Critical Illness
Neuromyopathy

Dr Alan Race
BSc (Hons) PhD FRCA
Objectives

1. What examiners say
2. Definitions
3. Aetiology
4. Risk Factors
5. Clinical Features
6. Prevention
Critical Illness Weakness
Pass Rate 30.4%, 46.6% of candidates received a poor fail

• Anticipated to be difficult
• Topical and an important consideration in the management of critically ill patients.
• Most no idea that the definition excluded pre-existing pathology, and that the weakness was symmetrical with cranial nerves sparing.
• Few candidates had knowledge of the use of nerve conduction studies and even fewer mentioned the MRC scale of scoring muscle power. The importance of preparing detailed notes on mandatory units of training when revising for the Final FRCA is exemplified by this question.
Definition

- Weakness in the critically ill patient *may be due to pre existing neuromuscular disorders*, a previously undiagnosed neuromuscular disorder, or as a complication of the critical illness.

- Critical illness neuromyopathy is a diagnosis of EXCLUSION.

- Under recognised (25 – 80% of patients).

- > 90 day stay, > 90% show EMG evidence of neuromuscular pathology 5 years later.
Definition

- Critical Illness Polyneuropathy (CIP)
- Critical Illness Myopathy (CIM)
- Critical Illness Neuromyopathy (CINM)
Aetiology

- Mechanism poorly understood
  - Microvascular damage
  - Direct neurotoxicity
  - Cytokine-mediated injury
Risk factors

- Sepsis
- Corticosteroids
- Neuromuscular blocking agents
- Hyperglycaemia
- Electrolyte derangements (K+, PO4- and Mg++)
- Immobility
# Risk Factors for CIP, CIM & CINM

<table>
<thead>
<tr>
<th>PROBABLE</th>
<th>POSSIBLE</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Severe sepsis/septic shock</td>
<td>• Age</td>
</tr>
<tr>
<td>• Multiorgan failure</td>
<td>• Female gender</td>
</tr>
<tr>
<td>• Prolonged mechanical ventilation/bed rest</td>
<td>• Severity of illness on admission</td>
</tr>
<tr>
<td>• Increasing duration of SIRS</td>
<td>• Admission APACHE II score</td>
</tr>
<tr>
<td>• Increasing duration of multiorgan failure</td>
<td>• Hypoalbuminaemia</td>
</tr>
<tr>
<td>• Hyperglycaemia</td>
<td>• Hyperosmolality</td>
</tr>
<tr>
<td></td>
<td>• Parenteral nutrition</td>
</tr>
<tr>
<td></td>
<td>• Renal replacement therapy</td>
</tr>
<tr>
<td></td>
<td>• Vasopressors</td>
</tr>
<tr>
<td></td>
<td>• Corticosteroids</td>
</tr>
<tr>
<td></td>
<td>• Neuromuscular blocking agents</td>
</tr>
<tr>
<td></td>
<td>• Aminoglycosides</td>
</tr>
</tbody>
</table>
Clinical features

• Diagnosis is DIFFICULT

• Suspect if
  • Unexplained weakness
  • Difficult wean – common

• Variable presentation
  • Motor deficit – symmetric: mild to quadriplegia
  • Face sparing – cranial nerve sparing
  • Reduced or absent reflexes
  • ~ 50% show sensory loss
<table>
<thead>
<tr>
<th>MRC scale</th>
<th>Simplified MRC scale</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 complete paralysis</td>
<td>0 complete paralysis</td>
</tr>
<tr>
<td>1 minimal contraction</td>
<td>1 severe weakness (&gt; 50% loss of strength)</td>
</tr>
<tr>
<td>2 active movement with</td>
<td>2 slight weakness (&lt; 50% loss of strength)</td>
</tr>
<tr>
<td>3 weak contraction</td>
<td></td>
</tr>
<tr>
<td>4 active movement</td>
<td></td>
</tr>
<tr>
<td>5 normal strength</td>
<td>3 normal strength</td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Clinical features

• Investigations – exclude, exclude, exclude...
  • Brain and spinal cord imaging – panning for central causes
  • EMG useful
    • Alternative diagnosis, or
    • Typical pattern – reduced action potential with normal conduction velocity
  • Muscle biopsy is not routinely performed
    • Only those with diagnostic uncertainty
    • Reduced actin/myosin ratio in CIMN
Compound Muscle Action Potentials, CMAP
<table>
<thead>
<tr>
<th>INVESTIGATION</th>
<th>CIP</th>
<th>CIM</th>
<th>CINM</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>EMG</strong></td>
<td>Spontaneous fibrillation potentials and sharp waves; + long duration, high-amplitude polyphasic MUPs (reinnervation)</td>
<td>Spontaneous fibrillation potentials and sharp waves; short duration, low-amplitude MUPs with early recruitment</td>
<td>Features of both CIP and CIM</td>
</tr>
<tr>
<td><strong>DIRECT MUSCLE STIMULATION</strong></td>
<td>Nerve: muscle ratio $&lt;0.5$; Normal direct muscle CMAP amplitude</td>
<td>Nerve: muscle ratio $&gt;0.5$; Reduced direct muscle CMAP amplitude</td>
<td>Variable depending on the relative components of CIP and CIM</td>
</tr>
</tbody>
</table>

Motor Unit Potential, MUP

Compound Muscle Action Potentials, CMAP
## INVESTIGATIONS IN CIP, CIM & CINM

<table>
<thead>
<tr>
<th>INVESTIGATION</th>
<th>CIP</th>
<th>CIM</th>
<th>CINM</th>
</tr>
</thead>
<tbody>
<tr>
<td>CPK</td>
<td>Normal or mildly elevated</td>
<td>Elevated in majority</td>
<td>Normal or elevated</td>
</tr>
<tr>
<td>CSF</td>
<td>Normal cell counts, Normal or slightly elevated protein (&lt;0.8g/L)</td>
<td>Normal</td>
<td>Normal or slightly elevated protein (&lt;0.8g/L)</td>
</tr>
<tr>
<td>NERVE CONDUCTION STUDIES</td>
<td>Reduced CMAP amplitudes; Reduced SNAP amplitudes; Normal conduction velocities and Latencies</td>
<td>Reduced CMAP amplitudes; Normal SNAP amplitudes; Normal conduction velocities and latencies</td>
<td>Reduced CMAP amplitudes; Reduced SNAP amplitudes; Normal conduction velocities and latencies</td>
</tr>
</tbody>
</table>

Sensory Nerve Action Potentials, SNAPS
Prevention

• No specific treatment exists
• Avoid risk factors...
• Most require prolonged respiratory wean
  • Complete functional recovery in ~ 70%
• Most some disability
  • Mild sensory impairment to complete function dependence
Prevention

- Treat sepsis aggressively
- Tight glucose control
- Avoid steroids
- Avoid NMB agents (Steroids > benzylisoquinolinium)
- Min sedation
- Physiotherapy
- Nutrition
Objectives

1. What examiners say
2. Definitions
3. Aetiology
4. Risk Factors
5. Clinical Features
6. Prevention