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INDÉPENDANCE ET EXPERTISE POUR LE PROGRÈS DE LA SANTÉ À DOMICILE

3rd INTERNATIONAL PEDIATRIC

NONINVASIVE VENTILATION CONFERENCE

ASSISTANCE PUBLIQUE HÔPITAUX DE PARIS

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Necker ENFANTS MALADES HÔPITAL UNIVERSITAIRE

Workshop:
Clinical Cases: complex OSA

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The child first and always

UCL INSTITUTE OF CHILD HEALTH

Great Ormond Street Hospital for Children NHS Foundation Trust

JK

- Born at term LSCS
- Antenatally diagnosed pericardial effusion
- Presented to local hospital collapsed needing CPR and intubation

- Dilated cardiomyopathy with severe BV dysfunction
- Multiple small apical VSDs

- PA banding, PDA ligation May 2012 (aged 2 months)

- Discharged with diuretics / ACE inhibitors after 3 weeks

JK

- Re-admitted to RBH in August 2012 with cardiac failure
- Left lower lobe collapse only responding to invasive ventilation
- Tracheostomy for long-term ventilation September 2012
- Listed for transplant

- Cardiac transplant Nov 2012 (aged 8 months)

- Discharged early 2013 with usual immunosuppression (Tacrolimus, MMF)

JK

- Tracheostomy decannulation March 2013 (1 y.o.)
- Since decannulation →

- Mild stridor on exertion during the day
- Struggling with breathing at night (tracheal tug, recession)
- Sleeping with neck hyper-extended
- Disturbed sleep
- Worsened by intercurrent upper airway infection
- Given prednisolone

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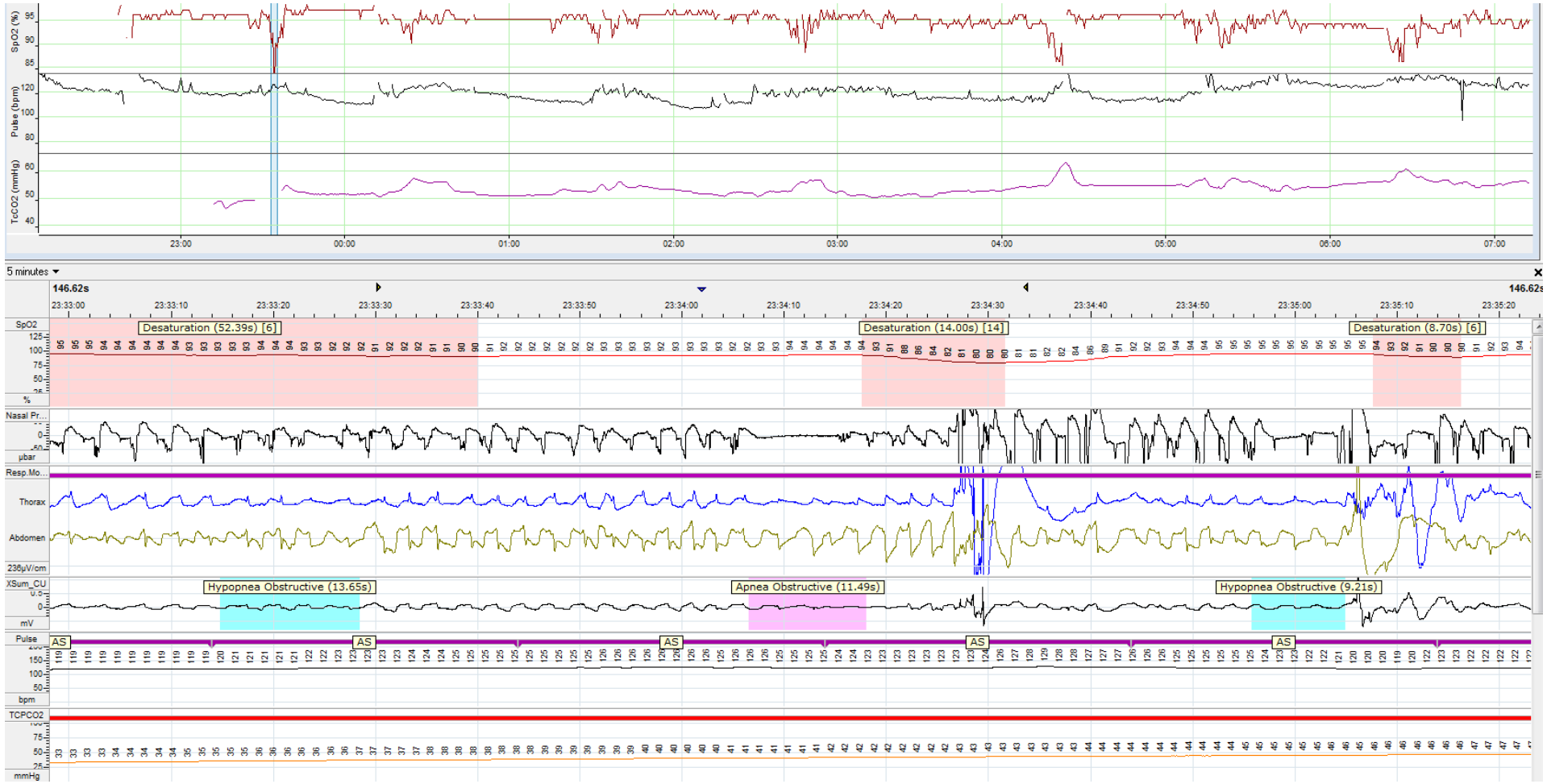
First study July 2013



