Pediatric Aspects of EDX

Albert C. Clairmont, MD
Associate Professor-Clinical
The Ohio State University

February 25, 2013

Objectives

- Overview of Pediatric Electrodiagnosis (EDX)
- Understand the different approach to EDX in children compared to adults
- Be aware of potential pitfalls
- Review how nerve conduction study values vary with age
Reasons for referral

• Evaluate the floppy infant or child
• Hypotonia
• Hypertonia
• Generalized or segmental weakness
• Normal or abnormal at birth
• Delayed milestones
• Evaluate the floppy infant or child

Duchenne Muscular Dystrophy

Myotonic Dystrophy

Duchenne Muscular Dystrophy
How to approach the examination

• Diagnostic challenge
• Plan the examination
• History and physical exam
• Electrodiagnostic exam

Immediate pre-EMG instruction

Patient and family education materials

A WORD ABOUT PITFALLS

How to approach the examination
Some Pitfalls

• Loose or insecure electrodes
• Measurement inaccuracies
• Volume conduction related inaccuracies
• Temperature related problems
• Measurement inaccuracies
• Loose or insecure electrodes

Corrective Actions

• Avoid skin lotion (nemesis #1)
• Papoose board
• Self-adhesive electrodes
• Use tape generously
• Secure the electrodes
Pitfalls

- Measurement
- Eight or 9 cms. available
- One centimeter error = 12% error in CV
- Careful measurement
- Volume conduction
- Scrutinize waveforms
- Spread through volume conduction
- Check stimulus characteristics
- Volume conduction

Pitfalls
Temperature-related Pitfalls

• Normal values determined between 32-36°C
• 2 mm/s°C, axilla to finger
• Sensory conduction
• Temperature varies

Pitfalls

• Temperature/g39°C
• Sensory conduction
• 2 m/s°C, axilla to finger

Temperature-related Pitfalls

• Small thermocouples respond to changes faster
• Allow time for internal/external temperature equilibration
• Difficult to keep warm sometimes
• 37°C
• Surface temp of 37-38°C = near nerve temp of 36-
• Normal values determined between 32-36°C
Eric Denys. AAEM Minimonograph # 14: Influence of Temperature (revised 1991)

Pitfalls

- Distal motor latency
  - Temperature Δ

- Ulnar, median and peroneal
  - 35°C to 25°C
  - 0.2 m/s°C

- Motor conduction
  - Temperature Δ

- Cooling of peroneal motor
  - 23.5°C to 35°C
  - 1.8 m/s°C

Pitfalls
Pitfalls: Correction factors

- 2.4 m/s/°C, 29-38°C median & ulnar motor (Henriksson)
- 2.1 m/s/°C, ulnar motor
- 1.6 m/s/°C, ulnar sensory (Halar)
- 2.0 m/s/°C, peroneal motor (Halar)
- 1.5 m/s/°C, median motor (Edelwjn)

Corrective Actions

- Electrical shielding in ICU
- Do EMG in lab
- Keep baby in incubator
- Use warming lights
- Control temperature

Warming is probably better

- 1.5 m/s/°C, median motor (Edelwjn)
- 2.0 m/s/°C, peroneal motor (Halar)
- 1.6 m/s/°C, ulnar sensory (Halar)
- 2.1 m/s/°C, ulnar motor
- 2.4 m/s/°C, 29-38°C median & ulnar motor
Additional preparation (optional)

• Cooperation can be a significant problem
  • Helpful strategies include:
    – Sedation
    – Thoughtful sequencing of testing
  
Sedation

- Advantages and disadvantages
  • Facilitates nerve conduction studies
  • Assessment of spontaneous activity
  • Assessment of MUP more difficult
  • Assessment of MUP more difficult
Sedation

• All can benefit
• Ages 1 through 4 years old
• History of "injection behavior"
• Do not retain memory of procedure
• Interval examinations likely to be successful
• Monitor vital signs

- Onset about 10 to 20 minutes
- 0.2 mg/kg to 1 mg/kg
- 2 mg/cc solution
- Oral midazolam syrup (Versed)

Sedation

- Oral midazolam syrup (Versed)
- 2 mg/cc solution
- 0.2 mg/kg to 1 mg/kg
- Onset about 10 to 20 minutes
- Maximum sedation about 60 minutes
- Do not retain memory of procedure
- Monitor vital signs
Sedation

