Pediatric Aspects of EDX

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

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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

Myotonic Dystrophy

Duchenne Muscular Dystrophy

How to approach the examination

- Diagnostic challenge
- Plan the examination
- History and physical exam
- Electrodiagnostic exam
- Patient and family education materials
- Immediate pre-EMG instruction

A WORD ABOUT PITFALLS

Some Pitfalls

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

Corrective Actions

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

Pitfalls

- Measurement
- Careful measurement
- Eight or 9 cms. available
- One centimeter error = 12 % error in CV

Pitfalls

- Volume conduction
- Check stimulus characteristics
- Spread through volume conduction
- Scrutinize waveforms

Temperature-related Pitfalls

- Normal values determined between 32-36°C
- Surface temp of 37-38°C = near nerve temp of 36-37°C
- Difficult to keep warm sometimes
- Allow time for internal/external temperature equilibration
- Small thermocouples respond to changes faster

Eric Denys. AAEM Minimonograph # 14: Influence of Temperature (revised by Rutkove July 2001)

Pitfalls

- Temperature Δs
- Sensory conduction
- 2 m/s/°C, axilla to finger
- 20 °C to 36 °C

Pitfalls

- Temperature Δs
- Motor conduction
- Cooling of peroneal motor
- 23.5 °C to 35 °C
- 1.8 m/s/°C

Eric Denys. AAEM Minimonograph # 14: Influence of Temperature (revised 1991)

Pitfalls

- Temperature Δs
- Distal motor latency
- $\Uparrow$ 0.2 m/s/°C
- $\Downarrow$ 35 °C to 25 °C
- Ulnar, median and peroneal

Pitfalls: Correction factors

- 2.4 m/s/°C, 29-38°C median & ulnar motor (Henriksen)
- 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)
- Warming is probably better

Corrective Actions

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

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

Sedation

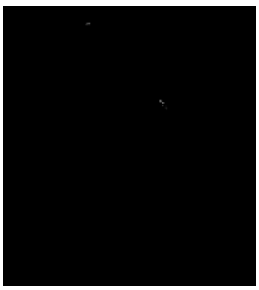
- All can benefit
- Ages 1 through 4 years old
- History of “injection behavior”
- ⇑ cooperation ⇓ anxiety
- Do not retain memory of procedure
- Interval examinations likely to be successful

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
- 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
- Monitor vital signs



Sedation

- Tylenol with codeine liquid + Ativan
- 120 mg acetaminophen / 5cc
- 12 mg codeine / 5cc
- 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

Sequence of Testing

- Sensory nerve conductions
- Motor nerve conductions
- Repetitive nerve stimulations
- Needle EMG

Sequence of Testing

- Sensory nerve conductions
- Least noxious
- Useful starting point in hypotonia
- If normal, check for motor neuron, NM junction, or muscle cell problems

Motor Nerve Conductions

- Needle cathode reduces artifact
- Motor NCV ⇓ in proportion to prematurity
- Newborn 50% CV of adults
- Adult values by age 4 or 5
- CMAP ⇑ from 3.7 mV to 10.5 mV ADM term to age 12

Ulnar motor NCV in Infants & Children

Number	Age	Age span	NCV (M/S)
6	Premature Infant	21- 40 days premature	18-22
42	Full Term	Newborn	21-33
98	Children	Up to 14 years old	47-73

Thomas & Lambert, 1959

Mean motor NCV Children 4-16 years and in Adults

Nerve	Age 4-16 years	Std. Dev	Adults	Std. Dev
Ulnar	58.2 M/s	± 9.7	55.1 M/s	± 6.4
Median	57.2 M/s	± 8.2	53.0 M/s	± 6.4
Peroneal	53.0 M/s	± 9.6	50.1 M/s	± 9.3
Tibial	47.9 M/s	± 9.2	50.2 M/s	± 9.3

N = 116. N = 80 < 4 y.o. N= 36 > 4 y.o. Baer & Johnson 1965

Peripheral motor & sensory NCS in normal infants and children

- N = 92 normal infants and children
- Age = 1 week to 6 years
- Surface electrodes
- Motor & sensory conduction velocity
- Corrected distal motor latency
- F-waves

Garcia et al. Clinical Neurophysiology, 2000

Age group, mean age and sex of subjects

Age group (months)	Mean age	Males	Females	Total
<1	18 days	6	6	12
1–6	3.5 months	7	8	15
6–12	9.5 months	12	2	14
12–24	20 months	9	6	15
24–48	34 months	16	2	18
48–72	59 months	13	5	18
Total		63	29	92