SS July 2013

- Clinical evidence of obstruction
- Moderate OSA with OAHI 8ev/hr
- ODI 18 dips/hour to a nadir of 81%
- Obstructive hypoventilation: CO2 mean 53mmHg (7.1kPa) – maximum 62mmHg (8.3kPa) – above 50 mmHg all night

What do you do next?

A) Refer back to ENT to investigate the cause for OSA

B) Start CPAP

C) Start BiPAP

D) Reinsert tracheostomy and ventilate via tracheostomy

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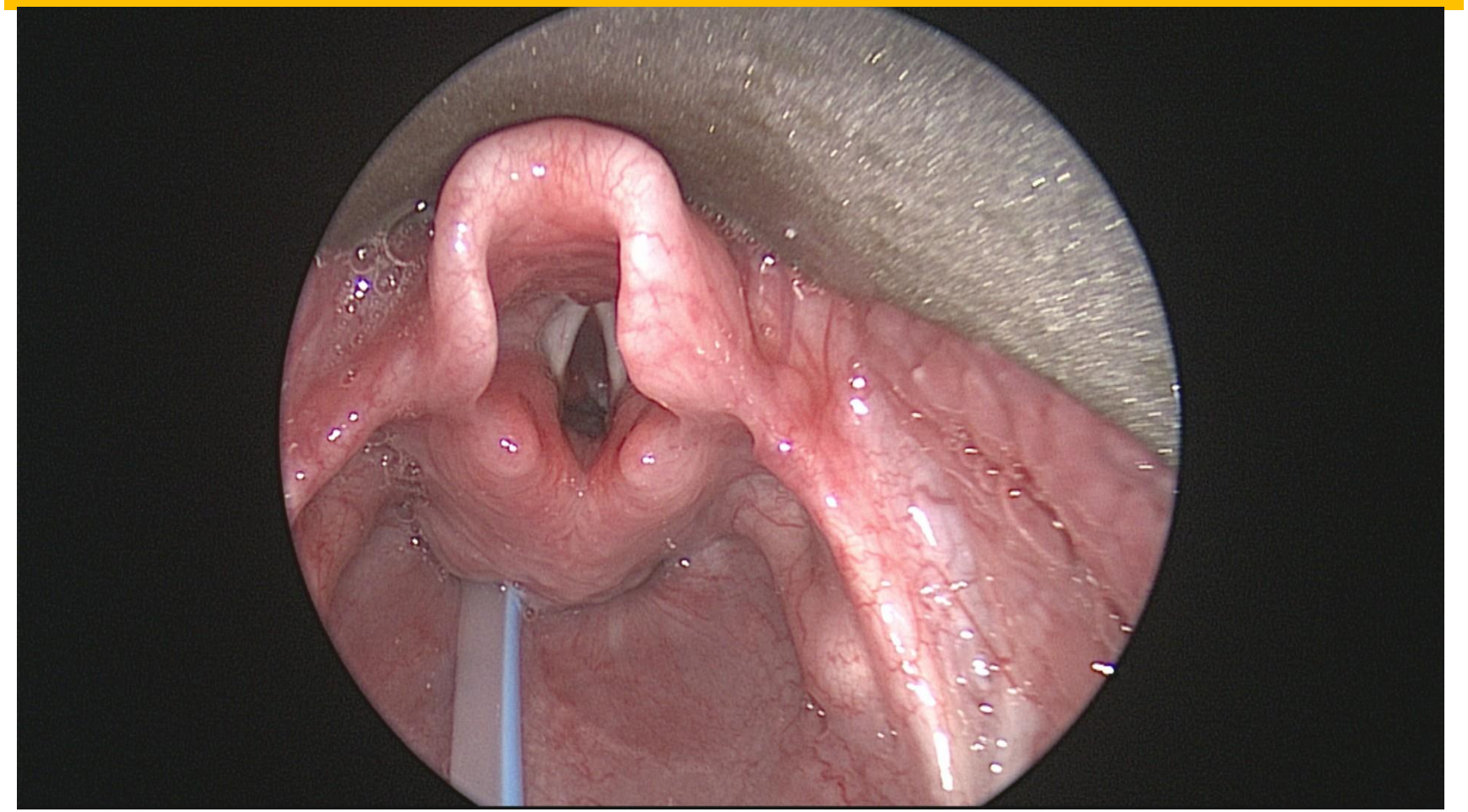
MLB



Mild laryngomalacia – mild subglottic granulomatous tissue
No intervention required

What do you do next?

