution of cardiac disorders in 37 patients in which myocardial infarction occurred as seen at autopsy. ALCA = anomalous left coronary artery from the pulmonary artery; AS = aortic stenosis; CM = cardiomyopathy; KD = Kawasaki disease; PA-IVS = pulmonary atresia with intact ventricular septum.**



1. Wide Q waves +/- notched Q
2. ST segment elevation
3. Prolonged QT

- KD is the most frequently acquired cause of acute MI in children

# Timing of AMI in KD



**Acute KD**

**Subacute KD (2 weeks)**

**Convalescent KD (Years)**

# Take Home Points

- You can only diagnose KD if you think about it
- Kawasaki Disease Shock Syndrome:
  - » For KD shock, getting to RCHSD promptly for IVIG is crucial
  - » Be cautious about fluid overload
- Acute MI:
  - » Ask families about KD history
  - » Aspirin and oxygen

# It Takes a Village...



- Patients and their families
- Attendings
- Housestaff
- Emergency dept. staff
- Echo techs
- Lab techs
- Nurses
- Study coordinators
- Pharmacy staff
- KD clinic staff
- Scientific collaborators
- Hospital administrators