Nuances of EMG interpretation in pediatrics

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Objectives

• Discuss:
  • Normal maturation of peripheral nerves & effect upon NCS
  • Repetitive nerve stimulation & SSFEMG
  • Normal maturation of muscle fibers & effect upon EMG
Peripheral nerve maturation

• Myelination begins as early as 15 weeks GA\textsuperscript{1}

• Diameter of axons & surrounding myelin sheaths gradually increase from birth until adult values attained at around 4-5 years of age\textsuperscript{2}

\textsuperscript{1}Gamble & Breathnach. J Anat 1965; 99: 573-84
\textsuperscript{2}Shroder. Pathology of Peripheral Nerves. 2001; 1-14
Nerve biopsies (toluidine blue staining, resin embedded) showing normal progression of peripheral nerve myelination with age.

A. **17-week old fetus** (normal lumbar plexus) shows essentially no myelination of large fiber axons;

B. **10-mos old infant** (normal sural nerve) shows thin myelin; and

C. **10-year-old child** (normal sural nerve) shows adult thickness of myelin.

Biopsy A was performed at the time of post-mortem study in a spontaneous delivery. Biopsies B and C were surgical specimen in patients with demonstrated either a muscular disease or a non-neurological systemic disease.

Photo credit: Dr. Jean Michaud, Department of Pathology, Children's Hospital of Eastern Ontario
# Peripheral nerve maturation

<table>
<thead>
<tr>
<th>Age</th>
<th>Ulnar nerve conduction velocity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Premature, 25 wks gestational age</td>
<td>&lt; 15 m/sec&lt;sup&gt;1&lt;/sup&gt;</td>
</tr>
<tr>
<td>Term newborn, 40 wks GA</td>
<td>20-36 m/sec&lt;sup&gt;2&lt;/sup&gt;</td>
</tr>
<tr>
<td>Adult, healthy</td>
<td>&gt;58 m/sec&lt;sup&gt;3&lt;/sup&gt;</td>
</tr>
</tbody>
</table>

<sup>1</sup>Lori S et al. Childs Nervous Syst. 2018: 34:1145-52
Peripheral nerve maturation
AANEM Consensus Statement
Electromyography in Pediatrics

• To be published soon in Muscle & Nerve

• AANEM’s Normative Data Taskforce criteria applied

• Good pediatric normative data

• Seven articles – published original, normative data on >100 patients (past 35 yrs)
  (eighth included, N=92)

<table>
<thead>
<tr>
<th>Study</th>
<th>Cohort</th>
<th>Sensory nerves</th>
<th>Motor nerves</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ryan et al, 2019&lt;sup&gt;1&lt;/sup&gt;</td>
<td>1,918 data sets 1,849 children (0-18 yo)</td>
<td>Median, ulnar, radial, LAC, MAC, sural, saphenous, peroneal, lateral &amp; medial plantar</td>
<td>Median, ulnar, radial, spinal accessory, femoral, peroneal, tibial</td>
</tr>
<tr>
<td>Wang et al, 2014&lt;sup&gt;2&lt;/sup&gt;</td>
<td>163 children (0-14 yo)</td>
<td>Saphenous</td>
<td>Femoral</td>
</tr>
<tr>
<td>Garcia et al, 2000&lt;sup&gt;3&lt;/sup&gt;</td>
<td>92 children (0-6 yo)</td>
<td>Median, medial plantar</td>
<td>Median, ulnar, peroneal, tibial</td>
</tr>
<tr>
<td>Smit et al, 1999&lt;sup&gt;4&lt;/sup&gt;</td>
<td>200 preterm infants (25-30 wks GA)</td>
<td>N/a</td>
<td>Ulnar, tibial</td>
</tr>
<tr>
<td>Cai et al, 1997&lt;sup&gt;5&lt;/sup&gt;</td>
<td>168 children &amp; adults (0-30 yo)</td>
<td>Median, ulnar, sural</td>
<td>Medial, ulnar, peroneal, tibial</td>
</tr>
<tr>
<td>Hyllienmark et al, 1995&lt;sup&gt;6&lt;/sup&gt;</td>
<td>128 children, young adults (0-20 yo)</td>
<td>Median, peroneal, sural</td>
<td>Median, peroneal</td>
</tr>
<tr>
<td>Parano et al, 1993&lt;sup&gt;7&lt;/sup&gt;</td>
<td>155 children (0-14 yo)</td>
<td>Median, sural</td>
<td>Median, peroneal</td>
</tr>
<tr>
<td>Lang et al, 1985&lt;sup&gt;8&lt;/sup&gt;</td>
<td>129 children (3-19 yo)</td>
<td>Radial, peroneal, sural</td>
<td>Median, peroneal</td>
</tr>
</tbody>
</table>

