Sickle cell crisis in Western Europe

Bart Biemond
Academic Medical Centre Amsterdam
Disclosures

- No disclosures
History

- Student from Grenada

- Symptoms:
  - Painful attacks
  - Chronic leg ulcers
  - Anemia
  - Jaundice

- Microscopy:
  - Sickle-formed red cells
  - “Herrick syndrome”

James Herrick 1910
Etiology

- Single nucleotide mutation in the $\beta$-globulin gene
- HbA to HbS
**Western society:**
US: 100,000 patients
UK: 15,000 patients
NL: 2,000 patients
Worldwide: >300,000 children born/yr
Survival children

- Vaccinations
- Antibiotics
- Transcranial Doppler screen
- Transfusions
- Neonatal screening
- Comprehensive care

Survival adults

Kaplan–Meier survival curves. (A) Survival curve by sickle genotype. (B) Survival curve for HbSS/β0, by hospitalization frequency.

HbSS/HbSβ0:
- 90% > 45 jr
- 80% > 50 jr
- 70% > 60 jr

## Clinical presentation

<table>
<thead>
<tr>
<th>Acute complications</th>
<th>Chronic complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute vaso-occlusive crisis</td>
<td>Osteonecrosis</td>
</tr>
<tr>
<td>Acute chest syndrome</td>
<td>Pulmonary hypertension</td>
</tr>
<tr>
<td>Infection</td>
<td>Renal failure</td>
</tr>
<tr>
<td>Hepatic complications</td>
<td>Cardiomyopathy</td>
</tr>
<tr>
<td>Priapism</td>
<td>Retinopathy</td>
</tr>
</tbody>
</table>
Acute crisis

- Extreme painful
- Relapsing & disabling
- Ischemia
- Particular located in bones
- No objective test!!
- Patients tells you the diagnosis!
- Often misunderstood…
Pain crisis treatment

- Immediate pain treatment!!

- Adequate triage at ER

- Oxygen if hypoxia (sat<95%)

- Antibiotics if fever (temp>38,5 C)

- Red cell transfusion (if Hb<3.5 mmol/l; 5,5 g/dL)
Retrospective study in Chicago

<table>
<thead>
<tr>
<th>Triage level</th>
<th>Site 1</th>
<th>Site 2</th>
<th>Site 3</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Level 1</td>
<td>0,5%</td>
<td>0</td>
<td>0</td>
<td>0,2%</td>
</tr>
<tr>
<td>Level 2</td>
<td>38%</td>
<td>25%</td>
<td>3,5%</td>
<td>27%</td>
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<tr>
<td>Level 3</td>
<td>61%</td>
<td>73%</td>
<td>91%</td>
<td>71%</td>
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<tr>
<td>Level 4</td>
<td>0,5%</td>
<td>1%</td>
<td>5,5%</td>
<td>1,5%</td>
</tr>
<tr>
<td>Level 5</td>
<td>0</td>
<td>9%</td>
<td>0</td>
<td>0,3%</td>
</tr>
</tbody>
</table>

Median time to analgesia 90 min (54-159)

Tanabe et al. Acad Emerg Med 2007
Prejudices

- Discordance in pain score and presentation
- Negative perception
- Stigmatized

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**Shapiro et al. J Pain and Symp Manag 1997**

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**Fig. 2.** Percentage of adults perceived as addicted. Responses of emergency department physicians are depicted by the solid bars; responses of hematologists are depicted by the gray bars.
Circle of harm

Poor experience of care

Delay seeking help

Poorly controlled pain

Increased opiate requirements

Negative staff perceptions
Acute Chest Syndrome

- Severe pulmonary complication
- Life-time risk: 30%-50%
- Mortality: 4.3% (ICU 19%)
- Caused by:
  - Infection
  - Fat emboli (2th day of admission)
  - Hypoventilation

*Minter et al. Am J Respir Crit Care Med 2001.*
*Johnson et al. Semin Respir Med 1988*
ACS
definition/pathogenesis

Definition:

- Respiratory symptoms
  - Fever
  - Coughing
  - Dyspnea
  - Hypoxia

- Chest X-ray: new infiltrate

*Rees D. Lancet 2010*
Treatment ACS

- Antibiotics
- Oxygen
  - (sat > 95%)
- On-top transfusion
  - (Max: Hb 6.2 mmol/L; 10 g/dl)
- Exchange transfusion (erythropheresis)
  - HbS% < 30%
Evolving new therapies

- Anti-adhesive therapies
- P-selectin inhibition
- Anti-sickl
‘Vicious circle’ of SCD

- Anti-adhesion
- Anti-oxidants
- HbF induction
- Anti-sickling

Thank you