### Kawasaki Disease Lori Thompson, APRN, CPNP-PC

### Introduction

This case study describes the presentation of an 18month-old male presenting to the clinic for an episode of acute illness.



# **Subjective**

Reason for selecting this case:

- KD is the leading cause of acquired heart disease in children 6 months to 5 years in the United States (U.S.) and developed countries
- The U.S. has 3000 to 5000 cases of KD per year with an overall incidence of 4-15/100,000
- Occurs in children younger than 8 years; 80% of cases occur under 5 years of age; peak incidence between 9 and 18 months



Seen in pediatric primary care office in rural underserved area

Multiple encounters since birth

Insurance: Blue Cross/Blue Shield

#### **Encounter**



## Subjective: Background Information

History

- 18-month-old male, presenting for an acute episode of illness
- HPI: Chief complaint of "five days of fever, vomiting, and a rash all over."
- PMH: 40 week gestation, vaginal birth without complications
- PSH: Circumcised at birth
- FH: Negative family history
- SH: Lives with parents and is an only child.
- Allergies: NKDA/NDFA
- Medications: None

# **Subjective: ROS**

- **General.** C. U.'s mother reports that he began running fever on Friday and has not been eating well over the last 5 days. He has been irritable and inconsolable all weekend and a rash began to develop 2 days ago.
- Skin/Hair/Nails. Red, rough rash beginning on groin area then spreading to torso, extremities, and face.
- **HEENT:** No history of head or neck injuries. Denies plaques, scaly patches, or rashes to scalp. Reports eyes red but denies discharge. Denies pain or discharge to ears. He has normal hearing. Denies nasal congestion or drainage. C.U's mother reports his lips very red, dry and cracking. His tongue and the inside of his mouth are very red. Denies problems with teeth.

# **Subjective: ROS**

- **CV:** Denies history of cardiac problems or congenital cardiac anomalies. Reports swelling of hand and feet.
- **Pulmonary:** Denies cough, SOB or difficulty breathing.
- **GI:** Reports vomiting for 2 days, diarrhea for 1 day, denies abdominal pain. Loss of appetite for the last 5 days.
- **GU:** Denies pain with or difficulty voiding.
- Integumentary/Hematologic. Reports red rash to trunk, face and extremities. No bruising or bleeding.
- Neurological: C. U. has been irritable and inconsolable for the last 5 days

# **Objective: Physical Exam**

- Vitals: wt 26 lbs, Ht 32 1/2 in., T 101.5 ax, HR 148, RR 28.
- General: The patient is lying on the exam table crying with his mother trying to calm him.
- Eyes: Conjunctiva injected, no exudate. Pupils equal, round, reactive to light and accommodation.
- Nose/mouth/throat: Nares patent no D/C. Pharynx red, tonsils 2+ with erythema, no exudate. Lips red, dry, and cracked. Buccal mucosa with diffuse erythema, + strawberry tongue. Mucous membranes moist. Gag reflex intact.

# **Objective: Physical Exam**

- Lymphatic: Normal, no palpable cervical or inguinal adenopathy
- Respiratory: Chest symmetrical, equal rise, lungs clear
- Cardiovascular: regular rate/rhythm, normal S1S2, no murmurs, equal peripheral pulses, 1+ edema noted to hands/feet
- Abdomen: soft, nontender, no masses, normal BS



# **Objective Physical Exam**

- Integumentary: Warm, dry and pink with capillary refill < 2 seconds. Red, rough rash is noted to his face, trunk, extremities, and the groin area red with peeling skin noted.
- Neurologic: normal tone, no abnormal reflexes





Scarlet Fever

Measles

Drug reaction

**Toxic Shock Syndrome** 

## **Assessment: Impression**

- Kawasaki Disease ICD-9: 446.1
- Presentation of C.U.'s symptoms:
  - High fever (103 ax) that did not remit with antipyretics
  - Red rash all over 2 days after onset of fever
  - The rash began in the diaper area then spread over his torso, extremities and face.
  - Swelling of his hands and feet 3 days after onset of fever
  - Red eyes and red lips began 4 days after the onset of fever



## Etiology



Cause of KD is unknown though many theories abound: Carpet shampoo, Autoimmune disease, Superantigen, Unusual immune response

Research has not supported any of these theories (Rowely & Shulman, 2010).

