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Recent advance of long term bed side EEG recording in critically ill patients: ccEEG

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The advancement of computer technology has allowed clinical application of long term EEG recording at bed side. This was not possible with “old fashioned” analog EEG. The digital EEG recording has been popularized only after the high resolution of video screen has become available for the last 10 years or so. This has brought a new era for clinical application of EEG. Besides the long-term EEG recording for the evaluation of epilepsy especially for the candidates of surgical treatment for the medically intractable epilepsy patients, there has been dramatic increase of bed side EEG recording for acutely and critically ill patients (ccEEG), especially at ICU settings. The ccEEG has found high incidence of seizures in patient with impaired consciousness, most importantly majority of these seizures are clinically silent seizures, i.e. non-convulsive seizure (NCS) or non –convulsive status epileptics (NCSE). Following are the indications of ccEEG

1) Presence of well defied interictal Spike/Spike-Wave, Periodic discharges (lateralized, bilaterally independent or generalized), Rhythmic Delta discharges, Brief (< 10 seconds) ictal like discharges.
2) After Convulsive seizures, Head trauma, Brian surgery, Intraparenchymal / Subarachnoid hemorrhage, CNS infection, Anoxic cerebral insult, Toxic/Metabolic encephalopathy
3) Assessment of treatment efficacy for NCS or NCIS
4) Monitoring of sedation or suppression therapy (Burst suppression pattern in EEG)
5) Prognostication of acute brain disease

Close to 90 % of seizures during acute cerebral dysfunction are captured within 48 hours recording.

The digital EEG allows technical modification or change such as sensitivity, filter setting, sweep speed and montage needed for the appropriate interpretation. Sophisticated computer program allowing qualitative/quantitative data analyses would be useful to facilitate the interpretation, however, these technology must be much improved in future for better accuracy and reliability.

Pathophysiology and treatment of facial synkinesis

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Facial neuritis occurs at the geniculate ganglion in the facial canal resulting from reactivation of herpes virus. Fascicular structure is lacking in the nerve trunk. So an aberrant circuit is easily generated at the time of regeneration. Nutrient vessels are running at outmost layer. If inflammation occurs here, nerve trunk is squeezed or entrapped with edema. At first, the vessels are occluded, so retrograde dying-back degeneration or axonotmesis develops. Then, much stronger compression ruptures inner membrane in association with Waller degeneration or neurotmesis.

An ENoG or electroneurogram representing axonal degeneration is inversely proportional to the severity of compression. Smaller is compression, bigger is ENoG. From a point of sequelae, it’s necessary to distinguish between axonotmesis and neurotmesis in axonal degeneration. Synkinesis appears in cases where ENoG is below 40%. In contrast, when ENoG is above 40%, synkinesis doesn’t develop. That is, at most 60% of axonal degeneration can be attributable to axonotmesis. For example, in a case with 15% ENoG, axonal degeneration account =85%. Furthermore, up to 60% of this is axonotmesis, therefore the remaining 25% is neurotmesis. This 25% is calculated in (40—ENoG), which means a possibility of Waller degeneration.

It is likely that regenerating neurites extend in the direction of voluntary movements. Since voluntary movement is the most effective for regeneration, even if Waller degenerating fibers are present, these regenerations are also promoted. Therefore, in cases where ENoG is below 40%, strong facial exercise is rather contraindicated for prevention of aberrant regeneration. In contrast, intensive stretching of the facial muscles may delay the nerve regeneration and extend to the original path, and there is a possibility of reducing aberrant re-innervation.

There is a fundamental relationship between clinical recovery curve and ENoG. The recovery is good and linear in the first 3 months, when dying-back fibers have arrived at the mimetics. On the contrary, it becomes a descending hyperbola after 4 months. This is because the Waller aberrant fibers have arrived at the mimetics.

Concerning treatment of synkinesis, there is no cure over prevention. In acute phase, do intensive stretching, which is likely to slow down regeneration and to reduce aberrant reinnervation in 12 months. In sequelae fixed phase, Botulinum toxin is effective. In the needle EMG for synkinesis, facial weakness is not re-innervated but rather simultaneous contraction of the agonist and the antagonist. To treat synkinesis, do weaken the agonist or the antagonist with Botulinum toxin.

Quantitative EMG

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Electromyography (EMG) reflects functional parameters of the motor unit, MU (axon, end-plate and muscle fiber) for diagnosis of nerve-muscle disorders. To achieve highest possible sensitivity to detect abnormalities, quantitative description of findings is of importance. Understanding of the relationship between generators and signals have emerged and analysis methods have moved from visual assessment to numerous ways of quantification. To compete with other methods for nerve and muscle studies e.g. ultrasound and to provide best medical value, EMGers need to practice electrophysiological methods skillfully and utilize new developments in signal analysis. Here focus will be on needle EMG, but advanced surface EMG will be mentioned.

Regarding nerve involvement, EMG describes motor denervation, reinnervation, nerve hyperexcitability, loss of axon and abnormal central activation of the neuron. Many of these parameters can be quantified.

End-plate abnormality is studied with repetitive nerve stimulation or better with SFEMG, not detailed here.

EMG is mainly used to study MU micro anatomy and function. Focus is to diagnose myopathy and neuropathy, their subgroups, severity and dynamics over time.

Modern EMG equipment provide robust, fast and objective methods for motor unit potential (MUP) analysis, many times with analysis time less than 3 minutes. MUPs from needle EMG electrodes are characterized with classical and new shape parameters, shape variability on repeated discharges, firing pattern. The signal pattern obtained with strong activation is analyzed with separate algorithms in the time domain (such as number of peaks and interpeak amplitudes) or in the frequency domain (such as FFT, frequency band description).

Methods using surface electrodes, are applied in EMG vs force correlations, multichannel recordings form multiple muscle groups in sports and physical medicine, activity dependent changes in frequency content in occupational medicine and pattern analysis in movement disorders. They are sometimes used in evaluation of neuromuscular disorders (analysis of tendency of EMG clustering in neurogenic disorders) or in mapping of MU activity with high density EMG (many hundred electrodes).

Other methods give new aspects about the MU (Macro EMG, Scanning EMG) or whole muscle (electrical impedance myography), not widely used.

To be useful QEMG needs good reference values. These are developed for many muscles for some equipment, and here quantification is commonly performed. Lack of such data bases is the main reason why QEMG is underutilized. The comment that quantification is too time consuming in busy routine, is not valid any longer with present analysis algorithms and procures.

On-Nerve Needle Sensory NCS for Studies of Peripheral Neuropathy

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Aims of ONN (On-nerve Needle) sensory NCS (Nerve conduction study):
■Direct comparison of sensory NCS and histology on the same nerve (sural nerve) by ONN sensory NCS.
■Diagnostic markers of axonal degeneration and demyelination by the sensory NCS.
■ Conversion factors in normal, axonal degeneration and demyelination

Materials: ONN Sensory Nerve Conduction in the Sural nerve during nerve biopsy was performed: in 89 cases
■50 cases: demyelinating neuropathy
■22 cases: axonal neuropathy
■5 cases: normal
■7 cases: mixed demyelinating and axonal neuropathy

Results:
1. Comparison of testing between ONN vs SE (Surface electrode techniques (N=89)
■In 55 (62%) cases, SNAP recorded with both techniques.
■In 21 (24%) cases, no SNAP in either technique
■In 13 (15%), SNAP absent in SE NC but recorded in ONN
2. Comparison of NC data between ONN vs SE techniques (N=55)
■CNAP amplitude : 2.9 x higher in ONN-NCS
■CNAP duration: 1.7 x longer in ONN-NCS
■NCV: No difference in NPNCV between two method
■Faster NCV in SE-NCS in maximal NCV
3. Conversion Factor
＊History
■ A linear relationship between the largest nerve fiber diameter and the fastest conduction velocity in animals: (Erlanger, Gasser, Grundfest & Hursh in 1930’s).
■Conversion factor (CF): the ratio between the largest fiber diameter and the fastest NCV
■NCV of myelinated fibers (m/s) = CF x outer diameter of myelinated fibers
■In 1975, Behse & Buchthal studies in 5 normal sural nerves Conversion factor :4.4
＊Conversion Factor Study (N=30 cases)
■Normal (N=2) : 4.3
■Axonal degeneration (N=4): 3.85
■Demyelination (N=24): 2.77: disproportional slowing
＊Conversion Factor (CF)

Conclusions:
■CF is helpful in differentiating demyelination from axonal degeneration
■36% decrease of NCV from the normal mean values (normal means – normal mean x 0.36) is a reasonable marker for demyelination:

4. Diagnostic Criteria of Axonal Degeneration vs Demyelination in Sensory NCS.
■ History: Sensory NCS not used as diagnostic markers of axonal degeneration and demyelination.
■ Present study: Axonal degeneration in 22 cases and Demyelination in 50 cases.
■Diagnostic Hallmarks of Sensory NCS
・Diagnostic sensitivity: 26% in SE-NCS vs 69% in ONN-NCS
・The most helpful diagnostic marker for demyelination: in both techniques
・Demyelination marker (>36% decrease of NCV from normal means)
＊Dispersion
・The most helpful diagnostic marker for Axonal degeneration: Low SNAP amplitude and No SNAP in ONN-NCS

Nerve Conduction Studies Workshop

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Over the years, we have come to recognize five simple principles in the practice of nerve conductions studies. These include 1) take a careful clinical history and examine the patient before initiating the study; 2) watch the muscle twitch and isolate the target under scrutiny before measuring the response; 3) compare responses elicited by shocks given distally and proximally to the site of lesion; 4) assess physiologic and pathologic changes by linearity and nonlinearity in segmental studies and 5) select a short distance for a focal lesion and a long passage for a diffuse process to improve sensitivity and reproducibility. This review discusses various uncommonly used methods to achieve the above objectives.

