Ondine’s curse and the anaesthetist.

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Background

- 4 year old girl
- CCHS, Marcus Gunn jaw winking syndrome

- Previous tracheostomy – decannulated Oct 2014.
- NIV overnight
- Sleep studies – periods of hypoventilation despite NIV – hypoxia and hypercarbia. Repeated chest infections.
Huh........?

- CCHS
- Inherited autosomal dominant
- 90% result from new mutation in the PHOX2B gene
What’s involved?

- **Breathing disorder** – hypoventilation unresponsive to hypoxia and hypercarbia
- **Autonomic disorder**
- Hirschsprung disease
- Oesophageal dysmotility
- **Eye disorders**
- Low body temperature (occasional perfuse sweating)
- Epilepsy
- Short, wide and flattened faces
Marcus Gunn Jaw-winking syndrome
Previous Anaesthetic History

- Examination of airway
  - Tracheostomy insitu – hypertensive and tachycardic
  - Commenced on atenolol for SBP > 220mmHg

- Failed fistula closure
  - Cardiac arrest at induction.
Plan – attempt 1

- Gas induction
- 22g cannula
- Intubate on Alfentanil (10mcg/kg)
- Arterial line
- Aim to extubate awake – PICU overnight
No notes
No echo
No PICU bed
Help requested in anaesthetic room - none available at that time
Patient travelled from distance.
What actually happened

- Resting heart rate 84-90 bpm
- **Gas induction** – O₂ + Sevo 5 %
- IV cannulation
- **Alfentanil** (10mcg/kg)
- Laryngoscopy Grade 1, **cords closed** but HR stable
- **Deepened** slightly and additional alfentanil bolus – sudden drop in HR to 50bpm.
- Reached for atropine – 10mcg/kg.

**BUT.....Too late – HR 20 bpm – CPR!**
Then what!? 

- HR increased to >90bpm in less than 45 seconds – no desaturation (according to monitor)

- Procedure abandoned

- NIV commenced in recovery

- One hour later – eating lunch on ward!
Learning points

Clinical
- Pre med with atropine
- Give atropine sooner
- Role of atenolol
- Spray cords vs alfentanil
- Postpone until adequate help available & notes
- Impact factors
  - Parents
  - Start times
  - Not familiar with trust

Operational
- Pre Op Assessment
- Rota manipulation
- PICU bed booked in advance
Plan - attempt 2

- Literature review
- Adequate support
- Investigations
- MDT approach
Case report

Heart block following propofol in a child

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Anesthetic Management of a Child With Congenital Central Hypoventilation Syndrome (CCHS, Ondine’s Curse) for Dental Treatment

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# Literature

## TABLE 2—Characteristics of Arrhythmias During Baseline Holter Recording in 14 Congenital Central Hypoventilation Syndrome Subjects and 17 Controls

<table>
<thead>
<tr>
<th>Arrhythmia Type</th>
<th>CCHS (322.9 hr recorded)</th>
<th>Controls (386.4 hr recorded)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Events</td>
<td>Events/hr</td>
</tr>
<tr>
<td>Sinus bradycardia</td>
<td>335</td>
<td>15.0</td>
</tr>
<tr>
<td>Sinus tachycardia</td>
<td>0</td>
<td>0.0</td>
</tr>
<tr>
<td>Sinus pauses</td>
<td>65</td>
<td>2.8</td>
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<tr>
<td>Premature atrial beats</td>
<td>364</td>
<td>15.5</td>
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<tr>
<td>Atrial couplets</td>
<td>0</td>
<td>0.0</td>
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<tr>
<td>Ectopic atrial rhythm</td>
<td>10</td>
<td>0.4</td>
</tr>
<tr>
<td>Accelerated junctional rhythm</td>
<td>0</td>
<td>0.0</td>
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<tr>
<td>Junctional rhythm</td>
<td>0</td>
<td>0.0</td>
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<tr>
<td>Ventricular premature beat</td>
<td>412</td>
<td>16.6</td>
</tr>
<tr>
<td>Ventricular bigeminy</td>
<td>5</td>
<td>0.2</td>
</tr>
<tr>
<td>Total</td>
<td>1,191</td>
<td>50.5</td>
</tr>
</tbody>
</table>
Literature

Central Hypoventilation Syndrome

Patient and Carer Information Booklet

1st Edition 2012
Not much available.

Try to avoid as many drugs as possible!
- Premeds
- Opioids
- NMB
- IV anaesthetic agents, esp. propofol (arrhythmias)
- N₂0

Gas induction with sevoflurane preferred

(low blood solubility is useful for rapid establishment of anaesthetic concentration and lack of airway irritant)
Literature

- Intubate
- Use regional where you can
- HDU / ICU back up
Plan – attempt 2

- Wean atenolol
- **Avoid** sedatives, opioids, muscle relaxation and propofol.
- Two anaesthetists for case and reduced theatre work load
- **Gas induction** with $O_2 + N_2O + Sevo$ 8%.
- **Lidocaine** to VC via MAD – intubate
- **Extubate awake** in recovery with non-invasive ventilation on standby.

- I.V Paracetamol and LA infiltration by surgeons
- Post op PICU bed available for monitoring.
What happened?

- Completion of surgery without cardiac arrest.
- Relatively hypertensive intraoperatively.
- Brief desaturation in recovery......
- Uneventful PICU stay but atenolol was eventually restarted.
Take home

- CCHS
- Trainee allocation
- MDT approach
- List booking and workload
- Dealing with poor outcomes
- Courage to cancel
- Requesting help
References


Sochala C, Van Deenen D, De Ville A, Govaerts MJM. Heart block following propofol in a child.

Thank you