The History of EDX in the Diagnosis of Demyelinating Polyneuropathies

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Financial Disclosure

• Sub-investigator for Ionis Pharmaceuticals (Familial Amyloid Neuropathy)
• Chapter author in *Medical Physiology*. eds: Rhoades & Bell. Wolters Kluwer
• Chapter author in *Rehabilitation Medicine Quick Reference series: Neuromuscular Disorders*, Prahlow & Kincaid. Demos Medical
NCS Development & Application

• 1948 Hodes, Larrabe & German describe Motor NCS techniques for arm & leg nerves using surface recorded responses

  Their velocity & amplitude normal values were similar those we use now

• 1956 Simpson showed application of NCS to focal neuropathy DX

• 1950’s Lambert applies NCS/ EMG techniques to wide range of neuromuscular disorders including inherited & acquired polyneuropathies

• 1957 Gilliatt & Thomas report very slow conduction times in Peroneal Muscular Atrophy patient: Ulnar motor wrist latency: 10 ms, elbow: 35 ms  CV = 11 m/s
Further EDX Evolution

• 1962 Bannister & Sears report prolonged distal motor latencies and sensory CVs in the mid-20 m/s range in acute idiopathic polyneuritis

• 1963 Dyck & Lambert report clinical & NCS findings in large CMT kindred: NCVs in 10-20 m/s range & distal latencies of 8-12 ms

• 1963 EMG Symposium in Copenhagen Lafayette & Mulder report findings in GBS pts: 14% were Normal, 61% had CV < 70% of normal mean & 25% had ↑ distal latencies with ~ Normal CVs Humphrey from Canada & Isch from France reported similar findings

• Early -mid-70’s papers by McQuillen from US, Eisen from Canada, Johnson from THE Ohio State & McLeod from Australia added to GBS EDX knowledge base
Conduction Velocities in a CMT Kindred

Fig. 4. Conduction velocity of motor fibers of the ulnar nerve between elbow and wrist in relation to age in the kindship studied. Each point represents 1 member of the family. The dotted line represents the lower limits of normal conduction velocity found in other studies in this laboratory. Symbols above this line represent persons with normal, those below, persons with low
The Flu  The Lawyers  The Government

• 1977 US Swine flu vaccine linked to ↑ incidence of GBS: Patients / Doctors / Vaccine makers / Lawyers / US Government are parties

• NIH & CDC calls on AAN/ANA expertise for DX Guidelines

• 1978 Guidelines for GBS DX by Asbury et al
  - 80% will have slowing or conduction block during the illness
  - CV is usually < 60% of normal but ABNLs are patchy
  - Distal latencies may be increased up to 3X normal
  - F-wave latencies give indications of proximal slowing
  - NCS may not become abnormal for several weeks
  - Up to 20% of patients may have normal NCS
Role of EDX Continues to Evolve

• Early to mid-1980s direct treatment for GBS becomes available
• Early mid-1980’s papers by McLeod, Albers, Donofrio, Miller, Cornblath, Brown & Feasby define EDX patterns, chronology & relation to prognosis
• 1982 Lewis & Sumner paper on EDX patterns in Familial vs Acquired Demyelinating Neuropathies
• 1984 Brown & Feasby paper on role of conduction block in GBS
• Which EDX parameter(s) correlate with prognosis for quicker / better recovery?
Figures 5 and 6. Representative median motor nerve conduction studies showing increased duration of compound action potential on proximal stimulation in an acquired patient. The familial patient had an increase in duration of 11%, whereas the acquired patient had dispersion of 47%. See figure 1 for comparison with normal.
Conduction Block

Brown & Feasby
Brain 1984; 107:219

FIG. 3. Examples o
sion in the maximum
op and bottom and
with Guillain-Barré
within two weeks of t
all 3 examples, the
durations extended t
control subjects betw
The Emergence of CIDP

• 1975 Dyck codifies CIDP
• Mid-80’s papers by Kelly & Albers begin process of better defining EDX parameters in acquired demyelinating neuropathies
• 1991 Research criteria developed by AAN task force: Cornblath et al
  o ↓ CV: < 70% LLN if CMAP< 80% LLN
  o Partial conduction block > 20% CMAP drop between prox & distal stim sites
  o ↑ Distal latencies: > 125-150% ULN depending on CMAP amp
  o ↑ F wave latency (if obtainable): >125-150% ULN depending on CMAP amp
  o Additional Sensory NCS & H reflex results
Some Real World Numbers

- CV LLN: Ulnar: 50 m/s 70% = 35 m/s Peroneal: 44 m/s 70% = 30 m/s

- Distal latency ULN: ulnar 3.5 ms 125% = 4.4 Peroneal ULN 6.5 125% = 8.2 ms

- F wave latency ULN ulnar 32 ms 125% = 40 Peroneal 56 ms ULN 125% = 70 ms
Sensitivity of EDX studies

• 1991 Bromberg paper demonstrates limits of sensitivity in EDX DX of CIDP: mid-60-ish % is the practical limit of sensitivity vs specificity in series of CIDP vs Hereditary Polyneuropathy vs Motor Neuron Disease vs Diabetic Polyneuropathy patients

• 1990 Cornblath paper gives history of EDX parameter development
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