Ordinary conduction studies suffice to approximate the site of involvement in entrapment neuropathies. For more precise localization, inching the stimulus in short increments in the range of one to several cms along the course of the nerve can further isolate the site of involvement. In the evaluation of a focal lesion, studies of a longer segment tend to lower the sensitivity of the test because the inclusion of the unaffected segments in calculation dilutes the effect of slowing at the site of lesion. With stimulation of a normal nerve in 1 cm increments, the latency-changes approximately 0.2 ms/cm for both sensory and motor conduction. In patients with an entrapment syndrome, a latency- increase exceeding 0.4 ms across a 1 cm segment usually signals a focal abnormality. An abrupt nonlinear change in waveform of the recorded response provides an additional, and perhaps more convincing evidence.

In contrast, a longer segment provides a better result in assessing a more diffuse or multi-segmental process such as polyneuropathies. A longer path has an advantage in accumulating all the segmental abnormalities, which individually might not show a clear deviation from the normal range. Assume a nerve impulse conducting at a rate of 0.2 ms/cm (50 m/s). A 20% delay for a 10 cm segment yields only 0.4 ms, whereas the same change for a 100 cm segment amounts to 4.0 ms, an obvious increase for easy detection. Thus, the longer the segment, the greater the delay of conduction in assessing a diffuse process. Evaluating a longer, as compared to shorter, segment also improves the overall accuracy because the same absolute measurement error constitutes a smaller percentage of change in latency and distance.

In essence, the lesion identified on clinical grounds provides the overall orientation for the subsequent physiologic evaluation. Short distances magnify focal conduction abnormalities despite increased measurement error, and long distances, though insensitive to focal lesions, provide better yields and reliability for a diffuse process.

Single Fiber EMG

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Single Fiber EMG (SFEMG) is a highly selective EMG method allowing the recording of action potentials from one or a few muscle fibers at voluntarily or electrical muscle activation. The most commonly used parameter to be obtained is the "jitter", caused by a variability in the arrival time to two muscle fibers from the same motor unit. This is mainly due to a variability in the motor end plate time to initiate a muscle fiber impulse. Therefore, in cases of disturbed neuromuscular transmission e.g. myasthenia gravis (MG), the jitter is increased. Sometimes intermittent transmission failure may occur. The method is widely accepted as the most sensitive method to detect disturbed neuromuscular transmission, not only in MG, but in all situations where the end plate function is abnormal e.g. reinnervation.

To achieve the high selectivity, a special electrode is used, with a very small recording surface, and by using filter restrictions removing low frequencies (setting 500Hz to 10KHz). The electrode is inserted into the voluntarily activated muscle and a position if sought, where 2 muscle fibers from the same motor unit can be obtained. The time variability between them for consecutive discharges, i.e. representing the combined jitter from the two end plates, is normally 20-50 μsec. Alternatively, the muscle is activated by minimal intramuscular axonal electrical stimulation or surface stimulation. In this case the jitter is the latency variation between the stimulus and the single fiber signal, i.e. one endplate is involved. Reference values for both types of activation are available.

In recent time, the use of disposable material has been imposed, and therefore the facial needle electrode (i.e. the smallest concentric needle electrode) has been introduced as a surrogate. The principles for recording and analysis is the same as with real SFEMG electrodes. Since it is likely that many fibers form the same motor unit are recorded, special criteria have been formulated for accepted signals; clean positive-negative going signal without notches or shoulders, parallel rising segments when 10 signals are superimposed and more than 350 μsec between the two peaks to be included. Filter setting is 1000Hz-10KHz. The measurements should preferably be made using so called peak detection method. The sensitivity to diagnose myasthenia gravis is similar for recordings with SFEMG and with concentric needle electrodes.

This demo will include recordings with concentric needle electrodes, since these are the most commonly used. Voluntary and electrical stimulation will be demonstrated.

TMS: MEPs

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Transcranial Magnetic Stimulation (TMS) has been widely used in central nervous system functional analyses and clinical practice in humans because it activates human central nervous system non-invasively. These include several methods, such as single pulse stimulation, paired pulse stimulation, paired coils stimulation, repetitive TMS and so on. In this live demonstration, we will show mainly single pulse stimulation methods and a few paired pulse stimulation methods. Issues discussed are as follows.

Mechanisms of TMS and why magnetic? TMS finally activates neurons by electricity. The skull, bone, protects the brain from not only mechanical injury but also electric stress because of its very high resistance to electricity. Magnetic field produced by TMS defeats this protect because of no resistance of bones to magnetism, and finally induces eddy currents in the brain, which activate brain tissues by electricity. It enables us to activate human brain noninvasively.

Central motor pathway analyses: TMS has been most frequently used for central motor pathway functional analyses. CMCT: Motor evoked potentials (MEPs) were elicited by motor cortical stimulation (CTX) and spinal nerve stimulation at most proximal part of spinal nerves (Sp). The latency difference between these two sites stimulation (CTX-Sp) (central motor conduction time (CMCT)) is used to evaluate the central motor pathway conduction. In addition, we activate the central motor pathways at the level of pyramidal decussation with a double cone coil (brainstem stimulation: BST). CMCT is divided to CTX-BST conduction time and BST-Sp conduction time both of which reflect the intracranial and extracranial conduction time, respectively. This has given more information about the central motor pathway than CMCT. Cortico-Conus conduction time (CCCT), Cauda-Equina conduction time (CECT)

We developed a magnetic augmented trans-sacral stimulation (MATS) coil for activation of the conus medullaris in the spinal canal. This adds one more information about the motor pathways to the lower limb muscles. The CCCT indicates the conduction time through only the corticospinal tracts without any peripheral components. The CECT reflects the peripheral nerve conduction time within the spinal canal and detects abnormal conduction in the cauda equina. It is useful to evaluate the most proximal part conduction time in peripheral nerve disorders.

Short interval intracortical inhibition (SICI): When a subthreshold conditioning stimulus precedes a suprathreshold test stimulus by 3-5ms, MEPs to the test stimulus are smaller than the control MEPs elicited by the test stimulus alone. This suppression is produced by BAGAA function of the primary motor cortex.

Botulinum toxin injection

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(canceled)

SEPs

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Somatosensory evoked potentials (SEPs) are useful tool to evaluate the somatosensory pathway from the peripheral nerve to the primary sensory cortex. By recording several potentials generated by structures constituting the somatosensory pathway, we can localize the lesion along the pathway. This methodology still has a concrete diagnostic utility even in this MRI era, since MRI may show frequent false-positives and many disorders would not present with structural changes that can be documented by MRI. SEPs, as well as other electrophysiological tests, have a definite advantage that they can see “function” that is directly linked to the symptoms and signs of the patient. To enable such localization, determination of the generator of each SEP component is necessary. This has been achieved by studies of many investigators. I myself have also made some contributions.

In median nerve SEPs, P9 is generated as a junctional potential due to the change of the volume conductor size from trunk to neck. Upper cervical N13 (ucN13) is generated by the cuneate nucleus. N18 is also generated at the cuneate nucleus due to primary afferent depolarization of the presynaptic terminal of the dorsal column afferent fibers. Scalp P13/14 complex is composed of 3 subcomponents. P13 and P14a are generated by the startup of the medial lemniscal fibers as junctional potentials, whereas P14b is related to the thalamocortical fibers. In tibial nerve SEPs, P15 is generated at the greater sciatic foramen as a junctional potential due to the change of the conductance from soft-tissue to bone.

In this live-demonstration, I will show you a tibial nerve SEP examination. We simultaneously record 5 channels, and can register N8, the potential at the popliteal fossa, P15, a junctional potential at the greater sciatic foramen, N21, a postsynaptic potential at the posterior horn of the spinal entry zone, N30, mainly generated by the medial lemniscus, and P38, a potential from the primary sensory cortex. The combination of N8-P15-N21 is particularly useful in that it can localize the lesion along the peripheral nerve. In CIDP, prolongation of the proximal segments, N8o (N8 onset)-P15, P15-N21, and N8o-N21, is a characteristic finding. In lumbar spinal stenosis, prolongation of the P15-N21 segment is documented, even in patients lacking sensory symptoms or signs.

I believe I can show you how easily and reliably we can record above components of tibial nerve SEPs. Average of only 100 responses is often sufficient to clearly register these components.