Garcia et al. Clinical Neurophysiology, 2000

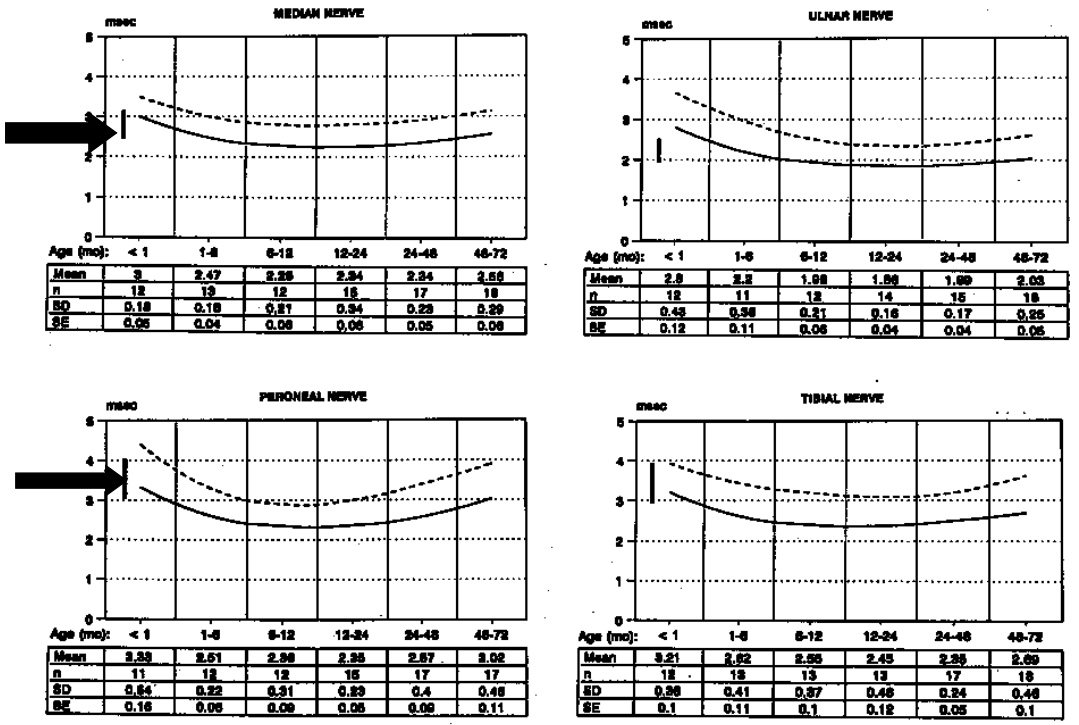


Fig. 1. Values of corrected DML for each age group (solid lines indicate mean values and broken lines mean + 2 SD). Vertical bars represent corrected DML in adults (mean + 2SD).