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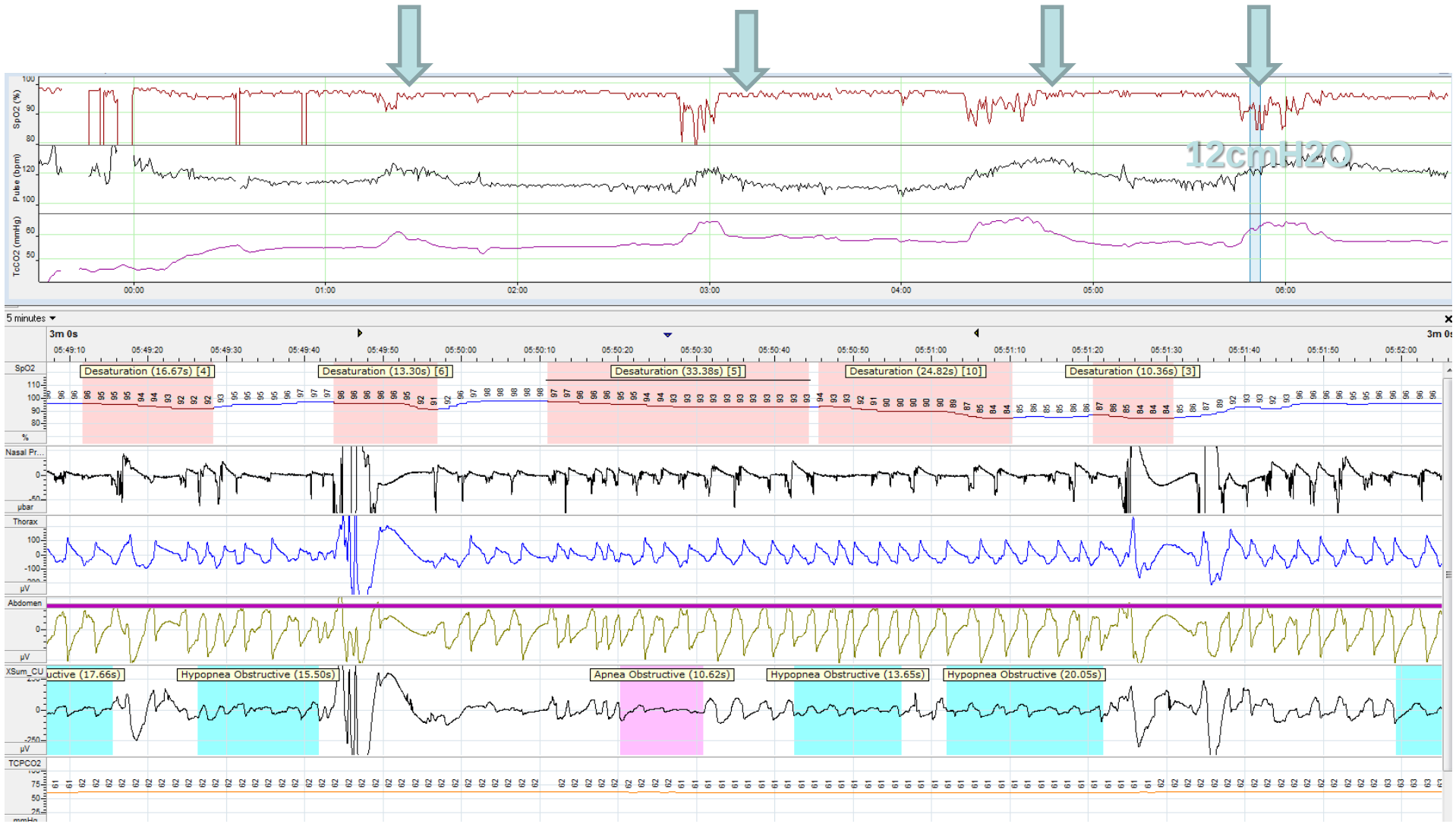
Rationale for clinical decision

1. The OSA is clearly the reason for the hypoventilation (obstructive hypoventilation) and there is a good chance that the hypoventilation will disappear if OSA is abolished – CPAP should be tried first

2. Tracheostomy would only be a last resort after NIV has been shown to fail

Follow up SS – CPAP trial (3 months later)



Follow up SS

- Sleep study on CPAP titrated up to 12cmH2O
- Clinical evidence of obstruction persist even on high pressure
- Moderate to severe OSA persist with OAHI 10ev/hr
- ODI 16 dips/hour to a nadir of 80%
- Obstructive hypoventilation: CO2 mean 53mmHg (7.1kPa) – maximum 62mmHg (8.3kPa) – above 50 mmHg all night

What do you do next?

