

Acute chest syndrome in children

A management protocol

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Audit of management of ACS in OLCHC

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

Outcomes

- No deaths
- Duration of stay
 - <5days; 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

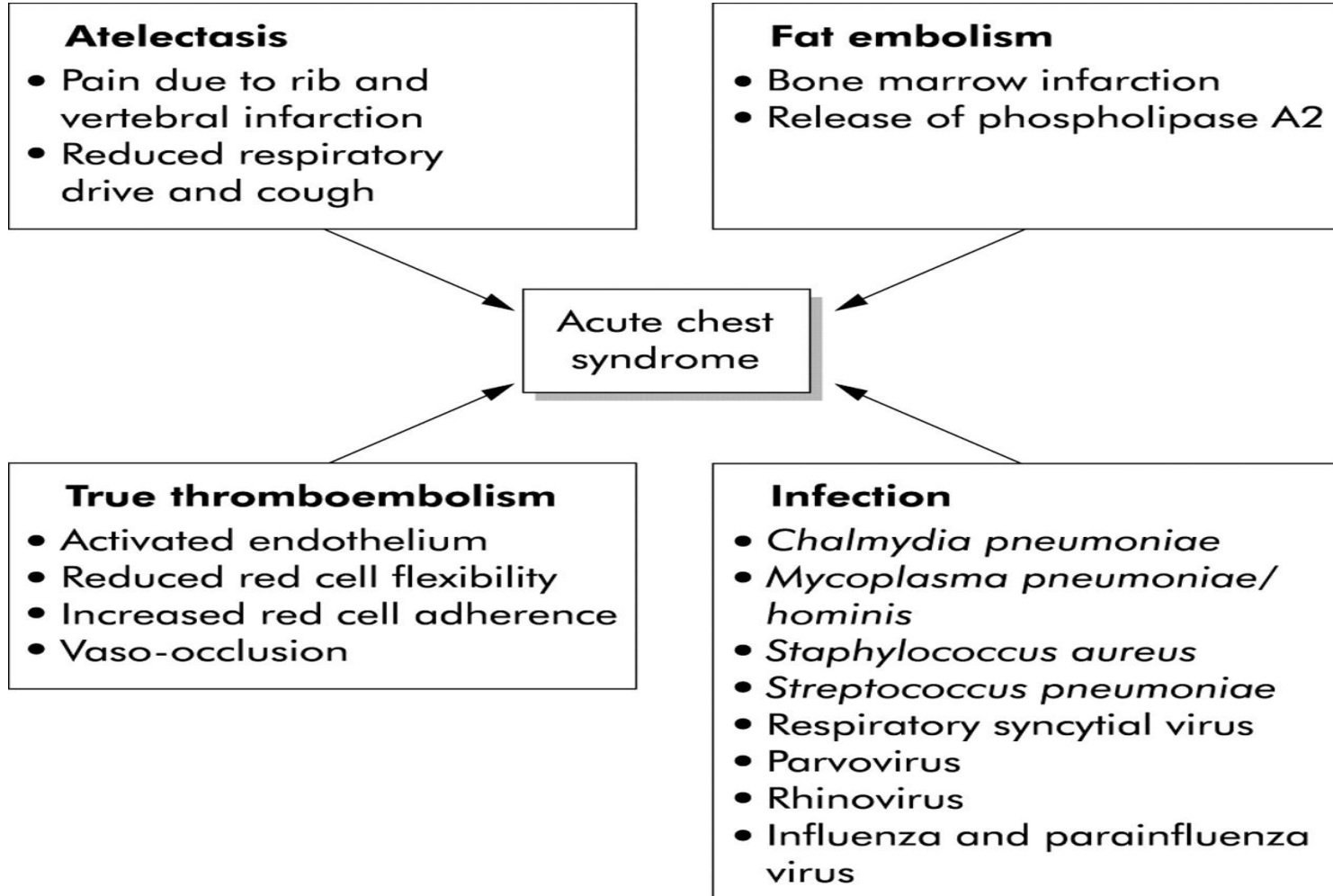
Day 1



Day 2



Causes of ACS



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

Knight-madden et al Paediatr Respir Rev 2014;15(1):13-6

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-
7days to improve outcome
without rebound pain
*Kumar et al J Pediatr Hematol Oncol
2010;32(3):e91-94*

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