P1-06-06

The isometric contraction of the index finger muscles shortens the cutaneous silent period in the first dorsal interosseous muscle

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Background: Muscle afferent feedback contributes to the maintenance of alpha motoneurons firing during sustained muscle contraction. The current view holds that position and movement signals come from a combination of sensory inputs from the muscles, joints and skin. We aimed to investigate how the isokinetic and isometric exercises of the index finger change sensorimotor modulation measured by cutaneous silent period (CuSP).
Methods: CuSP was recorded from the first dorsal interosseous (FDI) and abductor pollicis brevis (APB) muscles consecutively after stimulation of the 5th and 3rd digits on 6 healthy subjects before and after 2 exercises. The participants moved continuously the index finger of the dominant hand on a computer mouse left and right buttons for 5 minutes. Then, the non-dominant hand index finger was maintained in extension with FDI submaximal isometric contraction for 5 minutes. The thumb was fixed in neutral flexion position.
Results: CuSP measured on FDI after isometric contraction showed reduced average duration of 27.75ms in comparison with the baseline 47.2ms. In the control APB muscle CuSP was 42.25ms compared to 45.2ms obtained before the exercise. The isokinetic exercise didn´t produce semnificative changes in CuSP.
Conclusion: The sensorimotor circuitry of the CuSP might be affected by isometric exercise.

P1-12-01

The effect of body temperature on SEP latency during intraoperative neuromonitoring.

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Background: The latency of intraoperative somatosensory-evoked potentials (SEP) tends to be prolonged at the end of surgery than baseline. Few studies investigated the relationships between values in intraoperative SEP and various physiological parameter. Method: For 3 years, a total 343 patients performed with intraoperative SEPs monitoring. The demographic characteristics, systemic parameter and SEPs results were retrospectively reviewed. We divided into two groups according to the SEP latency change of baseline to end of operation. Results: A total of 325 cases data were obtained. At the end of operation, 195 (60%) showed prolonged left N20 SEP latency compared with baseline latency (left P40, 63.9%, right N20 60.8%, right P40 62.3%). To evaluate the systemic factors correlating to the change of SEP latencies, we performed the correlation analysis. Positive correlation: age and estimated blood loss; Negative correlation: the change of body temperature ($r=-.737$, $p<0.001$), the operating time, and the change of mean arterial blood pressure. Conclusion: In normal intraoperative SEP monitoring, the SEP latency is often delayed. The delay of SEP latency is most influenced by the change of body temperature. As the body temperature decreases by 1°C, about 4% of the SEP latencies rises.

P1-12-02

Factors associated with successful generation of baseline motor evoked potentials in patients with spine surgery

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Background: The aim of this study was to investigate clinical factors associated with the successful generation of baseline muscle motor evoked potentials (mMEPs) in patients undergoing spine surgery and to determine the relationship with postoperative functional outcome.
Methods: From July 2014 to June 2015, a total of 345 patients underwent spine surgery with intraoperative mMEPs monitoring and were consecutively included in this study.
Results: Two hundred and fifty-three (73%) patients showed the successful generation of baseline mMEPs at all recording muscles, and 92 (27%) patients failed to record baseline mMEPs at one or more limb muscles. Male sex, cervical/cervicothoracic lesion location the presence of T2-weighted HSI of cord, preoperative Nurick grade, and MRC grade showed a significant contribution for predicting the successful generation of mMEPs. When analyzing predictors for postoperative poor functional outcomes, the failure of mMEPs generation and malignant etiology were significantly related factors for the aggravation of the Nurick grades before and after surgery.
Conclusion: Various clinical parameters including lesion location, abnormal cord signal on MRI, preoperative motor power, and functional status were revealed to have a significant relationship. Additionally, the successful generation of mMEPs was one of the indicators for predicting the long-term functional outcome.

P1-15-07

Outcome of functional motor disability in ankle dorsiflexion in children with cerebral palsy following low dose botulinum toxin A injection

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Background: To assess the outcome of functional motor disability in ankle dorsiflexion in children with cerebral palsy who are having spastic diplegia following low dose botulinum toxin A (BTX-A) injection.
Methods: Ten children with cerebral palsy who are having spastic diplegia, who had received low dose BTX-A injection to gastrocnemius-soleus group of muscles were included. They have undergone a course of physiotherapy treatment following BTX-A injection. The outcome measures were range of motion (ROM) of affected joints, muscle tone, gait and selective motor control of dorsiflexion. These children were assessed before the injection and 3 months post-injection.
Results: Muscle tone (p=0.035), ROM of both right and left ankle dorsiflexion (p=0.001, p=0.002) showed a significant improvement. There is a marked improvement in selective motor control of dorsiflexion (p=0.016). In the physician rating scale for gait there was a significant improvement in the timing of heel rise (p0.089).
Conclusion: Standard BTX-A treatment recommended for cerebral palsy is expensive. Results of the present study shows that low dose BTX-A injection to gastrocnemius-soleus group of muscles decreases the spasticity and improves the functional ability of ankle in children with cerebral palsy as compared to the improvement reported in the literature using standard treatment.

P1-17-06

Clinical and neuro electrophysiological manifestations of two patients with sensorimotor neuron disease (FOSMN) syndrome presenting with facial symptoms

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Background: To characterize the clinical and neuro-electrophysiological manifestations of FOSMN syndrome.

Methods: Retrospectively analyzed two patients with FOSMN syndrome admitted in Peking Union Medical College Hospital in June 2018.

Results: Case 1, a 57-year-old male, developed facial sensory disturbance in 2016, followed by difficulty of drinking and chewing, dysarthria, upper limbs weakness, atrophy of temporalis, masseter, tongue, neck and upper limbs, hyperactive tendon reflexes. The corneal reflexes reduced. NCV was normal. EMG showed positive-sharp-waves and neurogenic MUAPs in sternocleidomastoid and upper limbs. Blink reflexes were absent bilaterally.

Case 2, a 63-year-old male, developed lips sensory disturbance in 2010, followed by cheek and left upper limb numbness, difficulty of drinking, chewing and swallowing. Tongue, temporalis and masseter were less plump, neck strength was weak, corneal reflexes were absent. NCV showed decreased amplitude of the median nerve. EMG showed neurogenic damage in the sternocleidomastoid and positive-sharp-waves in the parathorax muscles. Blink reflexes were absent bilaterally.

Conclusion: There were generalities of FOSMN: bulbofacial sensory disturbance, followed by bulbar, upper trunk and upper limbs symptoms; corneal reflex reduced or absent; abnormal Blink reflex; EMG showed neurogenic damages. The present two cases manifested FOSMN syndrome clinically. However, case 1 needed long-term follow-up for further confirmation.

P1-18-08

Characteristics of long term exercise test in patients with hypokalemic periodic paralysis

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Objectives: The exercise test can be used to detect the non-episodes of hypokalemic periodic paralysis. After exercise, the amplitude of composite muscle action potential (CMAP) decreases. The aim of this study is to find out the best recording time by analyzing the characteristics of the decrease in amplitude after exercise.

Methods: Subjects included 56 patients with periodic paralysis. We recorded the CMAP amplitude of the ulnar nerve at quiet state and 0, 20, 40, 60, 90 and 120 minutes after exercise. Continuously abduction about 45 seconds, rest 15 seconds as 1 sequences with a total of 5 sequence movements.

Results: We use the amplitude decreased by 33% was the positive index as reported previously in our EMG room[calculation method: (amplitude of pre exercise amplitude, post-motion amplitude) / pre-motion amplitude *100%]. There were 10 patients (17.9%), 36 patients (64.3%), 9 patients (16.1%), 0 patient and 1 patient (1.7%) were found decreased below 33% in 20, 40, 60, 90, 120 minutes, respectively.

Conclusion: Long term exercise test is an effective method for the diagnosis of periodic paralysis. Most of the positive results occur within 60 minutes. Thus, the time recording time can be shortened to 60 minutes.

P1-23-09

Nerve Conduction Study Profiling in HIV associated Neuropathy on Antiretroviral Treatment

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Background: Distal symmetrical polyneuropathy (DSP) is one of the most frequent neurologic complications of human immunodeficiency virus (DSP-HIV) infection that treated with antiretroviral treatment.. Distal axonopathy may be detected pathologically in DSP.
Methods: It was cross sectional study during 2015-2017 at Cipto Mangunkusumo Hospital Jakarta. Exclusion criteria were history or current use of stavudin, diabetes mellitus, Guillain Barré Syndrome, Morbus Hansen. Diagnosis of HIV-SN was made by AIDS Clinical Trial Group Brief Peripheral Neuropathy Screening Tools (ACTG-BPNST). Nerve conduction study (NCS) was performed for motor nerve of median, ulnar, peroneal, tibial nerves and sensory nerve of median, ulnar, superficial peroneal and sural nerves.
Results: HIV-SN was found in 9 of 92 patients (9.8%). Neuropathic pain was found in 6/92 subjects (6.5%). The NCS characteristics of DSP-HIV are SNAP amplitude of sural, superficial peroneal, median nerve were found significantly reduced, SNAP onset latency of superficial peroneal, median and ulnar were prolonged; superficial peroneal and sural sensory conduction (CV) velocity were reduced. Motor and sensory NCS was normal in neuropathic pain subjects.
Conclusions: DSP-HIV involves predominantly sensory nerve fibers rather than motor nerve fibers. Neuropathic pain as symptoms of DSP-HIV is not associated with nerve conduction abnormality

P1-24-01

The clinical and electrophysiological features of Guillain- Barré syndrome in Mongolia

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Background: No studies have been performed in Mongolia about Guillain Barré Syndrome (GBS). In recent years this disease can be classified into different clinical subtypes according to the electrophysiological criteria.
Purpose: we aimed to study the age, gender, preceding factors, clinical features and electrophysiological subtypes of GBS in our country.
Methods: 54 patients with acute weakness and suspicion of GBS were studied retrospectively. A careful disease history was taken, the degree of weakness was adjusted using the MRC scale. The GBS was classified electrophysiologically according to the Hadden criteria.
Results: The GBS affects all ages in our country. In about the half of patients no preceding factors could be evaluated, 43% had infections. The muscle tendon reflexes were absent in 44%, severe degree of paresis was revealed in 20%, in 32% the brain nerves were involved. According to the Hadden criteria the acute demyelinating polyradiculoneuritis was identified in 51% of patients, whereas the acute motor axonal neuropathy occurred in 36%. The remaining 13% could not be classified.
Conclusion: For the first time we inquired the subtypes of GBS in Mongolia. This study should be intensified in future.
Key words: Guillain Barré syndrome, acute motor axonal neuropathy, acute demyelinating polyradiculoneuropathy, Hadden criteria

P1-24-02

Epidemiology of Guillain-Barré Syndrome in State of Penang, Malaysia

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Background: Recent systematic analysis showed that incidence rate of Guillain-Barré Syndrome(GBS) worldwide ranges between 1.1-1.8 per 100,000 population/year.