A) Increase CPAP from 12cmH2O to 20cmH2O on second night

B) Start BiPAP at 18/12cmH2O and titrate as required

C) Start BiPAP at 12/6cmH2O and titrate as required

D) Reinsert tracheostomy as NIV is ineffective

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D) Reinsert tracheostomy as NIV is ineffective

Rationale for clinical decision

1. Despite a first night of titration up to 12cmH2O OSA was not affected and was even worse than on diagnostic study – there is no benefit in increasing pressure further

2. BiPAP works for refractory OSA not responding to CPAP – the extra inspiratory pressure (IPAP) can force open reluctant obstruction

3. You don't have to start with same CPAP as where you finished your titration on CPAP (i.e. 12cmH2O) – the benefit of adding IPAP means a lower CPAP can be tried

4. A differential of at least 6cmH2O between the two pressures is necessary to benefit from the IPAP effect

5. Tracheostomy will only be a last resort after BiPAP has been shown to fail

?Trial of BiPAP

Physiologists attempted to fit the interface numerous times throughout the night but Jakub would become increasingly distressed and would not allow for the interface to be fitted numerous times throughout the night

Jacob has become intolerant to NIV and OSA very severe (OAHI 45/hr)!

What do you do next?

A) Reduce BiPAP pressure to 10/4 to help with compliance

B) Change interface and add humidification

C) Investigate the reason why even BiPAP is not overcoming obstruction – repeat MLB

C) Enough is enough – the tracheostomy needs to go back in

What do you do next?

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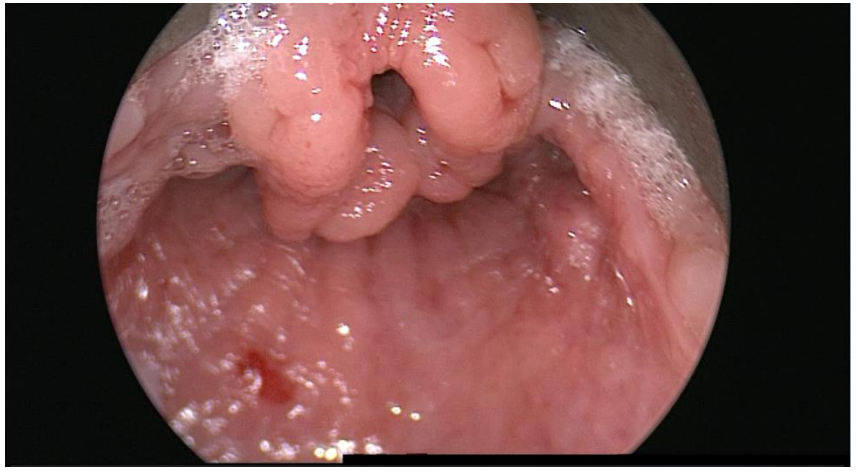
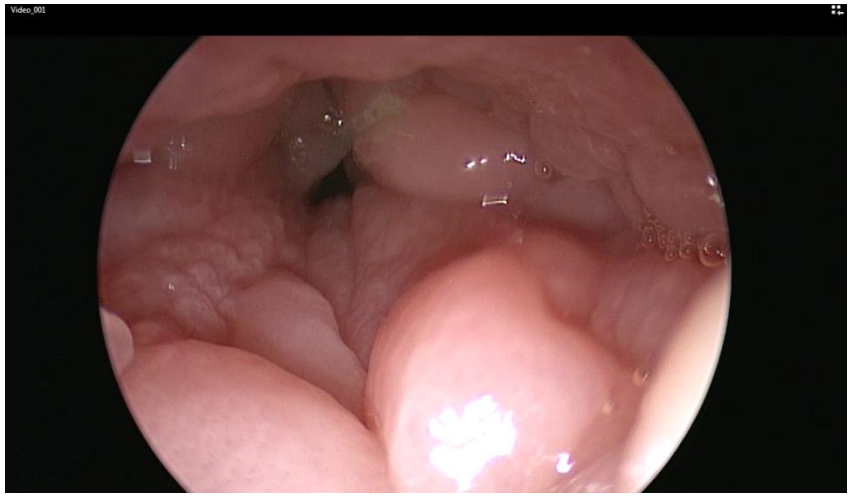
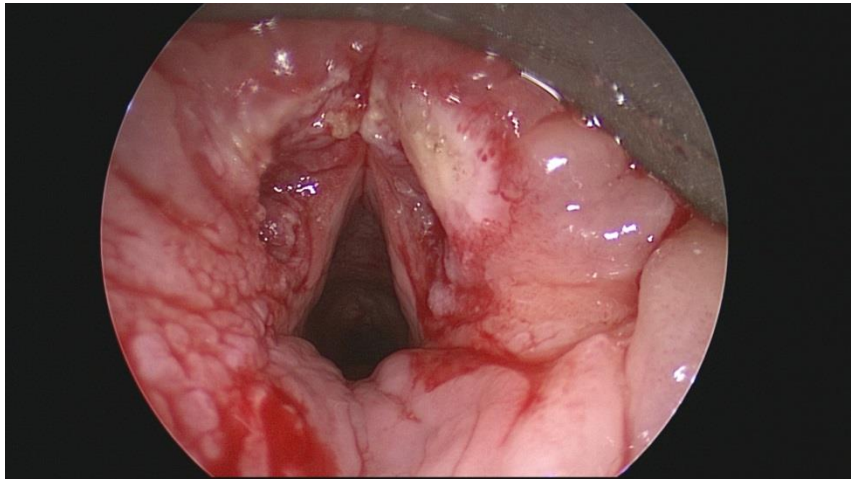




Severe laryngeal papillomatosis with almost complete obstruction of glottis



View post surgical debridement

Learning points

- Never lose focus on the patient and the underlying condition rather than the ventilation alone.