<sup>2</sup>Wang et al, Ped Neurol 2014: 50: 149-157  
<sup>7</sup>Parano E. et al. J Child Neurol. 1993: 8: 336-338  
<sup>8</sup>Lang HA et al. Muscle Nerve 1985: 8: 38-43
Approach to Pediatric Neuropathy

Is the neuropathy the only feature of the disease?
- no
  - Complex inherited neuropathy syndrome
  - Ataxia
  - Spasticity
  - Global neurodevelopmental impairment
- yes
  - CMT and related disorders
  - Other

What is the predominant phenotype?

<table>
<thead>
<tr>
<th>~30</th>
<th>~30</th>
<th>~30</th>
<th>&gt;100</th>
</tr>
</thead>
<tbody>
<tr>
<td>Friedreich</td>
<td>SPG</td>
<td>Giant axonal neuropathy</td>
<td>Eye (OPA1, NARP)</td>
</tr>
<tr>
<td>SCA</td>
<td>XL-ALD</td>
<td>Cockayne</td>
<td>Cranial nerve (BVVL)</td>
</tr>
<tr>
<td>ARSACS</td>
<td></td>
<td>Peroxisomal</td>
<td>Renal (Fabry)</td>
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<td></td>
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<td></td>
<td>Hepatic (tyrosinemia)</td>
</tr>
</tbody>
</table>

Approach to Pediatric Neuropathy

>100 genes
Charcot-Marie-Tooth disease + hereditary motor neuropathy + hereditary sensory autonomic neuropathy

Approach to Pediatric Neuropathy

- CMT
- Age artifact? (<5 yo)

Uniform slowing

- GBS
- CIDP
- Diphtheria
- Glue sniffing
- Krabbe disease
- Metachromatic leukodystrophy
- Cockayne syndrome
- Adrenoleukodystrophy
- HNPP
Approach to Pediatric Neuropathy

- CMT
- Age artifact? (<5 yo)

Uniform slowing
- GBS
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Diphtheria

- *Corynebacterium diphtheria* toxin
- Acute pharyngitis (grey pseudomembrane)
- ~15% develop multiple cranial neuropathies
- May develop sensorimotor polyneuropathy (NCS may mimic Guillain-Barré syndrome)

Solvent abuse

- 11 yo healthy boy with progressive weakness (x 3 months)
- Distal > proximal weakness & sensory loss.
- NCS/EMG – sensorimotor polyneuropathy demyelinating features
  - Sensory responses – absent to bilateral median, ulnar, sural
  - Motor studies – abnormal to bilateral median, ulnar, peroneal, tibial
    - Conduction velocity < 50% lower limit normal
    - Conduction block in right median & right ulnar nerve (>50% amplitude drop between wrist & below elbow)
  - Absent F-responses – right median, ulnar, tibial
- CSF WBC normal; CSF protein normal
- MRI brain & spine unrevealing (no nerve root enhancement)
Solvent abuse
Infantile botulism

- *Clostridium botulinum* ubiquitous
- Spores germinate in immature infant gut → grow → release toxin
- ~98% of affected infants present between 1 and 6 mos old
- Infant botulism reported as early as the 1st week of life & as late as 12 mos old
- Toxin impairs acetylcholine release
- Symptoms: hypotonia, weakness, constipation, sluggish pupils, respiratory failure

Infantile botulism

- Repetitive nerve stimulation:

<table>
<thead>
<tr>
<th>RNS results:</th>
<th>N=25</th>
</tr>
</thead>
<tbody>
<tr>
<td>High rates (20-50 Hz)</td>
<td></td>
</tr>
<tr>
<td>Increment</td>
<td>23/25, 92%</td>
</tr>
<tr>
<td>Decrement</td>
<td>1/25, 4%</td>
</tr>
<tr>
<td>No change</td>
<td>1/25, 4%</td>
</tr>
<tr>
<td>Low rates (2-5Hz)</td>
<td></td>
</tr>
<tr>
<td>Increment</td>
<td>5/25, 20%</td>
</tr>
<tr>
<td>Decrement</td>
<td>14/25, 56%</td>
</tr>
<tr>
<td>No change</td>
<td>6/25, 24%</td>
</tr>
</tbody>
</table>

