IHE Report: A Case of Rheumatic Heart Disease in Rwanda

Yao-Chieh “Jack” Cheng
03/15/2023
Learning Objectives

- Recognize some systemic differences between our health systems.
- Explain the pathophysiology of rheumatic heart disease (RHD).
- Understand the sequelae of rheumatic heart disease.
- Initiate general management for patients with RHD.
The Agaseke basket (uduseke is the plural) is a traditional Rwandan handwoven basket also known as a peace basket.
Land of a Thousand Hills

- Rwanda has 10,169 square miles (Connecticut=5543)
- Typical daily temperature is 54-81°F
- Three national parks (Volcanoes, Akagera, Nyungwe)
- Population 2021: 12.7 million
  - 1.15 million in Kigali
- Physician work force: 1350 (1:8919) in 2018
  - Target 1:7000 by 2024
  - Very few specialists
- Nurse/midwife work force: 12,000 (1:1000)
- Leading cause of death (2019):
  - LRTI, neonatal disorders, CVA, TB, ischemic heart disease, diarrhea, malaria, HIV/AIDS, cirrhosis, MVA
Case

- 20 y.o. man with known RHD (diagnosed 1 yr ago) p/w SOB and CP.
- Pt reported feeling SOB and weak which caused difficulty doing his normal activities. He was not on penicillin therapy.
- Vitals: Temp 98.4°F, BP 120/39, HR 90, RR 28, 89% on 15L NRB
- Initial Exam: Respiratory distress and diaphoretic. Diffuse crackles in all lung fields. Heart RRR - holosystolic murmur. Warm extremities, no LE edema. Confused mental status

What work up do you want?
Initial work up

- Labs:

<table>
<thead>
<tr>
<th>White Blood Count</th>
<th>Hemoglobin</th>
<th>Platelet Count</th>
<th>ESR</th>
</tr>
</thead>
<tbody>
<tr>
<td>140</td>
<td>101</td>
<td>6.2</td>
<td>-</td>
</tr>
<tr>
<td>3.8</td>
<td>-</td>
<td>0.86</td>
<td>-</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Total Bilirubin</th>
<th>Total Protein</th>
<th>Albumin</th>
<th>Creatinine</th>
<th>Urea</th>
</tr>
</thead>
<tbody>
<tr>
<td>12.7</td>
<td>13.8</td>
<td>41.1</td>
<td>225</td>
<td>14</td>
</tr>
<tr>
<td>26</td>
<td>0.8</td>
<td>-</td>
<td>-</td>
<td>1.6</td>
</tr>
</tbody>
</table>

- Bedside echo shows: Severe AR, moderate MR, mild TR, mild pHTN, large LA at 8.7cm, EF 60%, No effusion. Collapsible IVC 1.53
- Further imaging was not performed
Briefly, what is on your ddx?

Sombe with ugali
Briefly, what is on your ddx?

Where should this patient be triaged?

- **Vitals:** Temp 98.4°F, BP 120/39, HR 90, RR 28, 89% on 15L NRB
- **Initial Exam:** Respiratory distress and diaphoretic. Diffuse crackles in all lung fields. Heart RRR - holosystolic murmur. Warm extremities, no LE edema. Confused mental status
Briefly, what is on your ddx?
Where should this patient be triaged?
How do you want to manage him?
Hospital Course (15 days)

- No beds in the ICU, so remained on the medical floors while on 15L NRB
- On admission, he was started on:
  - Lasix IV 80mg TID for presumed pulmonary edema
  - Albuterol nebs
  - CTX and azithromycin (later switched to doxy) for presumed PNA and rheumatic fever
  - Hydrocortisone IV 200mg once for presumed acute carditis
- Cardiology primarily managed his care.
- BMP was checked two more times during his hospital course.
- Completed a 10-day abx course.
- Eventually weaned to room air and switched to PO lasix.
- D/c’ed with lasix PO 20mg and penicillin ppx.
- He was thought to have RHD with CHF
A note on Rwanda’s health system

- Universal insurance coverage with option to upgrade for increased coverage
- Income-based discount on insurance coverage
- Fee-for-service payment model
- Patients or families must pay for medications or studies before receiving.
- Referral based hospital system
- At CHUK, paper charts but electronic ordering for studies and results. One computer per ward.
- 3rd year medical students act as the interns. Residents oversee rounds and run the list with the attending later.
Epidemiology of RHD

- Global prevalence of 40.5 million in 2019
- Causing 306,000 deaths and 10.7 million DALYs
- US prevalence unknown given lack of data
  - US ARF estimated prevalence of <2 per 100,000 school-aged children
  - One study screening low-risk children showed 4/500 with borderline RHD (2013)
- In Rwandan school children in Gasabo district, RHD prevalence of 6.8/1000 school-aged children (2016)
## Revised Jones Criteria