Methods: This is a retrospective analysis of patients with GBS, diagnosed from Year 2010 till 2017 at Penang General Hospital. Information regarding distribution by gender and race, age of onset, presence of GBS bulbar palsy and presence of antecedent infections are collected from Penang General Hospital Neurology Clinic archive. Annual incidence rate is calculated using population data provided by Department of Statistics Malaysia(http//: pqi.stats.gov.my).

Results: A total of 73 patients are diagnosed with GBS from 2010 till 2017. 48%(n=35) of patients are male and 52%(n=38) are female. Majority of subjects(43%) being diagnosed with GBS at age range of 61-70 years(n=25). There is preponderance of Malay patients(45%, n=33) who are diagnosed with GBS, with Chinese and Indians constituting 30%(n=22) and 23%(n=17) respectively of the cohort. Within 2010 to 2017, the incidence rate among Penang population ranged from 0.17 to 0.78 per 100000 population. 21%(n=15) patients had GBS with Bulbar Palsy.

Conclusion: GBS Incidence rate among Penang population is lower compared to GBS overall worldwide incidence rate. GBS presents commonly individuals within age range of 61 to 70 with Malay ethnicity being commonly affected.

Recent Developments in the Diagnosis and Management of Myasthenia Gravis

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Myasthenia gravis (MG) is the most common acquired disorder of neuromuscular transmission at the neuromuscular junction (NMJ. It is increasingly seen as a syndrome than a single disease. The pathogenesis is related to immune defects in regulatory T and B cells. Multiple targets at the NMJ known include the AchR, MuSK, LRP4, and other others lesser known domains. The thymus can be the site of immune sensitization or antigenic production in the presence of thymomas.

Diagnosis of MG can be clinically challenging, especially in antibody negative cases limited to ocular manifestations. Previous studies in this area contained confounders of nonblinding, incorporation and spectrum biases. Lacking a diagnostic gold standard, neurological examination, treatment response, ice test, repetitive nerve stimulation (RNS) and single fiber electromyography (SFEMG) are collectively used to diagnose MG. A combination of the ice test and SFEMG was found to provide the most reliable diagnosis of MG.

In a large Asian cohort of 191 patients, we found that the conversion rate of ocular to generalized MG was unexpectedly low (10.6%) on 40.8 months follow up and 7.7% on 24 months follow up. Predictive factors for conversion are presence of AchR antibody, thymoma and a positive RNS study.

Recent emergence of immune checkpoint inhibitor MG, in tandem with myositis and myocarditis, is due to its increasingly widespread usage. While the majority of cases arises de novo, exacerbation of pre-existing clinical or subclinical MG should be excluded.

Treatment of acute MG exacerbation consists of supportive therapy, plasma exchange or intravenous immunoglobulin. Apart from corticosteroids, use of immunosuppressive agents for steroid sparing effect is derived mainly from retrospective data. However, a recent randomized trial has demonstrated the benefit of thymectomy in terms of clinical improvement and steroid reduction.

Emerging treatment strategies include targeting T-cell signaling pathways, T/ B cell interactions, B cell and plasma cells. Monoclonal antibodies against Fc receptors, interleukins and complement are under development. Other adjuvant research treatments are vaccination, autologous stem cell transplantation and drugs acting at the muscular level

E8-1 (PA-01)

Sequential changes in clinical symptoms, EEG and ECG during ictal asystole

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Background: Ictal asystole has been intensively investigated because of its possible contribution to SUDEP. However, the sequential changes in clinical symptoms, EEG, and ECG during ictal asystole remain to be elucidated.
Methods: We analyzed clinical symptoms, EEG, and ECG in three epilepsy patients with ictal asystole (total seven seizures with ictal asystole).
Results: Clinical ictal symptoms appeared 5 to 22 sec after EEG seizure onset. Tachycardia started 8 to 26 sec after the clinical onset, and it was followed by bradycardia with 11 to 35 sec interval. Ictal asystole started 9 to 21 sec after bradycardia. After the onset of ictal asystole, general atonia and generalized EEG suppression appeared within 9 to 10 sec. Heartbeat restarted 3 to 4 sec after generalized EEG suppression. The asystole lasted for 6 to 19 sec. After the restarting of heartbeat, EEG showed generalized irregular delta waves within 3 to 16 sec, and multifocal myoclonus occurred within 6 to 18 sec.
There were tachycardia in 4 seizures, bradycardia in all, muscle atonia in 2, generalized EEG suppression in 3, and myoclonus in 2.
Conclusion: There are stereotyped sequential changes in symptoms, EEG and ECG during ictal asystole.

E8-2 (PA-02)

Magnetoencephalography features of Parkinson’s disease

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Background: Studies using electrocorticography have revealed that phase-amplitude coupling (PAC) is exaggerated in the primary motor cortex (M1) in patients with Parkinson’s disease (PD) and that PAC relates to motor symptoms of PD. We investigated the whole-brain PAC of patients with PD and healthy controls (HCs) non-invasively using magnetoencephalography (MEG) to demonstrate that PAC could be a physiological biomarker o PD.
Methods: We recorded the MEG signals of 21 patients with PD and 34 HCs in both a resting state and during a fingertapping task. We evaluated resting-state PAC, PAC during movement preparation and time series of PAC during a movement task.
Results: In a resting-state, β-low γ PAC on the primary visual area was significantly higher in patients with PD than in HCs. β -low γ PAC of patients with PD on M1 significantly related to motor symptoms. During movement preparation, no significant PAC was found in patients with PD while α-γ PAC were found on a wide range of areas in HCs. During a fingertapping task, α-low γ and β-low γ PAC showed significant time variance on the sensorimotor areas both in patients with PD and HCs.
Conclusions: PAC characterizes PD and thus can be a biomarker of PD.

E8-3 (PA-03)

Effect of acute optic neuropathies on Light Peak: Dark Trough ratio (Arden ratio) of electro-oculography

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Background: Electro-oculography (EOG) is an electrophysiological test of measure of retinal pigment epithelium (RPE). There are only very few studies have been reported regarding the effect optic neuropathies (ON) on EOG. This study aimed to determine the influence of acute ON on Light Peak: Dark Trough (LP: DT) ratio of EOG.

Methods: We recruited 40 patients referred to the Neurophysiology unit, Teaching Hospital, Peradeniya, Sri Lanka with a provisional diagnosis of having acute ON. We recorded EOG waveforms in all of them using the Natus NCV/EMG/EP machine. LP: DT ratios were determined in each patient and compared with the similar values of 40 control subjects using the Mann -Whitney U test.

Results: Seventeen out of 40 patients (42.5%) had reduced LP: DT ratio with a median LP: DT ratio (± inter quartile range) of 1.40±0.15, compared to the subsequent value of 1.75±0.20 of the controls. The difference was statistically significant ($p < 0.001$).

Conclusions: Some patients with acute ON had reduced LP: DT ratio, which indicates that the RPE can be affected in acute episodes of ONs. This can be due to edema of optic nerve head or changes in choroidal circulation occur in ON, causing abnormalities in RPE.

E8-4 (PA-04)

Predicting children with ADHD using prefrontal cortex activity

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Background: Many studies have supported the notion that executive function impairments explain the core symptoms of attention deficit hyperactivity disorder (ADHD). Their neural basis is generally accepted to be in the prefrontal cortex. Thus, methods to quantify brain functions may ultimately assist both in the differential diagnosis of ADHD and in assessing novel interventions. To establish valid, objective biomarkers for ADHD using machine learning for brain function is necessary.

Methods: Machine learning was used to predict disorder severity from new brain function data, using a support vector machine (SVM). A multicenter approach was used to collect data for machine learning training, including behavioral and physiological indicators, age, and reverse Stroop task (RST) data from 108 children with ADHD and 108 typically developing (TD) children. Near-infrared spectroscopy (NIRS) was used to quantify the change in prefrontal cortex oxygenated hemoglobin during RST. Verification data were also collected from 62 children with ADHD and 37 TD children from six facilities in Japan.

Results: The SVM general performance results showed sensitivity of 88.71%, specificity of 83.78%, and an overall discrimination rate of 86.25%.

Conclusions: A SVM using an objective index from RST may be useful as an auxiliary biomarker for diagnosis for children with ADHD.

