Case Presentation
Duchenne's Muscular Dystrophy

Partners in Paediatrics 2016
15 year old male
Duchenne’s Muscular Dystrophy
Fall 24 hours before with impacted femoral #
Admitted for analgesia and physiotherapy
Unwell
No access
NK organising emergency transfer
Unwell...
RR 30-40
Saturations 78% on air, high 90s with O2
HR 125
BP 100/70
Temp 38
GCS 15
1915

Background history

Florid Cushing’s Syndrome
Able to transfer and stand aided
Last echo EF 50%, asymptomatic
Recent admission to PICU Telford pneumonia requiring inotropes but not ventilated
Complex family dynamics
2015

Access established

Examination, warm peripherally but CRT 3-4 seconds, bronchial breathing L side ++ Low BM

CXR and bloods
2015-2115

Working DX

Pneumonia and sepsis

?? PE/Fat embolism

Treatment and assess

Fluid bolus/ steroid/ Antibiotics/ glucose

Response modest initially

Definitive PICU required
Bed in Stoke secured

Scoop and run?

Intubate when KIDS team arrives?

Intubate/stabilise now?

Call a team

Call a colleague
Anaesthetic room 6
2230

Induction went exactly as anticipated

Stormy
- Non compliant patient
- Awkward intubation
- Difficult access
- Unstable BP
- High inflation pressures

KIDS Team arrived at 2400
Left at about 0230
Debrief organised for following Tuesday

Good Teamwork
Appropriate and timely escalation
Excellent nursing of frightened patient
Calm and co-ordinated resuscitation
Transfer policy followed and it worked

No change following morning

Phone call 2030- patient had arrested and died
What could of be done better?

- Diagnosis?
- EWS system?
- Time to intervention?
- Documentation?
- Missed fracture @RSH?
- Anaesthetic care RJAH?
- PICU care in Stoke?
Diagnosis Fat Embolism- made at post mortem.

Not uncommon finding at PM for unexplained deaths in DMD
Not always a fracture or significant trauma
BIPHASIC pathophysiology- “embolic” then inflammatory
Learning Points

High index of suspicion of Fat Embolism in DMD patients with unexplained illness.

KIDS team takes time to mobilise.

Always be able to call another colleague.