- Progression in treatment modalities in refractory OSA
  CPAP -> BiPAP -> Tracheostomy as a last resort

- Don't be afraid to reassess the aetiology of SDB in the face of challenges

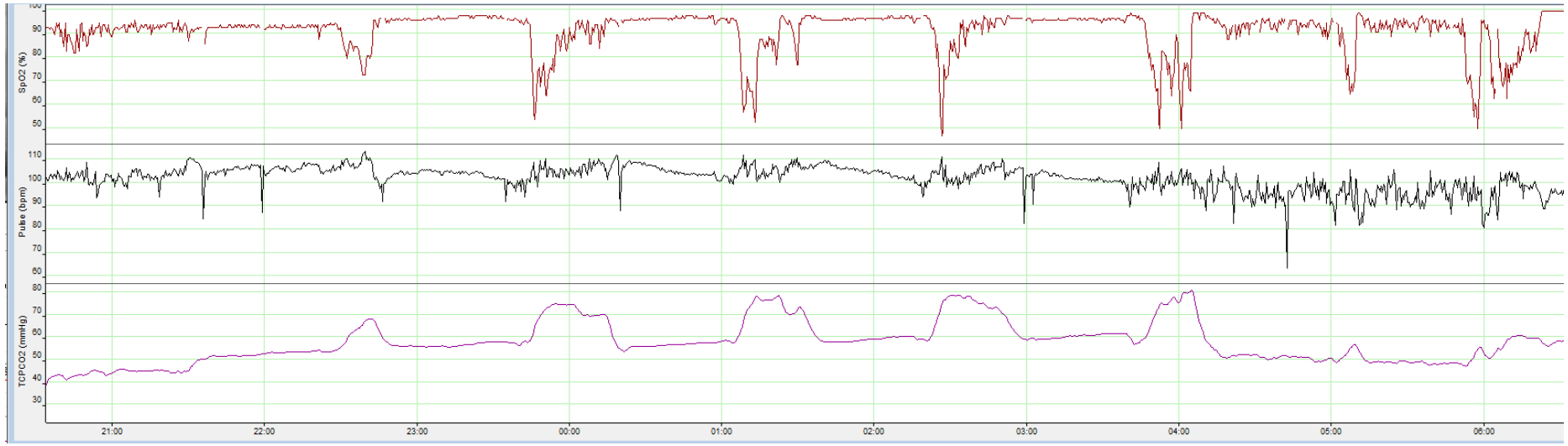
2 contrasting clinical cases with CP

LB

- 15 year old young man
  - X-linked hydrocephalus
  - CP (GMFCS V)
  - seizures,
  - GORD
  - PEG-fed
  - Adenotonsillectomy in 2015 (afterwards PICU admission)
  - scoliosis
  - snoring and daytime tiredness

LB

Baseline sleep study: severe obstructive hypoventilation



- Mean SpO_2 93%
- ODI 29 dips/hr (absolute nadir of 47%)
- Mean $TcCO_2$ 60mmHg (Max 80mmHg)
- Obstructive apnoea hypopnoea index = 21.7 events/hr (significantly raised)

What is your management plan?

1. Re-refer to ENT for revision adenotonsillectomy
2. Start CPAP
3. Start BiPAP
4. Consider alternative treatment options such as nasopharyngeal airway

What is your management plan?

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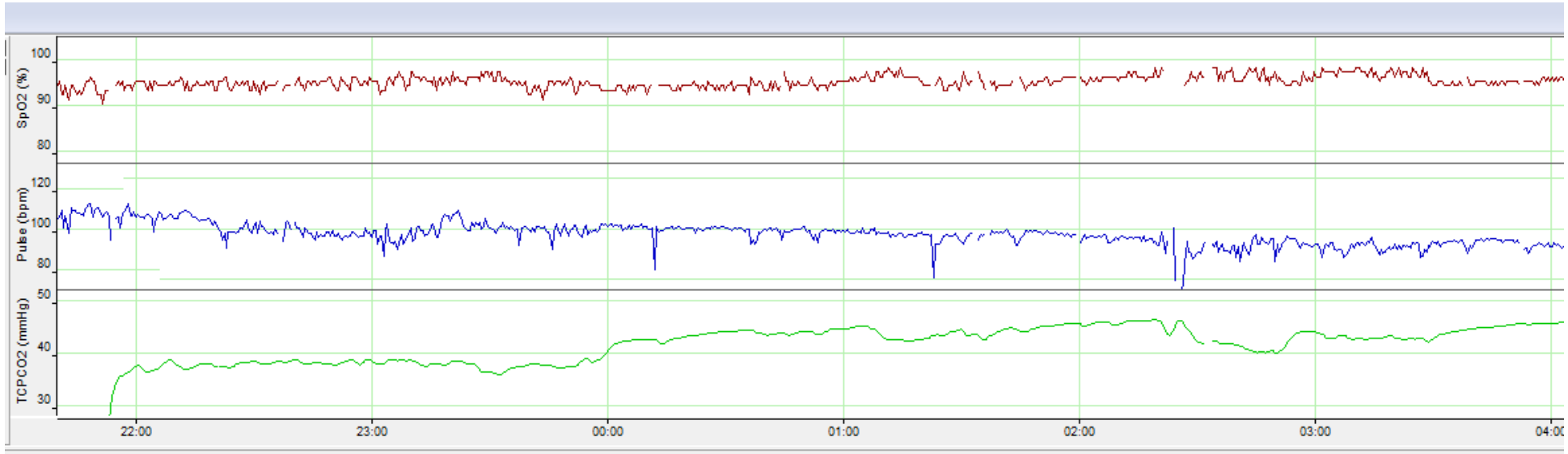
LB