E8-5 (PA-05)

Individualized transcranial alternating current stimulation improves motor memory consolidation in humans: A double-blinded sham-controlled study

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Background: Previous studies have demonstrated that the frequency-dependent functional coupling between the primary motor cortex and the motor neurons of target muscles is increased following motor skill practice. This could reflect processes related to motor memory consolidation, but this is unknown. Here, we speculated that entraining this frequency dependent communication following motor practice through transcranial alternating current stimulation (tACS) would lead to a greater motor memory consolidation in humans.

Methods: Twenty young individuals were participated in this study. Motor skill learning task was a visuo-motor tracking task involving tonic ankle dorsiflexion. tACS was individually targeted at EEG-EMG peak beta-band frequency at 2mA for 10 minutes. EEG was measured from the vertex (Cz) and 5cm frontal to Cz. EMG was measured from the right tibialis anterior muscle. Corticomuscular coherence was estimated during a tonic 2-min dorsiflexion at 10% MVC.

Results: Skill acquisition did not differ between groups. Peak corticomuscular coherence in the beta-band was increased following motor practice. The changes in performance from end of motor practice to a day and 7 days after practice were greater in tACS group compared to sham-tACS group.

Conclusions: Individualized tACS improves motor skill retention. Future studies could seek to translate these findings into patient populations.

E8-6 (PA-06)

Diagnosis of the lumber radiculopathy using magnetospinography.

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Background:

The biomagnetic field measurement is not affected by the surrounding tissue and theoretically has high spatial resolution. We previously reported noninvasive visualization of neural activities in the lumbar spine using magnetospinography (MSG) in healthy subjects. We report here MSG findings of patients with lumbar radiculopathy.

Methods:

We measured the neuromagnetic fields of five patients with L4/5-disc herniation or L5/S-disc herniation at the surface of the lumbar spine area after stimulation to the peroneal nerves and tibial nerve (duration 0.3 ms, averaging 2,000 times). Neural currents were estimated using spatial filter techniques and were superimposed on X-ray images or MRI of the lumbar spine.

Results:

On the healthy side, the nerve action current calculated from MSG passed through the intervertebral foramen and propagated in the caudal-to-cranial direction. On the affected side, the nerve action current attenuated at the L4/5 or L5/S disc herniation area.

Conclusion:

Thus, MSG visualizes propagating spinal nerve excitation with high spatial and temporal resolution, allowing us to quantitatively evaluate the decreased action current in lumbar radiculopathy. MSG is expected to be a clinically useful modality.

E8-7 (PA-07)

Pathophysiology of eculizumab treatment for Guillain–Barré syndrome: an electrophysiological study.

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Background: A previous study suggested the efficacy of eculizumab therapy for patients with Guillain-Barré syndrome. To further elucidate the pathophysiology of the efficacy, we investigated the nerve conduction study (NCS).

Methods: We investigated the NCS of 33 patients who participated in the Japanese eculizumab trial for Guillain-Barré syndrome (JET-GBS). Antiganglioside IgG antibodies (GM1, GD1a, GalNAc-GD1a, GQ1b, GM1/GD1a, GM1/GalNAc-GD1a, GM1/GQ1b and GD1a/ GQ1b) were measured at entry. Reversible conduction failure (RCF) was defined as CMAP amplitudes at 4W recovered > 150% compared with that at entry.

Result: RCF occurred in 26% (34/136) of nerves, and the frequency of RCF was higher in patients with antibodies than in those without antibodies (29% vs 5%, $p < 0.05$). In the antibody-positive group, RCF occurred more frequently in the eculizumab group than in the placebo group (38% vs 15%, $p < 0.05$), and the patients with RCF were able to run within 4 weeks more than the patients without RCF ($p < 0.05$).

Conclusions: In patients with antiganglioside antibodies, eculizumab may inhibit complement activation and block the nerve damage caused by the antibodies before axonal degeneration to improve outcomes during the sub-acute phase of GBS. How eculizumab might affect patients without antiganglioside antibodies requires further investigation.

E8-8 (PA-08)

Development of CTS SCORE for Rapid Assessment of Carpal Tunnel Syndrome Diagnosis and Severity

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Background: Carpal Tunnel Syndrome (CTS) is the most common entrapment neuropathy among adults. The aim of this study is to develop a scale that can be done at the clinics to predict its diagnosis and severity.

Methods: 310 consecutive patients referred for CTS were evaluated. Using laboratory values, EMG-NCV was performed to confirm diagnosis. Univariate/Multivariate logistic regression analysis was done to identify significant parameters and to determine cut-off point.

Results: 343 hands were CTS positive. A multiplier of 1.75 was used as coefficient value. A scale was derived using Phalens (3 points), Tinels (2 points), thumb strength (2 points), age> 40 (2 points), tingling (2 points), numbness (2 points), nocturnal awakening (1 point), and extramedian symptoms (1 point). The mean cutoff value for CTS positive (mild CTS) was 4.35, moderate CTS at 7.35 and severe CTS at 11.48. The area under the curve shows that the cutoff point is 82% sensitive and 81% specific.

Conclusions: This simple scale can be used at bedside to predict the diagnosis and severity of CTS. However, some limitations would be that it does not account the possibility of concomitant CTS mimickers and that it should not replace an EMG-NCV as standard of care.

E8-9 (PA-09)

Frequency of CRD in a variety of disease and individual muscles

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Background: CRD occurs in various myopathy and chronic lower motor neuron diseases. Since it also occurs in normal healthy cases, CRD is considered to be a nonspecific abnormality. Clinical significance of CRD has been debated. We analyzed the frequency of CRD in a variety of disease and individual muscles. From this examination, we considered clinical usefulness of CRD.
Method: 1277 muscles in 330 consecutive cases were retrospectively analyzed.
Results: CRD was observed in 40 muscles in 28 cases. The occurrence was higher in the paraspinal muscle (11.8%) and iliopsoas muscle (8.2%), and lower in the triceps brachii (0%) and extensor digitorum muscle (0.7%). In each diagnostic category, the appearance ratio of CRD was higher in motor neuron diseases and myopathy, at 11/39 cases (18/285 muscles) and 6/22 cases (7/93 muscles), respectively. No CRD was observed in the normal cases. Among eight cases who recorded CRD in two or more muscles, four of them were ALS.
Conclusion: Presence of CRD should be first considered as associated with some kind of neuromuscular disease. Appearance of CRD in normal healthy cases is less than expected.

E8-10 (PA-10)

Comparison of muscle motor evoked potential changes between cervical and thoracic ossification of the posterior longitudinal ligament surgery

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Backgrounds: The aim of this study was to compare intraoperative changes in muscle motor evoked potential (mMEP) between cervical and thoracic ossification of the posterior longitudinal ligament (OPLL) surgery in terms of the predictive value for postoperative motor deficits (PMD).
Methods: In total, 88 patients with OPLL were identified at a single tertiary hospital between January 2010 and December 2015. One hundred and forty-one limbs from 78 patients with cervical OPLL and 15 limbs from 10 patients with thoracic OPLL were included in the analysis. PMD was assessed at immediate postoperative period and 1-year follow-up.
Results: PMD was more frequently developed in thoracic OPLL than in cervical OPLL (immediate PMD, 6.60% vs. 2.80%; 1-year PMD, 20.0% vs. 1.40%). As for the mMEP disappearance criteria, the sensitivity for 1-year PMD in cervical OPLL was higher in tibialis anterior (TA) muscle than that in abductor hallucis (AH) muscle, in contrast, both the sensitivity and positive predictive value for 1-year PMD in thoracic OPLL was higher in AH muscle than those in TA muscle.
Conclusions: The alarm criteria for mMEP changes should be differentiated between cervical and thoracic OPLL surgery in practical intraoperative monitoring.

Mismatch negativity as a biomarker reflecting pathogenesis in the superior temporal gyrus

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Automatic change-detection system has been developed in human acoustic brain, probably for survival competition in the primitive times. Such a system enables us to notice the incoming sound-changes of warning even if ignored. This system has been revealed by investigating the mismatch negativity (MMN) in the brain (Naatanen et. al., 1978). MMN is considered to be generated by the comparison process between sound change and the memory trace of the preceding sounds stored in sensory memory of the brain. This "memory trace theory" has been supported by plenty of studies. MMN can be elicited by various types of sound changes, such as change of frequency, intensity, duration, spatial location, phonetic change, and even a stimulus omission. Actually, omission-MMN was not elicited with SOAs of 160 ms or longer, suggesting the function of temporal window of integration (TWI). Namely, the duration of the neural trace encoded in sensory memory corresponds to TWI of 160-170ms (Yabe et al., 1997).

Recently, MMN is expected to be one of the promising neurophysiological bio-markers in schizophrenia, because the impaired MMN reflects the cognitive dysfunction and/or psychosocial impairment in the patients with schizophrenia. Interestingly, a definite reduction in early schizophrenia was evident in MMN to "duration" but not frequency deviants (Todd & Michie et al., 2007). The impaired duration-MMN might be caused by the dysfunction of TWI. Most importantly, MMN might provide the prediction of conversion to psychosis when the duration MMN was recorded in clinically at riskmental state (ARMS) individuals (Sumiyoshi & Higuchi et al., 2013; Natanen, Shiga & Yabe et al., 2015).