Garcia et al. Clinical Neurophysiology, 2000

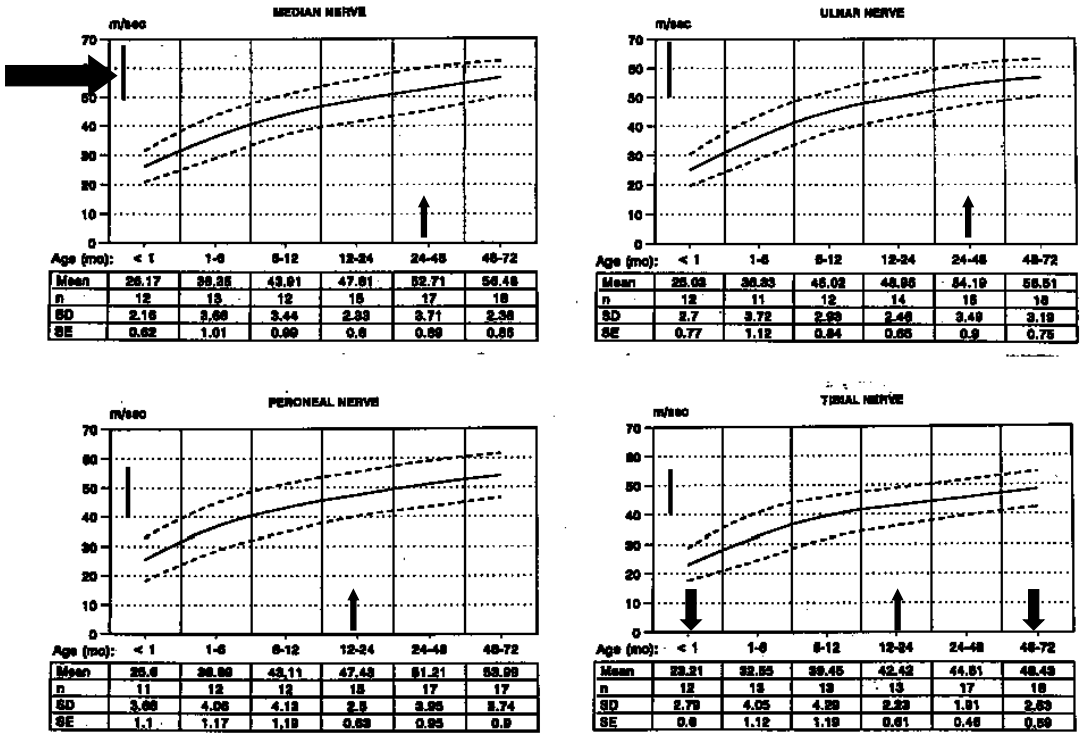


Fig. 2. Values of MCV for each age group (solid lines indicate mean values and broken lines mean ± 2 SD. Vertical bars represent adult MCV (mean ± 2 SD).

Garcia et al. Clinical Neurophysiology, 2000

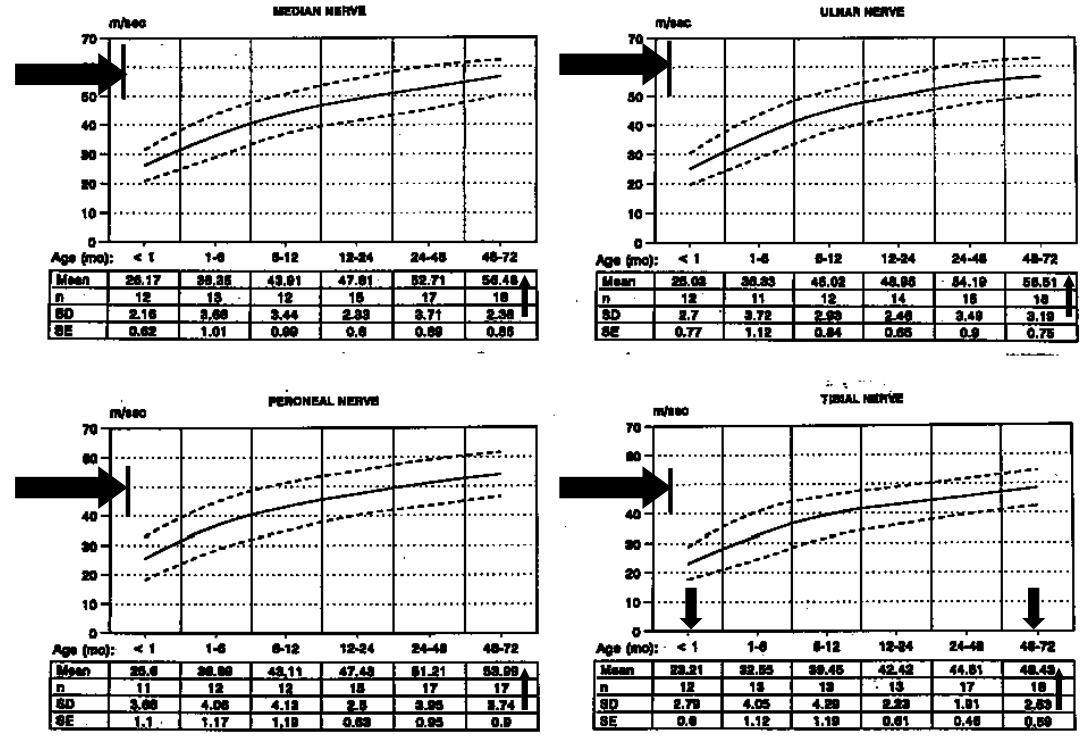


Fig. 2. Values of MCV for each age group (solid lines indicate mean values and broken lines mean ± 2 SD. Vertical bars represent adult MCV (mean ± 2 SD).