- Intranasal midazolam (Versed)
  - Dosage: 0.2 mg/kg divided between nares
  - 5 mg/cc solution
  - Use 1 cc syringe without needle
  - Onset: about 5 minutes
  - Maximum sedation: at 10 minutes

- Tylenol with codeine liquid + Ativan
  - 120 mg acetaminophen / 5 cc
  - 12 mg codeine / 5 cc
  - Codeine: 0.5 mg/kg to 1.0 mg/kg
  - Ativan (Lorazepam) syrup: 0.05 to 0.1 mg/kg
  - Give one hour prior to procedure

Monitor vital signs

Dosage: 0.2 mg/kg divided between nares

Intranasal midazolam (Versed)
• Sensory nerve conduction

Sequence of Testing

• Least noxious
• Useful starting point in hypotonias
• If normal, check for motor neuron, NM
• Junction, or muscle cell problems

Sequence of Testing

• Needle EMG
• Repetitive nerve stimulations
• Motor nerve conduction
• Sensory nerve conduction

Sequence of Testing
### Motor Nerve Conductions

- Needle cathode reduces artifact
- Motor NCV/gram in proportion to prematurity
- Newborn 50% CV of adults
- Adult values by age 4 or 5
- Newborn 50% CV of adults
- Motor NCV in proportion to prematurity
- Needle cathode reduces artifact

### Table: Ulnar Motor NCV in Infants & Children

<table>
<thead>
<tr>
<th>Number</th>
<th>Age (Years)</th>
<th>Age Span</th>
<th>NCV (m/s)</th>
<th>Prematurity</th>
<th>Full Term</th>
<th>Premature Infant</th>
<th>Premature Infants 21-30 days</th>
<th>Premature Infant 21-40 days</th>
<th>Newborn 21-33</th>
<th>Full Term 47-73</th>
<th>Premature 18-22</th>
</tr>
</thead>
</table>

| 42     | 21-33       |          |          | Full Term   |           |                 |                             |                             |                |                 |               |
| 6      | 18-22       |          |          | Premature Infant |        |                 |                             |                             |                |                 |               |

Thomas & Lambert, 1959
- Motor & sensory conduction velocity
- Corrected distal motor latency
- Surface electrodes
- Age = 7 weeks to 6 years
- N = 92 normal infants and children

Normal infants and children
Peripheral motor & sensory NCS in

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Std. Dev.</th>
<th>4.7 M/s</th>
<th>5.0 M/s</th>
<th>5.3 M/s</th>
<th>5.7 M/s</th>
<th>5.8 M/s</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tibial</td>
<td>± 9.4</td>
<td>± 9.3</td>
<td>± 9.2</td>
<td>± 9.6</td>
<td>± 9.0</td>
<td>± 8.2</td>
</tr>
<tr>
<td>Peroneal</td>
<td>± 9.3</td>
<td>± 9.3</td>
<td>± 9.6</td>
<td>± 9.0</td>
<td>± 8.2</td>
<td>± 7.7</td>
</tr>
<tr>
<td>Median</td>
<td>± 9.7</td>
<td>± 9.4</td>
<td>± 9.1</td>
<td>± 8.2</td>
<td>± 7.4</td>
<td>± 6.9</td>
</tr>
<tr>
<td>Ulnar</td>
<td>± 9.4</td>
<td>± 9.4</td>
<td>± 9.4</td>
<td>± 9.4</td>
<td>± 9.4</td>
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</tbody>
</table>