The main generator of MMN is located in the superior temporal gyrus (STG) as demonstrated by magnetoencephalographic (MEG) studies and electroencephalographic (EEG) studies in humans, electrical recording in cats, and monkeys (Naatanen, 1992). Many neuroimaging studies have revealed structural abnormalities of STG in schizophrenia (Kasai et al., 2003). MMN also suggests the impaired function of NMDA receptors (Javitt et al., 1996). However, little postmortem study has paid attention to STG in schizophrenia. DARPP-32 and calcineurin (CaN) measurable in the postmortem brain reflect the activities of the dopamine and glutamate systems. By combining MMN findings in STG with the postmortem study, the pathogenesis as reflected by DARPP-32 and CaN has been revealed more in STG than previously found in the prefrontal cortex (Kuni & Yabe et al., 2011, Wada & Yabe et al., 2017). Thus, MMN is a promising neurophysiological biomarker in schizophrenia.

Distinguishing Features of the Repetitive Nerve Stimulation Test between Lambert-Eaton-Myasthenic Syndrome and Myasthenia Gravis 50 years Reappraisal

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Objective: To reappraise the distinguishing features of the repetitive nerve stimulation (RNS) tests in the abductor digiti quinti muscle between myasthenia gravis (MG) and Lambert-Eaton myasthenic syndrome (LEMS) 50 years after the 1965's Lambert seminal paper.

Methods: The various parameters of the RNS test were compared between 34 LEMS patients and 140 MG patients to assess their diagnostic sensitivity.

Results:

- RNS test was abnormal in all (100%) LEMS patients and 76 (54%) MG patients.
- The diagnostic hallmark of LEMS, ≥ 60% increment at high-rate stimulation (HRS) or post-exercise facilitation (PEF), was present in 33 (97%) of patients.
- The diagnostic hallmark of MG, decrement at low-rate stimulation (LRS), was present in 65 (46%) of MG patients.
- The most prominent difference in the various parameters of RNS tests was noted between LEMS versus MG and normal controls.

Table 1 Diagnostic sensitivity of test parameters in the repetitive nerve stimulation test

	LEMS (N=34)	MG (N=140)	Normal limit (N=40)
Abnormal	34 (100%)*	76 (54%)	
Diag. marker	33 (97%)*	65 (46%)	
CMAP (mV)	Low: 30 (88%)*	Normal: 137 (98%)	4.8
LRS:	33 (97%)*	63 (45%)	-6.8%
2 Hz	28 (82%)	47 (34%)	-6.8%
3 Hz	30/32 (94%)	48 (34%)	-6.7%
5 Hz	26/32 (81%)	49 (35%)	-5.0%
PTE:		2 (1.4%)	-11.1%
50 Hz (%)	Increment: 33 (97%) ≥ 60% increment: 32 (94%)	Decrement: 36 (26%)	+43% LEMS -19% MG
PEF: increment	26/31 (84%) ≥ 60% increment: 21/31 (68%)		+37% LEMS
HRS or PEF: increment	≥ 60% increment: 33 (97%) †		

* P value: <0.0001 vs MG † Diagnostic marker for LEMS Diagnostic marker for MG

Conclusion:

1. Distinguishing features of the RNS test in LEMS and MG are confirmed in this direct comparison study:
- the low CMAP amplitude, decrement at LRS, and increment at HRS or PEF for LEMS and normal CMAP amplitude and decrement at LRS for MG.
- Distinguishing features of the RNS test are most prominent between normal and LEMS and less prominent between normal and MG.

3. In MG, RNS test in proximal muscle are essential to achieve the meaningful diagnostic sensitivity: 75% in RNS test

Guillain-Barré syndrome: current concepts and perspectives

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Conduction block can be produced by either demyelination or axonal dysfunction, if the safety factor for impulse transmission is critically impaired. Guillain-Barré syndrome (GBS) is classified into the two major categories; acute inflammatory demyelinating polyneuropathy (AIDP) and acute motor axonal neuropathy (AMAN), presenting demyelinating and axonal conduction block respectively.

Over the past 20 years, major advances have been made in understanding the immunopathogenesis and pathophysiology of AMAN. AMAN is characterized by pure motor involvement, antecedent Campylobacter jejuni enteritis, and serum anti-ganglioside antibodies. Compared with AIDP, AMAN has more rapid progression and earlier nadir, and two patterns of recovery (rapid improvement by resolution of conduction blocks, and slow recovery by axonal degeneration). Electrophysiological studies frequently reveal, as well as axonal degeneration, rapidly reversible nerve conduction block/slowing (reversible conduction failure); the time-course suggests functional or microstructural changes at the nodes and paranodes, termed as “nodopathy”. It is now established that AMAN is caused by molecular mimicry of human gangliosides by the bacterial lipo-oligosaccharide. An animal model of AMAN immunized by GM1 was developed, and its histological studies show disruption of nodal sodium channel clusters, and paranodal myelin detachment; these changes are likely to account for conduction block in human AMAN.

In therapeutic aspects, complement inhibition is a promising treatment for severe GBS. A recent clinical trial of eculizumab, a monoclonal antibody against complement C5, showed better recovery in the eculizumab group than in the placebo group. The mechanisms for axonal or demyelinating conduction block due to complement activation are discussed.

Waveform Analysis for Electromyographic Studies

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Electromyographic studies analyze spontaneous and voluntarily activated muscle action potentials. Following brief injury potentials coincident with the insertion of the needle, a relaxed muscle remains electrically silent except for the endplate activities, which consist of end-plate noise and associated spikes. Denervated muscle fibers fire at rest independent of the volitional neural control, which provides one of the most distinct abnormalities of the needle study. Several types of spontaneous discharges, though often nonspecific as to a disease entity, all signal disorders of the nerve or muscle.

Both fibrillation potentials and positive sharp waves represent spontaneous excitation of individual muscle fibers. The complex repetitive discharges comprise high-frequency spikes derived from multiple single muscle fibers, which discharge sequentially maintaining a fixed order. Fasciculation potentials represent spontaneous discharges of a single motor unit. Myokymic discharges relate to intermittent bursts of 30 to 50 Hz repetitive firing of a single motor unit, or grouped fasciculation potentials. Neuromyotonia derives its name from continuous firing of a motor nerve axon at 200 to 300 Hz, causing high pitched sounds resembling myotonia.

Isolated discharges of single motor unit give rise to motor unit potentials, the smallest functional element of volitional contraction, as defined by Sherrington. Diseases of the nerve or muscle cause its structural or functional disturbances leading to alterations in its waveform and discharge patterns; a late recruitment and rapid firing of large units for neurogenic lesions in contrast to an early recruitment of small units in myopathies. The study of motor unit potentials helps categorize motor abnormalities associated with clinical weakness into upper and lower motor neuron disorders and myogenic lesions.

As a clinical tool, electromyography and nerve conduction study work well only if the examiner conducts the procedure in the light of the patient's history and physical examination. Thus, the electrodiagnostic studies constitute an extension of clinical assessments rather than an independent laboratory test. As such, it serves best if performed by a physician thoroughly familiar with the patient’s clinical findings with the aim to prove or disprove the diagnostic impression. Although waveform analysis and pattern recognition of discharging units form the basis of electromyographic studies, one must interpret the results in the context of the clinical findings.

Intraoperative neurophysiological monitoring in Korea: Current status & its role in Asian-Oceanian region

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History & background

The first use of IONM dates back to mid-1990s in Korea. IONM practices have been carried out mainly at university affiliated hospitals since then.

Two IONM societies were organized in 2013 and 2014 respectively. One is Korean Society of Intraoperative Neurophysiological Monitoring (KSION), originally a research group of Korean Society of Clinical Neurophysiology (KSCN), and the other is Korean Society of Intraoperative Neuromonitoring, a research group of Korean Neurosurgical Society. Korean national medical insurance policy only reimburses IONM tests which are supervised and read by neurologist or physiatrist. It recognizes that clinical neurophysiology needed for IONM is included only in these specialists' formal educational and training curriculums. And this is why these doctors are playing a main role in this field in Korea.

Current status

The number of IONM practices has increased substantially, so most of university-affiliated hospitals use them on regular basis. The 2 IONM societies organize their own annual meetings, which pursue an education and training of doctors and technologists involved in this field. Also KSION hosted a successful 6th International Society of Intraoperative Neurophysiology (ISIN) Congress & Educational Course 2017 in Seoul under the auspices of KSCN, which was the first meeting of ISIN in Asian-Oceanian region.

Future perspectives

IONM practices have been increased mainly in quantity for decades in Korea. So devoted neurophysiologists and associated societies need to focus more on a promotion of IONM quality throughout the nation for a patient safety. Also, International contribution for an education and development of IONM in Asian-Oceanian region in which large number of countries are still in their beginning stages would be essential.

History and Perspective of EEG Science

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In 1875, Richard Caton from Liverpool, England first discovered the electrical activity recorded from the animal brain. Fifteen years later, his discovery was confirmed by Fleischl von Marxow rom Vienna, Austria and Adolf Beck from Krakow, Poland. After Richard Caton's first description of animal brain electrical activity, it took more than 50 years before the electrical activity from human brain was described by Hans Berger from Jena, Germany. The title of his first publication in 1929 was entitled as "Uber das Elektrenkephalogram des Menschen" (About Human Electroencephalography). He named about 10 Hz activity as "alpha rhythm "recorded during quiet wakefulness with eques closing. However, his discovery was initially viewed with skepticism. But in 1934 Adrian and Matthews from London approved Berger's work and called the EEG waves as "Berger rhythm". This has brought surge of interest in EEG throughout the world.