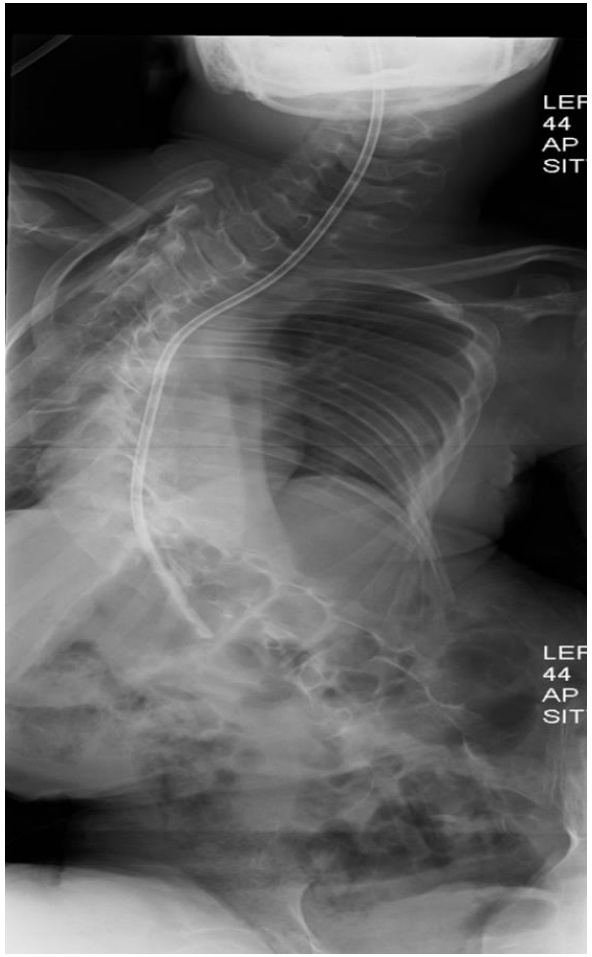
Sleep study on nocturnal BiPAP

- 

- $TcCO_2$ normalised
- BiPAP obliterated obstructive episodes

SM

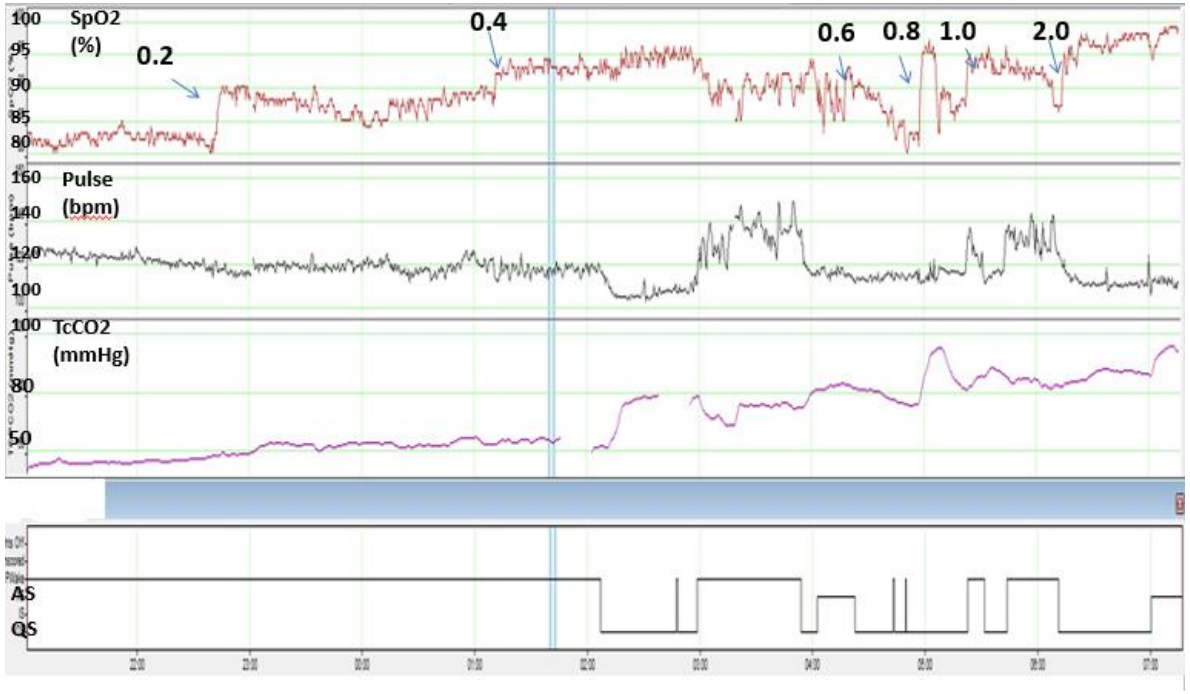
- 11 yr old girl
  - Cerebral palsy (GMFCS level V)
  - Severe scoliosis
  - Seizure disorder (daily seizures)
  - Very severe neurodisability
  - Gastrostomy fed
- 3 PICU admissions (May 2016 LRTI)
- Only rarely needed oral antibiotics for chest infections
- Secretion management
- PEG fed
- Occasional vomits
- Referred for spinal pre-op assessment including sleep study