Mean motor NCV Children 4-16

Garcia et al. Clinical Neurophysiology, 2000

F-waves

<table>
<thead>
<tr>
<th>Age (months)</th>
<th>Median nerve (µV)</th>
<th>SD</th>
<th>Tibial nerve (µV)</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>48-72</td>
<td></td>
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</tr>
<tr>
<td>2.27 ± 0.83</td>
<td>6.99 ± 5.49</td>
<td></td>
<td>6.12 ± 1.53</td>
<td></td>
</tr>
<tr>
<td>3.81 ± 1.53</td>
<td>5.89 ± 2.80</td>
<td></td>
<td>2.4 ± 1.52</td>
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</tr>
<tr>
<td>3.63 ± 1.44</td>
<td>4.50 ± 1.15</td>
<td></td>
<td>6-12</td>
<td></td>
</tr>
<tr>
<td>2.70 ± 1.43</td>
<td>3.45 ± 0.00</td>
<td></td>
<td>4-6</td>
<td></td>
</tr>
<tr>
<td>3.13 ± 1.48</td>
<td>3.62 ± 1.06</td>
<td></td>
<td>1-6</td>
<td></td>
</tr>
<tr>
<td>1.71 ± 0.74</td>
<td>2.33 ± 0.86</td>
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<table>
<thead>
<tr>
<th>Age (months)</th>
<th>SNAP amplitude (µV): mean ± SD</th>
<th></th>
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</thead>
<tbody>
<tr>
<td></td>
<td>48-72</td>
<td></td>
</tr>
<tr>
<td>9.48 ± 2.39</td>
<td>5.96 ± 2.33</td>
<td></td>
</tr>
<tr>
<td>9.57 ± 3.54</td>
<td>4.72 ± 1.42</td>
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</tr>
<tr>
<td>9.07 ± 2.12</td>
<td>2.69 ± 1.32</td>
<td></td>
</tr>
<tr>
<td>6.83 ± 2.69</td>
<td>2.4 ± 1.63</td>
<td></td>
</tr>
<tr>
<td>6.16 ± 2.44</td>
<td>1.72 ± 0.62</td>
<td></td>
</tr>
<tr>
<td>4.40 ± 1.73</td>
<td>1.88 ± 0.74</td>
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<table>
<thead>
<tr>
<th>Age (months)</th>
<th>CMAP amplitude (µV): mean ± SD</th>
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<tbody>
<tr>
<td></td>
<td>48-72</td>
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</tr>
</tbody>
</table>
Sensory nerve conduction velocity studies

Garcia et al., Clinical Neurophysiology, 2000

Pediatric neurologist was standard
Clinical history, examination and review by
Identified and studied
Visits and final clinical diagnoses were
27 cases with muscle biopsies, labs, follow up
Years (1999-2005)
Retrospective study of 550 EMG/NCV over 5

Biopsy in Childhood
EMG Accuracy Compared to Muscle

Rabie et al., Journal of Child Neurology, July 2007
EMG Accuracy Compared to Muscle Biopsy in Childhood

Classifications:
- Myopathic
- Neuromuscular junction disorder
- Neurogenic
- Nonspecific
- Normal

Nerve conduction studies:
- 1 sensory + 1 motor in the upper limbs
- 1 sensory + 2 motor in lower limbs
- 2 proximal + 2 distal upper limb muscles
- 2 proximal + 2 distal lower limb muscles

Needle EMG:
- 2 proximal + 2 distal upper limb muscles
- 2 proximal + 2 distal lower limb muscles

Rabie et al; Journal of Child Neurology, July 2007
EMG Accuracy Compared to Muscle Biopsy in Childhood

- 5 congenital myopathies: 40% normal, 40% children < 2 years old
  - Improved detection rate 3/4 (75%) for myopathies when patient > 2 years old
  - Low EMG detection rate 1/7 (14%) for congenital myopathies, 40% normal, 40%.

EMG Accuracy Compared to Muscle Biopsy

- Low EMG detection rate (1/7) or 14% for myopathies when patient < 2 years old
  - Improved detection rate 3/4 (75%) for children > 2 years old
  - 11/11 myopathies: 26% (7/27) discordant with Muscle biopsy: 87% (20/23) concurred with EMG: 74% (20/27) concurred with final Dx.
  - Muscle biopsy: 87% (20/23) concurred with discordant EMG were myopathies 26% (7/27) discordant
  - 5 congenital myopathies: 40% normal, 40% normal, 40% normal, 40%.
5 Year old boy

Long H/O ataxia

Obligatory wheelchair user for a number of years

Long H/O ataxia

Work-up has included muscle biopsy, results unknown

C/O weakness of lower limbs x 1-2 weeks

Case

Muscle stretch reflexes absent

Nerve conduction studies: Absent sural & superficial peroneal sensory

Muscle stretch reflexes absent

Uncooperative with physical exam

Motor NCS: See next 2 slides

Needle EMG: ↓ #, ↓ duration, ↓ polys

No abnormal spontaneous activity recorded

Case
Chronic axonal sensory and motor polyneuropathy, with no evidence of acute change

SUMMARY

- Different approach in infants & children
- Can be technically difficult
- Sedation might be necessary
- Be aware of pitfalls
- ACCurate, detailed H & P essential
- EDX useful in infants & children

Case
Selected References