In 1936, Gibbs, Davis and Lennox (USA) discovered that 3 Hz spike-wave bursts are diagnostic pattern for absence (petit mal) seizures. This had triggered the EEG to be import and non-invasive diagnostic tool for various brain diseases, especially for epilepsy.

Recent advances of computer technology have brought a new era of EEG utilization, which allows us to record EEG continuously at bed side, especially at ICU setting(ccEEG). Because EEG reflects dynamically changing brain function sensitively and instantaneously, for which no other functional tests (PET, SECT, fMRI) can compete with. This lead to the recognition of non-convulsive seizure or non-convulsive status epileptics in acutely ill patients with impaired consciousness. Overall the incidence of non-convulsive seizures in such patients reported to be about 20-40 %, which could never be found without ccEEG. The indication of long term ccEEG includes post convulsive seizure, traumatic brain injury, post brain surgery, acute ischemic/hemorrhagic stroke, encephalitis and any unexplainable mental status change. The ccEEG can also be used for therapeutic assessment and prognostication of acute cerebral dysfunction.

Other capabilities of digital EEG include qualification and quantification of EEG data, ability to change recording parameters as of off-line and remote accessibility to EEG recordings. With further advancement of computer technology EEG could become "Window of Mind" as Hans Berger dreamed.

Berger's dream: How did EEG become the EEG?

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There were three pivotal events in the earliest stage of the human EEG history. First, Hans Berger first succeeded in recording human EEG in 1924. Second, he published the first EEG paper five years later (1929). Finally, after another five years of silence, Edgar Adrian, a Nobel Prize winner in physiology and medicine for the discovery of the all-or-none law of nerves, confirmed Berger's EEG result (1934). After that, EEG rapidly spread across the world as a physiological and neurological research tool of the brain.

Two questions have arisen about the above EEG history. Why did it take five years for Berger to publish the first EEG paper? What was going on in the next five years? In my talk, I would like to answer these questions by illustrating the following issues:

・Berger's EEG traces recorded with primitive EEG machines
・Adrian's EEG demonstration experiment at Cambridge (1934) and the subsequent gentlemanlike supports for Berger.

We will learn much about what is important in science from Berger's and Adrian's attitude to the discovery of EEG even today.

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A historical perspective on clinical EEG in Japan

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In 1935, Kazumi Yamagiwa, later appointed to prof. of Physiology at the Tokyo Medical and Dental Univ., wrote a paper on "the origin of the Berger rhythm" at the Adrian's Lab. in Cambridge Univ. Although he was the first Japanese researcher contributing to early human EEG study, he could not continue EEG study in Japan because of the outbreak of the World War II in 1941.

During the World War II, Koichi Motokawa, prof. of Physiology at the Tohoku Univ., and Seizo Katsumura, prof. of Internal Medicine at the Nagoya Univ., had emphasized the importance of EEG research, and the Japan Society for Promotion of Science organized the first official EEG committee in 1942. At that time, EEG was recorded by handmade string galvanometer and developed into photographic paper. Also in 1942, Satoru Kuwabara, a neurosurgeon wrote the first EEG textbook in Japanese titled "No-denki-zu" supervised by Kikuo Ohtsuki, prof. of Neurosurgery at the Tokyo Univ. Thereafter, Prof. Katsumura proposed the Japanese word "Nouha", which was widely used as a translation of “electroencephalogram”.

After the World War II, the Japanese Ministry of Education formed the national EEG committee in 1946, and prof. Koichi Motokawa again served as a chairman. The members were consisted from seven National University of Medicine and the official report was published in 1947. The reported topics were the construction of EEG amplifier using vacuum tubes and the period analysis of EEG waves from the Tokyo Univ., the EEGs of brain tumors from the Kyoto Univ., the EEGs of myoclonic epilepsies from the Hokkaido Univ., etc.

In 1950, a sub-committee of the national EEG committee standardized an ink-written recording device, and Hideo Itokawa, prof. of Institute of Industrial Science of the Tokyo Univ., produced a prototype of EEG machine. In 1951, a Japanese company "Sansei-Denki", under the guidance of the Sakamoto Lab., dept. of Technology at the Tokyo Univ., marketed a 2-channel ink-written EEG machine named "Mokusei-go", which was first delivered into the Psychology Lab. of the Nihon Univ. In 1953, the Rockefeller Foundation presented two Grass 8-channel EEG machines and delivered into dept. of Neurosurgery at the Tokyo Univ. and dept. of Psychiatry at the Nagoya Univ. Thereafter, domestic inkwritten EEG machines were on the market and widely distributed in Japan. During the years since then, the neurophysiology markedly flourished in Japan.

New developments in quantitative EEG analysis

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Less than a century has passed since Hans Berger’s first report in 1929 about Electroencephalography (EEG) with a human being. Even now, EEG is still one of the principal methods for extracting information on human brain function for research and clinical purposes. EEG has both advantages and disadvantages. Advantages are minimal invasion, low cost, high temporal resolution and convenience. On the other hand, disadvantages are limited spatial resolution and difficulty in inspection. In the early days of EEG, inspection of EEG was performed by only visual observation. However, visual observation is subjective because it is influenced by observer’s experience and ability. Accordingly, quantitative EEG (qEEG) analysis has developed accompanied by progress of computer technology from the 1960s. qEEG is a procedure that processes the digitized EEG signal which was converted from analog EEG signal from multi-electrode using various algorithms. The derived qEEG information can be interpreted as parameters of brain function. The beginning of qEEG approach mainly based on frequency domain analysis using multi-electrodes. Recently, many novel analytic approaches have appeared such as LORETA (low resolution brain electromagnetic tomography) which estimates the electric neural activity distribution upon Talairach space using inverse solutions and EEG microstate analysis which elucidates the minimal spatio-temporal components of the EEG, and so on. In addition, brain functional connectivity, lagged coherence and lagged phase synchronization can be estimated from cortical electric neural activity which is obtained using LORETA from scalp electrodes, too. In recent times, the predominance of EEG might have decreased because of the development of other neuroimaging methods. Nonetheless, EEG still has solid superiority over other neuroimaging methods by its fundamental characteristics and newly developed methods of analysis. Especially, safety, economy and high temporal resolution of EEG can’t be replaced by other modalities. EEG has great potential as a tool for further investigating brain function from a multidisciplinary manner. In this presentation, I would like to talk about some new qEEG analysis methods, principally LORETA, and applications mainly in psychiatry.

Clinical EEG in 21st century: a research topic or a tool ?

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Clinical EEG represents the dynamic change of mainly EPSPs arising from the pyramidal neurons in the cerebral cortices, and it consists of oscillatory wave forms such as delta, theta, alpha, beta and gamma frequencies. Since its fundamental generator mechanisms were elucidated and since various pathological situation such seizure, focal abnormality, coma and brain death have been well correlated with each EEG findings, EEG is already an established tool for clinical practice and clinical research in analogue EEG era in 20th century.
Since 1990s digital EEG has been rapidly introduced and almost replaced analogue EEG, and currently furthermore wide-band EEG is clinically available. It ranges from infraslow or DC shifts (since 1990s) to high frequency oscillation (HFO) such as over 300Hz or even 1kHz (since 2000s) depending on the degree of sampling rate. Therefore, currently clinical EEG may belong to the 2nd generation of digital EEG as “wide-band EEG era”. Therefore it is very important to differentiate those digital EEG features as to whether they are already established tools or still research topics.
1) Research topics can be divided into biological research topics. It includes infraslow or DC shifts. Ictal infraslow or DC shifts has been extensively investigated as the research topics in basic and clinical fields, and at least a part of them (invasive ictal DC shifts) is very close to a tool (Nakatani et al, 2018). Among pathological HFO, interictal, invasive HFO is at least partly regarded as a tool. Physiological HFO such as language and memory is actively investigated as the research topics.
2) Mathematical EEG data analysis with clear hypothesis or condition is also regarded as methodological research topics (3D source analysis, Granger causality analysis, etc.).
3) Clinical tools are also listed as follows; quantitative EEG analysis such as frequency band mapping, time-frequency analysis and voltage map. Those are strong tools for clinical practice and for clinical research for CNS function and its pathological state.

In the clinical filed, we could often apply novel EEG technology as the research topic in the clinical situation, and then often and usually immediately, the outcome, feedback or the degree of clinical correlation is obtained. Therefore, nevertheless, we should be cautious not to obscure the boundary between research topic and clinical tool until it is at least partly established appropriately.

Automatic Integrated Interpretation and Reporting of the Adult Waking EEG

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Automatic interpretation of EEG has faced significant difficulties because of a large amount of spatial and temporal information contained in the EEG, continuous fluctuation of the background activity, occurrence of paroxysmal activities, contamination with various artifacts, and use of different electrode montages. Therefore, the previous attempts of automatic EEG interpretation have focused on a certain feature of EEG such as the background activity and paroxysmal abnormalities. The primary purpose of this study is to establish a computer-assisted, off-line system for automatic integrated interpretation of EEG, which takes into account all features of the adult waking EEG. First, we extracted features of EEGer’s visual inspection to quantitatively score the background activity, and the parameters for each frequency band were determined. Automatic detection of artifacts and spikes, and automatic judgment of vigilance and attention level of subjects were incorporated into the system. This system automatically provides the results in a written form either in English or in Japanese soon after completion of actual recording. This system can be used as a supplementary tool for EEGer’s visual inspection and save the time for writing the report, and its modified form can be applied for training EEGers and EEG technicians.