SM

Baseline sleep study result:

- Increased WOB
- Moderate obstructive hypoventilation
- AHI 8.6evs/hr
- Worsening of hypoventilation in oxygen
- In air: mean SpO2 83-84%, TcCO2 7.4kPA
- in 1.2L O2 mean SpO2 90%,TcCO2 12.5kPa



What would be your management plan?

1. Give antibiotics for presumed respiratory infection
2. Additional investigations such as chest imaging
3. Consider non-invasive ventilation
4. Alternative management method such as nasopharyngeal airway
5. All of the above

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SM

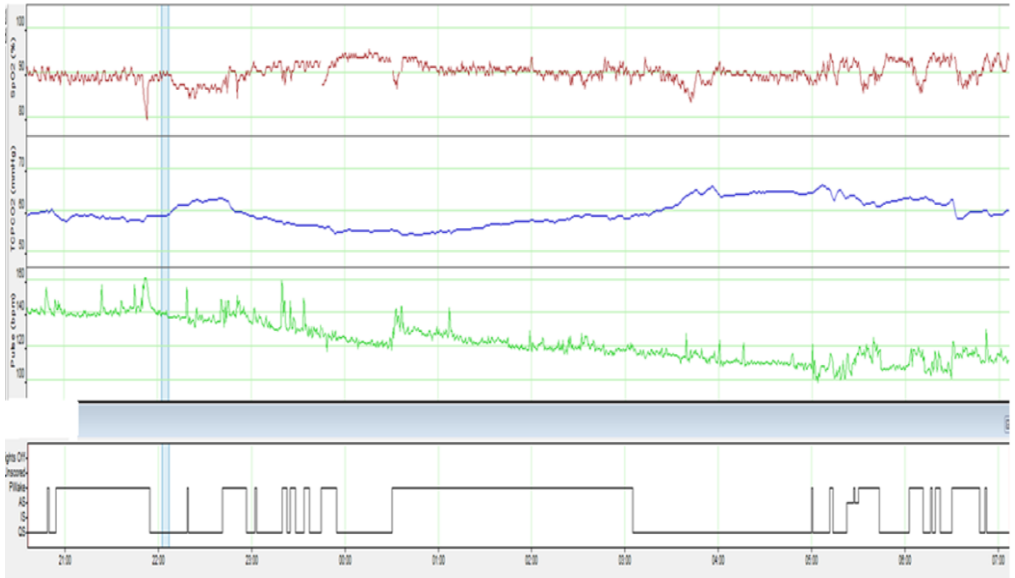
Acute or chronic?

Antibiotics given for presumed respiratory infections
No acute changes on chest x-ray
Nasopharyngeal prong unsuccessful
Pros and Cons of NIV discussed
Decision for BiPAP trial

SM

BIPAP titration study:

- via Wisp (L) nasal mask
- Pressure 12/6 (16) to 16/8 (16)
- AHI 3.8 evs/hr (OAHI 0, UnAHI 3.8)
- SpO2 91% to nadir 83%
- Mean $TcCO_2$ 7.8kPA



Mother completed training to use NIV ventilator
Plan for discharge and follow up

< 24 hours of discharge:
Found dead in her bed at home first night after discharge