#### Epidemiologic features:

Age group affected Occurrence of epidemic, with wavelike spread of the illness during an epidemic

Support an infectious cause.

#### Genetics play a role:

tenfold increased risk in Asian populations verses Caucasian populations.

occurs in patients with certain genetic predispositions who contract an infectious agent

## **Clinical Features**

- No prodromal period
- Diagnostic Criteria
  - fever of 5 days or more and the presents of at least four of the following conditions:
    - bilateral (non-purulent) conjunctivitis
    - polymorphous rash
    - changes in the lips and mouth (reddened, dry or cracked lips, strawberry tongue, diffuse erythema of oral/pharyngeal mucosa)







# **Clinical Features**

- changes in the extremities (erythema of palms or soles), edema of hands and feet, desquamation of skin on hands, feet, perineum (during convalescence)
- cervical lymphadenopathy (more than 1.5 mm)
- These classic clinical features are present in 80 to 90% of cases with the exception of cervical lymphadenopathy, which is only present in 50 to 75% of the cases







The course of KD consists of three clinical phases:

Acute

#### Sub-acute

#### Convalescent

#### **Clinical Features**

# **Acute or Febrile Phase**

- Lasts 7 to 14 days ending with the resolution of the fever
- fever is typically high
  - Does not remit with antibiotics/antipyretic medications
  - Characterized by conjunctivitis, mouth and lip changes, presents of the rash, edema of the hand and feet, and cervical lymphadenopathy
  - Cardiovascular manifestations occur
  - Aneurysms occur most commonly one to three weeks after the onset of fever



## **Sub-acute Phase**

- The sub-acute phase is the
  - period of time from the resolution of the fever to day 25
  - consists of;
    - desquamation of the fingers and toes
    - arthritis
    - thrombocytosis



## Convalescent

- Resolution of clinical symptoms until the erythrocyte sedimentation rate (ESR) returns to normal
- Usually 6 to 8 weeks after the onset of KD



## **Evidence Based Practice**



Critical to identify patients with KD and initiate treatment within 10 days of symptom onset

•Early initiation of treatment decreases the rate of coronary artery abnormalities

Peak mortality between
15 to 45 days of the onset
of fever

### **Evidence Based Practice**

Baseline Echocardiogram
High dose Aspirin (80-100 mg/kg/d)
Intravenous immunoglobulin (IVIG)
(2-4 mg/kg) in a single dose

•Goal of Tx: decrease the inflammation within the coronary arteries and prevent thrombosis

Steroid tx is contraindicated

## **Evidence Based Practice**



**Complications:** coronary artery stenosis, thrombosis, myocardial infarction, death In pts with early coronary artery changes on initial echo •40% develop coronary artery ectasia •27% develop coronary artery aneurysms (< 5mm to > 8mm) 20% develop myocarditis •0.07% develop pericarditis

 Later changes on echo: healing and fibrosis of arteries leading to stenosis



### Plan

•C.U. was referred to a North Texas children's hospital

•Labs on admission: •ESR: 47, CRP: 14 •White blood cell count 11.6, 57 poly's and 13 bands demonstrating a shift to the left •Cultures were negative •Echo was normal

•Given high dose ASA and IVIG

### Outcome

•Discharged on day 4 of hospitalization

•Continued low dose ASA for 6 weeks

•No long term cardiac sequela

Cardiology follow up PRN

•High risk for coronary artery disease

#### **Question 1**

What are three of the classic clinical features of Kawasaki Disease?



### **Question 2**

What is the first line treatment for Kawasaki Disease?



### **Question 3**

What role does steroid therapy have, if any, in Kawasaki Disease?

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