### A. For all patient populations with evidence of preceding GAS infection

| Diagnosis: initial ARF | 2 Major manifestations or 1 major plus 2 minor manifestations |
| Diagnosis: recurrent ARF | 2 Major or 1 major and 2 minor or 3 minor |

### B. Major criteria

#### Low-risk populations*  
- **Carditis**
  - Clinical and/or subclinical
- **Arthritis**
  - Polyarthritis only
- **Chorea**
- **Erythema marginatum**
- **Subcutaneous nodules**

#### Moderate- and high-risk populations
- **Carditis**
- **Arthritis**
  - Clinical and/or subclinical
- **Monoarthritis or polyarthritis**
- **Polyarthralgia**
- **Chorea**
- **Erythema marginatum**
- **Subcutaneous nodules**

### C. Minor criteria

#### Low-risk populations*
- **Polyarthralgia**
- **Fever (≥38.5°C)**
- **ESR ≥60 mm in the first hour and/or CRP ≥3.0 mg/dL**
- **Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)**

#### Moderate- and high-risk populations
- **Monoarthralgia**
- **Fever (≥38°C)**
- **ESR ≥30 mm/h and/or CRP ≥3.0 mg/dL**
- **Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)**
Sequelae of RHD

- Mitral regurgitation and/or stenosis
- Afib
- Thromboembolism
- Aortic regurgitation and/or stenosis
- Heart failure
- Pulmonary hypertension
- Tricuspid regurgitation and/or stenosis
- Increased risk of infective endocarditis
General management

- Structural cardiology referral
  - Standard indications for valve interventions are not changed by RHD
- Secondary prophylaxis of ARF
- Annual flu and dental care
- Pregnant people with RHD are at higher risk
Pathophysiology of RHD

- Valvular damage caused by abnormal immune response to *Streptococcus pyogenes* (GAS) infection
- Exact mechanism is unclear.
  - Antigenic mimicry of rheumatogenic GAS strain, genetically susceptible host, and aberrant host immune response.
- Multiple episodes of ARF increase risk of RHD.
- Dx does not require episode of ARF.
## Secondary prevention of ARF

<table>
<thead>
<tr>
<th>Scenario</th>
<th>Antibiotic choice(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preferred treatment in endemic areas where IM penicillin is available at low cost</td>
<td>▪ IM penicillin G benzathine given every 28 days</td>
</tr>
<tr>
<td>Alternative treatment in nonendemic areas where IM penicillin is unavailable or prohibitively expensive</td>
<td>▪ Oral penicillin V</td>
</tr>
</tbody>
</table>
| Confirmed penicillin allergy*                                            | ▪ Preferred – Oral azithromycin  
  ▪ Alternative – Oral sulfadiazine                                               |
| Severe symptomatic RHD                                                   | ▪ Preferred – Oral penicillin V  
  ▪ Alternatives – Oral azithromycin or oral sulfadiazine                           |
| Bleeding problems following IM injection that cannot be addressed        | ▪ Preferred – Oral penicillin V  
  ▪ Alternatives – Oral azithromycin or oral sulfadiazine                           |
| Other barriers to using the preferred treatment that cannot be resolvedΔ | ▪ Oral penicillin V                                                               |
| Patients at low risk of recurrence♂                                     | ▪ Oral penicillin V                                                               |
| Breakthrough infection while on prophylaxis                               | ▪ For treatment of acute infection – Oral clindamycin   
  ▪ For ongoing prophylaxis – IM penicillin G benzathine given every 21 days     |
Secondary prevent of ARF Duration

- Ppx against ARF is unlikely to alter natural hx of RHD once symptomatic valve dysfunction occurs
- Secondary ppx of latent RHD reduces the risk of echocardiographic progression

<table>
<thead>
<tr>
<th>Category</th>
<th>Duration after last attack</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rheumatic fever with carditis and residual heart disease (persistent valvular disease*)</td>
<td>10 years or until 40 years of age (whichever is longer)</td>
</tr>
<tr>
<td></td>
<td>Sometimes lifelong prophylaxis (refer to UpToDate topics on treatment and prevention of acute rheumatic fever and management and prevention of rheumatic heart disease)</td>
</tr>
<tr>
<td>Rheumatic fever with carditis but no residual heart disease (no valvular disease*)</td>
<td>10 years or until 21 years of age (whichever is longer)</td>
</tr>
<tr>
<td>Rheumatic fever without carditis</td>
<td>5 years or until 21 years of age (whichever is longer)</td>
</tr>
</tbody>
</table>
References

Thank you!!!
(Murakoze)
Questions?