Garcia et al. Clinical Neurophysiology, 2000

CMAP amplitude (mV): mean ± SD

Age (months)	APB muscle	ADM muscle	EDB muscle	AH muscle
<1	1.27 ± 0.74	1.88 ± 0.92	1.77 ± 0.62	4.40 ± 1.73
1–6	2.37 ± 1.27	3.11 ± 1.45	2.68 ± 1.04	6.16 ± 2.44
6–12	2.94 ± 1.17	2.73 ± 1.09	2.64 ± 1.32	6.83 ± 2.69
12–24	4.12 ± 1.90	4.55 ± 1.53	3.69 ± 1.27	9.07 ± 2.12
24–48	5.96 ± 2.01	5.48 ± 1.42	4.25 ± 1.59	9.57 ± 3.54
48–72	6.96 ± 2.33	5.50 ± 1.55	3.78 ± 1.23	9.48 ± 2.39

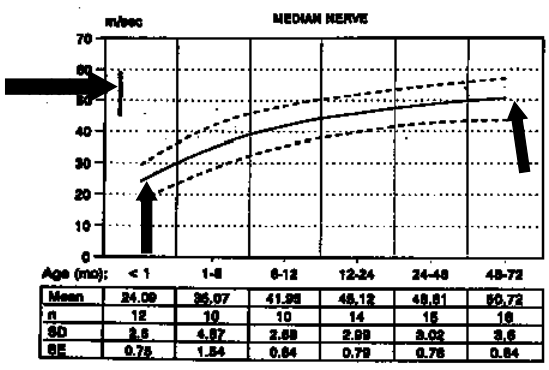
Garcia et al. Clinical Neurophysiology, 2000

SNAP amplitude (μV): mean ± SD

Age (months)	Median nerve	Tibial nerve
<1	4.86 ± 2.23	1.71 ± 0.74
1–6	10.66 ± 3.62	3.13 ± 1.48
6–12	9.00 ± 3.45	2.70 ± 1.43
12–24	15.72 ± 4.50	3.63 ± 1.14
24–48	12.02 ± 5.89	3.81 ± 1.53
48–72	14.04 ± 5.99	2.27 ± 0.83

Garcia et al. Clinical Neurophysiology, 2000

Sensory nerve conduction velocity studies



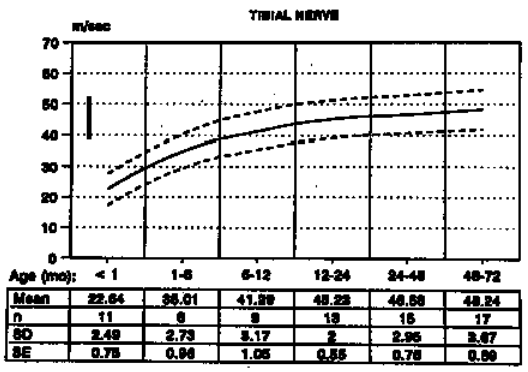


Fig. 5. Values of SCV for each age group (solid lines indicate mean values and broken lines mean ± 2 SD). Vertical bars represent adult SCV (mean ± 2 SD)

Garcia et al. Clinical Neurophysiology, 2000

EMG Accuracy Compared to Muscle Biopsy in Childhood

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

Rabie et al; Journal of Child Neurology, July 2007

EMG Accuracy Compared to Muscle Biopsy in Childhood

- Nerve conduction studies:
- 1 sensory + 2 motor in lower limbs
- 1 sensory + 1 motor in the upper limbs
- 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

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

Rabie et al; Journal of Child Neurology, July 2007

EMG Accuracy Compared to Muscle Biopsy in Childhood

- EMG: 74% (20/27) concurred w final Dx
- 26%)7/27) discordant
- Discordant EMG were myopathies
- Muscle biopsy: 87% (20/23) concurred with final Dx.
- 11/11 myopathies. 8/8 normal. 1 of 4 neurogenic

Rabie et al; Journal of Child Neurology, July 2007

EMG Accuracy Compared to Muscle Biopsy in Childhood

- 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
- 5 congenital myopathies: 40% normal, 40% myopathic, 20% neurogenic

Rabie et al; Journal of Child Neurology, July 2007

Case

- 5 Year old boy
- Long H/O ataxia
- Obligatory wheelchair user for a number of years
- Work-up has included muscle biopsy, results unknown
- C/O ⇑ weakness of lower limbs x 1-2 weeks

Case

- Muscle stretch reflexes absent
- Uncooperative with physical exam
- Nerve conduction studies: Absent sural & superficial peroneal sensory
- Motor NCS: See next 2 slides
- Needle EMG: ⇓ #, ⇑duration, ⇑ polys
- No abnormal spontaneous activity recorded