Infantile botulism

- Repetitive nerve stimulation left median nerve (APB):
  - At 5 Hz decrement, 8%
  - At 10Hz increment, 25%
  - At 20 Hz increment, 38%
  - At 50 Hz increment, 94%

Stimulated single fiber EMG

- Increasingly used NMJ disorders
- Eg. congenital myasthenic syndrome
Electromyography

• In children:
  • Study clinically, most important muscles first
  • Study “withdrawal muscles” (tibialis anterior, iliopsoas) for active recruitment & their antagonists (gastrocnemius, quadriceps) for spontaneous denervation

• In infants:
  • Developmental considerations for predicting recovery post-injury
Electromyography

- Infant (at 3 months old)
  - Diameter of muscle fiber = 17 µm³
  - More than 3-fold smaller than adult muscle fiber diameter

- Due to smaller cross-sectional area,
  infant muscle contains 11x motor units compared to adult

- Infant EMG can give "overly optimistic" impression of axonal continuity
EMG in root avulsion:
• Normal sensory studies
• Absent motor studies (Absent CMAPs)
• Profuse fibrillation potentials
• Few / absent MUAPs
• No improvement on f/u EMG (no nascent units)

MRI in root avulsion
• Pseudomeningocoel
• Nerve rootlets not seen

EMG provides useful information for management of these cases

Severe neonatal brachial plexus palsy
Severe neonatal brachial plexus palsy

MRI – no obvious abnormality?

MRI – thinning of left C5 & C6 nerve roots just proximal to upper trunk

Role of EMG is less clear in management / prognostication of these cases
Acute flaccid paralysis

- EMG – critical role localizing to motor neuron
- In 2018 – 285 cases of polio-like AFP reported (236 in USA; 49 in Canada)
- Cases annually, peaks in 2018, 2016, 2014

Other motor neuron disorders

- 16 yo, healthy male
- Progressive, painless weakness of left hand (x 12 months)
- No sensory symptoms
- Exam: Muscle wasting: left hypothenar & thenar eminence, FDI

<table>
<thead>
<tr>
<th></th>
<th>Deltoid</th>
<th>Infraspin</th>
<th>Biceps</th>
<th>Triceps</th>
<th>WE</th>
<th>WF</th>
<th>EDC</th>
<th>EIP</th>
<th>EPB</th>
<th>FPL</th>
<th>APB</th>
<th>FDI</th>
<th>ADM</th>
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<tbody>
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<td>Right</td>
<td>5</td>
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<td>4+</td>
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<tr>
<th></th>
<th>Illosoas</th>
<th>Quad</th>
<th>Hip Add</th>
<th>Ham</th>
<th>FA</th>
<th>Gast</th>
<th>TP</th>
<th>PL</th>
<th>EHL</th>
<th>Toeflex</th>
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<tbody>
<tr>
<td>Right</td>
<td>5</td>
<td>5</td>
<td>5</td>
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</table>
## Other motor neuron disorders

**Nerve conduction studies**

<table>
<thead>
<tr>
<th>Sensory</th>
<th>Normal</th>
<th>Right</th>
<th>Left</th>
<th>Normal</th>
<th>Right</th>
<th>Left</th>
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<tbody>
<tr>
<td>Median</td>
<td></td>
<td></td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td>PL (ms)</td>
<td>&lt;3.2</td>
<td>2.6</td>
<td>2.1</td>
<td>&lt;3.2</td>
<td>3.7</td>
<td>3.9</td>
</tr>
<tr>
<td>SNAP (mV)</td>
<td>&gt;14.0</td>
<td>29.0</td>
<td>35.8</td>
<td>&gt;14.0</td>
<td>12.4</td>
<td>7.9</td>
</tr>
<tr>
<td>Ulnar</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CV (m/s)</td>
<td>&gt;50</td>
<td>56</td>
<td>55</td>
<td>&gt;50</td>
<td>11.5</td>
<td>5.4</td>
</tr>
<tr>
<td>PL (ms)</td>
<td>&lt;3.3</td>
<td>2.0</td>
<td></td>
<td>&lt;3.4</td>
<td>2.5</td>
<td>3.0</td>
</tr>
<tr>
<td>SNAP (mV)</td>
<td>&gt;9.0</td>
<td>25.1</td>
<td></td>
<td>&gt;5.9</td>
<td>61</td>
<td>59</td>
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<tr>
<td>Dors Ulnar Cut</td>
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<td></td>
</tr>
<tr>
<td>PL (ms)</td>
<td>&lt;3.3</td>
<td>1.3</td>
<td></td>
<td>&gt;50</td>
<td>4.4</td>
<td>4.3</td>
</tr>
<tr>
<td>SNAP (mV)</td>
<td>&gt;4.0</td>
<td>16.1</td>
<td></td>
<td>&gt;50</td>
<td>5.4</td>
<td>3.2</td>
</tr>
<tr>
<td>Radial</td>
<td></td>
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<td></td>
<td></td>
<td></td>
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<tr>
<td>DML (ms)</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>PL (ms)</td>
<td>&lt;2.9</td>
<td>2.2</td>
<td></td>
<td>&gt;50</td>
<td>63</td>
<td>50</td>
</tr>
<tr>
<td>SNAP (mV)</td>
<td>&gt;11.0</td>
<td>38.7</td>
<td></td>
<td>&gt;50</td>
<td>5.4</td>
<td>3.2</td>
</tr>
</tbody>
</table>