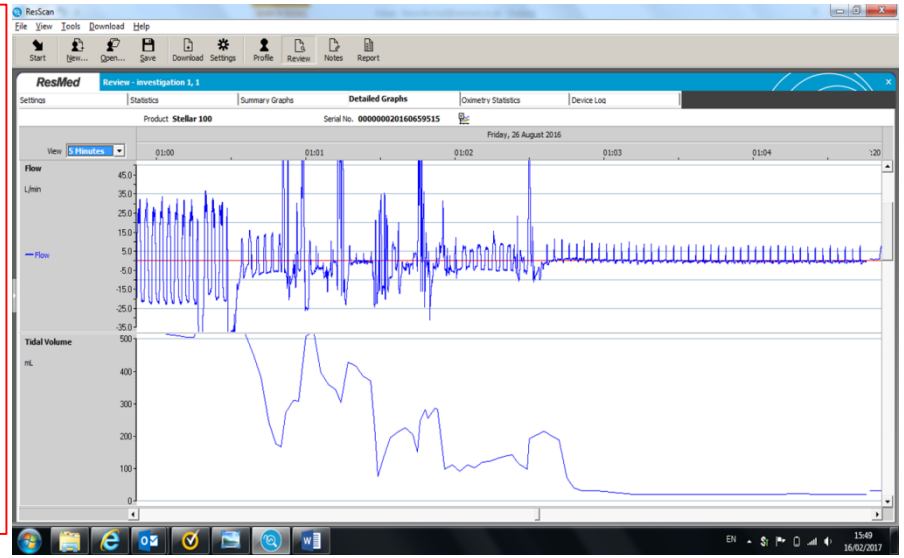
SM

Inquest

- PM inconclusive
- Ventilator download

Cause of death:

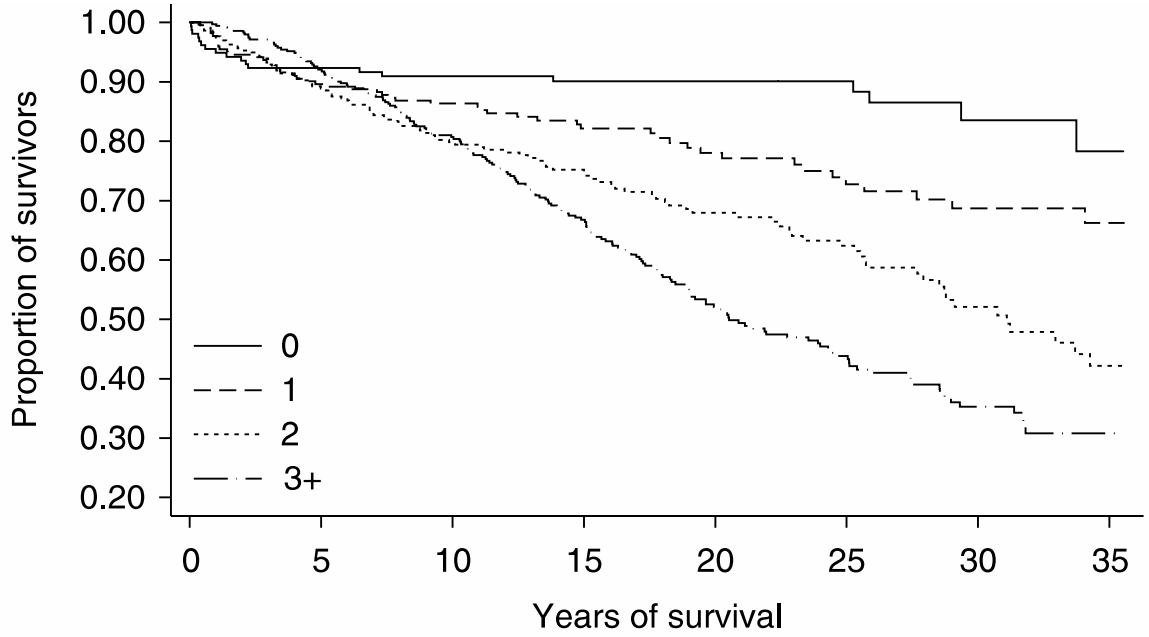
1. Epilepsy
2. Cerebral palsy



Lessons learnt

- Parental perception:
  - Parents perceived NIV being the cause of death
- In particular, in severe CP with multiple comorbidities:
  - Safety issue
  - Risk in altering previously adapted chronic hypoventilation
  - ?Goal post for effectiveness

Survival of patients with CP (GMFSC IV-V)



Number at risk

0	156	144	121	96	68	53	27	12
1	222	199	164	123	89	61	44	23
2	297	264	199	145	100	70	40	16
3+	352	324	267	186	118	77	43	18

Reid et al Develop Med Child Neuro 2013

Increased risk for dying during sleep?

- Single centre study (1993-2011)
- 177 patients who died during this period
- 19 were "discovered dead during sleep" at home
- Average age was 17 years and 6 months (range 6 – 25 years)

- Patient characteristics:
  - 19/19 had gastrostomy, epilepsy, non-ambulatory (GMFSC IV-V)
  - 11/ 19 had severe mental retardation
  - 19/ 19 had more than one respiratory co-morbidity
  - 14/19 had respiratory failure
  - 16/19 had nocturnal respiratory failure

Combination of aspiration risk, chronic lung disease, SDB and epilepsy that makes these patients vulnerable at night

Karatas AF J Pediatr Rehabil Med 2013 Jan ; 6:225-31

Thank you for you attention
Any questions?

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