0V
0.1ms
1pps
1
3Hz
10kHz
Stim Intensity
R TIBIAL (KNEE) - AH
Ankle 1
50ms 5mV
8
Knee 2
50ms 5mV
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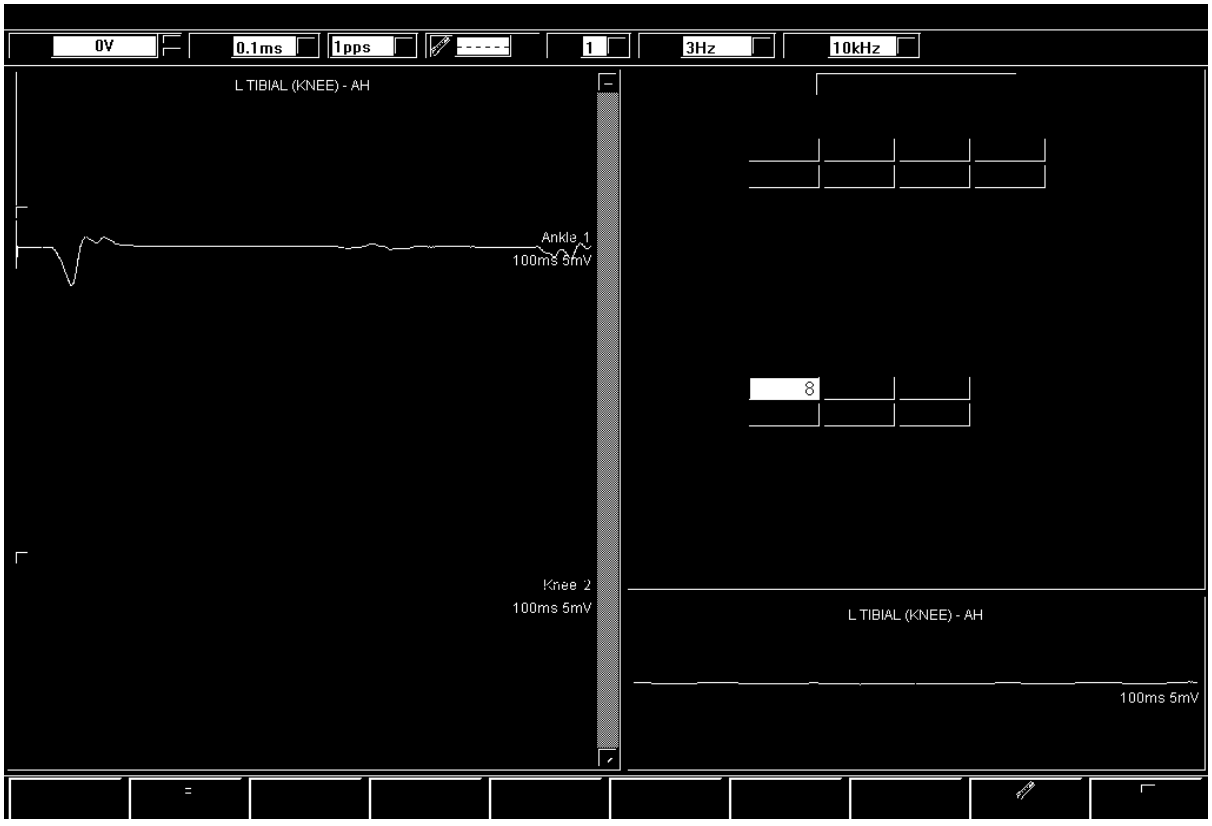
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L TIBIAL (KNEE) - AH
Ankle 1
100ms 5mV
8
Knee 2
100ms 5mV
L TIBIAL (KNEE) - AH
100ms 5mV

Case

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

SUMMARY

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

Selected References

Miller, RG; Kuntz, N: Nerve conduction studies in Infants and Children. Journal of Child Neurology. Vol 1 January 1986.

Bryant, P; Eng, G: Normal Values for the Soleus H-Reflex in Newborn Infants 31-45 Weeks Post Conceptional Age. Archives of Phys. Med. Rehabil Vol 72 January 1991

Wagner, AL; Buchtal, F: Motor and sensory conduction in infancy and childhood: Reappraisal. Dev Med Child Neurology 1972: 14: 189-216

Garcia et al; Peripheral motor and sensory nerve conduction studies in normal infants and children. Clinical Neurophysiology 111 (2000) 513-520

Selected References

Malcolm Rabie, Joseph Jossiphov and Yoram Nevo. Electromyography (EMG) Accuracy Compared to Muscle Biopsy in Childhood. J Child Neurol 2007 22:803

Thank you