| MAC     |        |       |      |        |       |      |
| PL (ms) | <3.3   | 1.3   | 1.7  | <3.3   | 3.7   | 3.9  |
| SNAP (mV)| >4.0  | 13.1  | 8.5  | >4.0   | 11.5  | 5.4  |

**Concentric needle EMG**

<table>
<thead>
<tr>
<th>Side</th>
<th>Muscle</th>
<th>Root</th>
<th>Insert</th>
<th>Fibr</th>
<th>PSW</th>
<th>Amp</th>
<th>Dur</th>
<th>Poly</th>
<th>Recruit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left</td>
<td>First dorsal interosseous</td>
<td>C8-T1</td>
<td>Incr</td>
<td>2+</td>
<td>Nml</td>
<td>Incr</td>
<td>Incr</td>
<td>Nml</td>
<td>2- to 3-</td>
</tr>
<tr>
<td>Left</td>
<td>Flexor carpi radialis</td>
<td>C6-C7</td>
<td>Incr</td>
<td>Nml</td>
<td>1+</td>
<td>Incr</td>
<td>Incr</td>
<td>Incr</td>
<td>1-</td>
</tr>
<tr>
<td>Left</td>
<td>Flexor pollicis longus</td>
<td>C7-C8</td>
<td>Incr</td>
<td>Nml</td>
<td>3+</td>
<td>Incr</td>
<td>Nml</td>
<td>Nml</td>
<td>1-</td>
</tr>
<tr>
<td>Left</td>
<td>Extensor digit commun</td>
<td>C7-C8</td>
<td>Incr</td>
<td>1+</td>
<td>1+</td>
<td>Incr</td>
<td>Nml</td>
<td>Nml</td>
<td>2- to 3-</td>
</tr>
<tr>
<td>Left</td>
<td>Biceps</td>
<td>C5-C6</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
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<tr>
<td>Left</td>
<td>Triceps</td>
<td>C6-C8</td>
<td>Nml</td>
<td>Nml</td>
<td>Incr</td>
<td>1+</td>
<td>Incr</td>
<td>Nml</td>
<td>1-</td>
</tr>
<tr>
<td>Right</td>
<td>First dorsal interosseous</td>
<td>C8-T1</td>
<td>Nml</td>
<td>1+</td>
<td>Nml</td>
<td>Incr</td>
<td>Incr</td>
<td>Nml</td>
<td>1-</td>
</tr>
<tr>
<td>Right</td>
<td>Abductor pollicis brevis</td>
<td>C8-T1</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
<td>Incr</td>
<td>Incr</td>
<td>Nml</td>
<td>1+</td>
</tr>
<tr>
<td>Right</td>
<td>Extensor digit commun</td>
<td>C7-C8</td>
<td>Nml</td>
<td>Nml</td>
<td>Incr</td>
<td>Incr</td>
<td>Incr</td>
<td>Nml</td>
<td>1-</td>
</tr>
<tr>
<td>Left</td>
<td>Vastus lateralis</td>
<td>L2-L4</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
</tr>
<tr>
<td>Left</td>
<td>Tibialis anterior</td>
<td>L4-L5</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
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Hirayama syndrome
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