Acute chest syndrome in children
A management protocol

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## Audit of management of ACS in OLCHC

**January-August 2016**

<table>
<thead>
<tr>
<th>N = 16 /36 events M=F; Age 3-16yrs</th>
<th>number</th>
<th>percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Antibiotics</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ceftriaxone</td>
<td>14</td>
<td>87.5%</td>
</tr>
<tr>
<td>clarithromycin</td>
<td>14</td>
<td>87.5</td>
</tr>
<tr>
<td></td>
<td>13</td>
<td>81</td>
</tr>
<tr>
<td>Supplemental oxygen</td>
<td>12</td>
<td>75%</td>
</tr>
<tr>
<td><strong>IV fluids (60-80% maintenance)</strong></td>
<td>16</td>
<td>100%</td>
</tr>
<tr>
<td>antipyretics</td>
<td>9</td>
<td>56.2%</td>
</tr>
<tr>
<td>analgesics</td>
<td>11</td>
<td>68.7%</td>
</tr>
<tr>
<td>Prednisolone</td>
<td>16</td>
<td>100%</td>
</tr>
<tr>
<td>bronchodilators</td>
<td>15</td>
<td>93.7%</td>
</tr>
<tr>
<td>Simple transfusion</td>
<td>6*</td>
<td>37.5%</td>
</tr>
<tr>
<td>Exchange transfusion</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Incentive spirometry</td>
<td>15</td>
<td>93.7%</td>
</tr>
<tr>
<td>Chest physio</td>
<td>15</td>
<td>93.7%</td>
</tr>
<tr>
<td>Chest drain</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Icu admission/mechanical ventilation</td>
<td>0</td>
<td></td>
</tr>
</tbody>
</table>
Outcomes

• No deaths

• Duration of stay
  – <5 days; 1 patient
  – >5 days; 15

• Readmission within 30 days
  – N = 2 (12.5%)
  – No pain events
Definition ACS

• An acute illness
• Fever
• Increased work of breathing
• New radiological density on chest X-ray

• Hypoxia and decline in respiratory status (often within 24 hours) is a useful predictor of severity and outcome.
• Fall in platelets signals more severe disease

De Baun et al Lancet 2016;387:2545-2553
Howard et al BrJHaematol 2015,169;492-505
Radiological changes

Day 1

Day 2
Causes of ACS

Atelectasis
- Pain due to rib and vertebral infarction
- Reduced respiratory drive and cough

Fat embolism
- Bone marrow infarction
- Release of phospholipase A2

Acute chest syndrome

True thromboembolism
- Activated endothelium
- Reduced red cell flexibility
- Increased red cell adherence
- Vaso-occlusion

Infection
- Chlamydia pneumoniae
- Mycoplasma pneumoniae/ hominis
- Staphylococcus aureus
- Streptococcus pneumoniae
- Respiratory syncytial virus
- Parvovirus
- Rhinovirus
- Influenza and parainfluenza virus
Inflammatory disorder

- Significant morbidity
- Significant mortality
- Association with asthma
- SCD also associated with bronchial hyperreactivity
- Combination of atopic asthma and SCD is associated with worse patient outcomes

Investigations

- History (previous diagnosis of asthma/wheezy episodes)
- Examination
- Pulse oximetry
- FBC + Biochemistry testing (Na, Creatinine, LFTs)
- Blood cultures, sputum culture, NPA for viral testing
- Group and crossmatch (Full Rh & kell)
- CXR

*De Baun et al Lancet 2016;387:2545-2553*

*Howard et al BrJHaematol 2015,169;492-505*

*Lazarus et al Am J Hematol 2016,91;8: E332-E333*

*Miller ST Blood 2011,117(20):5297-5305*
Treatment
(the consensus)

- Prevention (incentive spirometry for VOC)
- Antibiotics
  - Cephalosporin and macrolide
- Pain relief
  - “adequate to prevent hypoventilation”
- Supplemental oxygen (keep O2>95%)
- Simple (top up) transfusion
  - for hypoxia
  - Deterioration in respiratory status
  - ↓Hb level ≥ 2g/dl below baseline
- Exchange transfusion
  - If Hb over 9g/dl
  - ?worsening status despite the above
- ICU admission + supported ventilation
The supporting data? None!!!
Other possible treatments (controversial)

- Bronchodilators
- Corticosteroids
- Inhaler nitric oxide
Bronchodilators?

Argument for
- ↑incidence of asthma in children with SCD
- ↑association of asthma with ACS
- Often bronchial hyperreactivity with ACS

Argument against
- If no wheeze no logical reason to use

If hx of asthma or evidence of wheeze – add bronchodilator
Corticosteroid?

For

• Dexamethasone 0.3mg/kg/dose 12 hourly for 4 doses attenuated the course of ACS (mild-moderate)
  Bernini et al Blood 1998;92(9):3082-3089

• Prednisone 2mg/kg/day x5-7 days to improve outcome without rebound pain

Against

• Increased readmission rate with pain events post steroids

• Prolonged admissions (retrospective analysis)
  Strouse et al 2008
  Sobota et al 2010
  Quinn et al 2011

But it is appropriate therapy for Asthma!
“Approaches to monitoring of end organ damage within Paediatric SCD”
Long-term outcomes of ACS

• Early death
  – Recurrent ACS
    • Asthma
    • 1st ACS <4 years

• Pulmonary function abnormalities
  – Normal
  – Obstructive/reversibility
  – Restrictive

• Pulmonary fibrosis/chronic lung disease
Longer term strategies

• Annual flu vaccine/pneumococcal vaccines
• Avoid smoking
• Identify those with asthma, treat adequately
• Pulmonary function tests
• Joint respiratory/sickle clinics
• Offer HU to anyone who has had ACS
• Blood transfusion programme if HU failure
• Consider HSCT