P2-06-01

Early mild cognitive impairment identification using support vector machine-based analysis of resting-state functional connectivity

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Mild cognitive impairment (MCI) is the prodromal stage of Alzheimer's disease (AD). Despite the fact that there is consensus that diagnosis and intervention for early stage MCI (eMCI) are urgently needed, it is difficult to diagnose because the cognitive impairment is very mild or insignificant. Resting-state functional connectivity (rsFC) provides complementary information that can be used to enhance our understanding of brain cognitive function decline from normal aging and brain disorders. In the current study, we aimed to apply machine learning techniques such as Support Vector Machine (SVM) in rsFC to classify eMCI and a healthy control (HC). A rsFC matrix was computed for all participants (eMCI 148, HC 249). This matrix acted as the source for the initial classification between eMCI and healthy controls. Then a Least Absolute Shrinkage and Selection Operation (Lasso) was performed for feature shrinkage. The features corresponding to non-zero Lasso regression coefficients were retained as crucial features for classification. Finally, the classification was carried out using SVM with leave-one-out cross validation. We were able to reach an average of 71% accuracy when differentiating eMCI from the HC. The current study indicated that our approach has the potential of distinguishing between eMCI and HC.

P2-09-02

Comparison of intra- and extra-operative lateral spread responses in microvascular decompression surgeries of hemifacial spasm

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Background: The aim of this study was to compare the prognostic value of intra- and extra-operative lateral spread response (LSR) in microvascular decompression (MVD) surgeries of hemifacial spasm (HFS).
Methods: A retrospective review was made of consecutive 25 patients who underwent continuous intraoperative monitoring during MVD. Extraoperative LSRs were done pre- and postoperatively.
Results: In 19, the intraoperative LSRs disappeared during surgery. In 5, the intraoperative LSRs were absent before incision. In one, the intraoperative LSR intraoperatively persisted. For extra-operative LSRs, they disappeared after surgery in 10 but persisted in 6. LSRs were absent before and after surgery in 7. For 5 of 19 patients, intraoperative LSRs disappeared during surgery but extra-operative LSRs were persisted despite adequate MVD. In one of 5 patients, intraoperative LSRs were absent before incision and remained during the surgery. But extra-operative LSRs were present before surgery and disappeared after adequate MVD. Statistically, the extraoperative disappearance of LSR was correlated with the HFS relief in 4days after surgery and the 3-month follow up period. However, the intraoperative disappearance of LSR was not.
Conclusion: In our study, extra-operative LSR monitoring may be more predictive of the surgical outcome compared with intraoperative LSR during the 3-month follow up.

P2-11-01

Cortical silent period shows larger ending latency and duration than cutaneous silent period on abductor pollicis brevis muscle in healthy human subjects

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Background: The voluntary contraction of the abductor pollicis brevis (APB) muscle can be suppressed cortically with a suprathreshold TMS pulse (cortical silent period CSP) and by cutaneous noxious stimulus (cutaneous silent period CuSP). CSP measures the intracortical inhibition produced by the activation of the GABAA interneurons that synapse on pyramidal neurons. CuSP is mediated by A-delta afferents that inhibits the C7-T1 motoneurons postsynaptically, through an oligosynaptic spinal circuit. We aimed to characterize both CSP and CuSP in APB muscle.
Methods: In 6 healthy subjects, we assessed the APB silent period to cortical magnetic single pulse stimulation at 140%RMT applied on the contralateral APB motor primary cortex region while the participants performed submaximal isometric contraction. CuSP was recorded after 2nd finger dermatome electrical stimulation (10x sensory threshold) using ring electrodes.
Results: The CSP showed the average duration of 160 ms (SD 20) larger than CuSP duration of 44 ms (SD 4) with average CSP end latency of 180 ms in comparison with CuSP end latency of 124 ms.
Conclusions: The CSP shows larger end latency and duration in APB muscle compared to CuSP, with a duration ratio between CSP and CuSP of more than 3 to 1 (3.64).

P2-14-04

Comparison of Polyneuropathy in Levodopa-Treated Parkinson’s Disease Patients

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Background: Parkinson’s disease is the most common disease of neurodegeneration, which impacted quality of life of those patients. Lots of recent studies showed that another non-motor symptom including polyneuropathy, that not yet known the etiology. This study, we hypothesize that polyneuropathy might occur from the side effect of Levodopa.
Methods: We included 30 patients who have Parkinson’s disease. 15 were treated with Levodopa for more than 3 years and 15 patients who were treated 3 years or less. Toronto Clinical Neuropathy Scoring System (TCSS) was done to screen the patients. The eligible patients who TCSS more than 6, would be conducted a nerve conducting study test (NCS) for both Radial nerves and Sural nerves.
Result: This study is a preliminary for the whole research which will be finished in September, 2018.7 were enrolled, 2 were excluded. 5 patients passed TCSS and experienced the clinical of polyneuropathy but NCS of all patients were normal.
Conclusions: This preliminary of our research shown that patients of this study experienced the clinical of polyneuropathy from TCSS without an abnormal large fiber sensory conduction study. This may show the evidence of small fiber polyneuropathy which cannot be detected by performing a conventional NCS.

P2-19-01

Quantitative analysis of core muscle activities by surface EMG

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Background: Functions of core muscles are important for stable and safe execution of all movements. An objective quantitative assessment of core muscles has not been established. For limb muscles, muscle strength can be estimated using surface electromyogram (s-EMG), based on a positive linear correlation between the generated force and s-EMG activity. In this study, we examined whether this correlation also exists for core muscles.
Methods: Ten healthy volunteers aged 18-40 years were recruited. Surface-EMG were recorded from rectus abdominis and paraspinal muscles and analyzed quantitatively. Force generated by core muscles was measured by strain gauge connected to the back of a legless chair. Participants repeated consecutive isometric trunk flexions (ISM) at maximal and submaximal efforts. Surface-EMG during free movements without constraint were recorded in some participants.
Results: During ISM, the rectus abdominis functioned as the agonist, and a positive linear correlation was observed between force and magnitude of s-EMG. However, the relation was different during free movements.
Conclusion: A linear force-s-EMG relation was observed in rectus abdominis and paraspinal muscles during ISM, and muscle strength may be estimated from s-EMG activity. Because the s-EMG patterns during free movement differed from ISM, careful interpretation of data considering specific movements is needed.

P2-21-01

Sensory neuropathy associated with long term turmeric consumption

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Backgrounds :Sensory neuropathy from vitamin B6 toxicity was recognized since 1983. The lowest dose has been reported was 50 mg/day. High intakes of vitamin B6 from food and herb have not been reported to cause adverse effects. Turmeric is a herb that very rich source of pyridoxine. The data from United States Department of Agriculture National Nutrient data base indicates that turmeric 100 g contained pyridoxine 1.8 mg
Methods :This case report present A-76 year old woman present with progressive gait difficult for 3 year after turmeric consumption 1,000mg/day for 5 year. Her physical examination showed sensory ataxia ,impaired vibration and hyporeflexia both feet.
Results :Electrophysiological study showed evidence of severe distal sensory neuropathy on lower extremities. Laboratory investigation showed high level of vitamin B6 in serum.
Conclusions :After cessation turmeric consumption 2 months, we follow up physical examination the sensory conduction study. The sensory conduction study was improved from previous study. We also follow up the vitamin B6 level but still pending for the result.

P2-23-01

Myasthenia gravis associated with Charcot-Marie-Tooth neuropathy: report of two cases

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Objective: To analyze the clinical and electrophysiological characteristics of patients with Charcot-Marie-Tooth neuropathy and MG.
Method: We conducted a search of medical records at Peking Union Medical College Hospital for coexistence of Charcot-Marie-Tooth and MG, retrospectively analyzed their clinical manifestations together with the neurophysiological characteristics.
Results: We presented 2 patients in our database. Case 1: a 22-year-old male, inability to open eyes widely at 3-year-old. He had complained since childhood of general faintness related to physical exercise. AchRab (-). After treatment with pyridostigmine, the symptoms were alleviated and corticosteroid treatment was rejected. Case 2: a 47-year-old male with left eyelid ptosis and diplopia for 2 months, oral pyridostigmine was effective. The 17p12 region large fragment repeat variation includes the PMP22 gene. AchRab(+). Diplopia disappeared after treatment with corticosteroids. Their electrophysiological characteristics are similar : NCV showed peripheral nerve damage in the upper and lower extremities, and the motor and sensation were both involved. EMG showed chronic neurogenic damage. RNS has not seen diminishing or increasing phenomena.
Conclusion: Concurrence of MG and CMT1 is rare and the mechanism is unknown. Both of the patients were ocular muscle type, and RNS showed no declination of low frequency, and pyridostigmine was effective.

P2-23-02

A case of congenital myasthenia with false positive anti-AChR antibody

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Background: Anti-acetylcholine receptor (AChR) antibody is pathogenic and highly sensitive and specific for autoimmune myasthenia gravis (AMG). There were only few false positive results reported. We described a young child with positive anti-AChR antibodies on 2 separate sera treated as refractory AMG but revealed to have COLQ-mutant congenital myasthenic syndrome (CMS).
Case report: A 4-month-old boy with hypotonia and bilateral ptosis investigated for neuromuscular disorder. His anti-AChR antibody determined via ELISA (Euroimmun AG, Germany) at 6 months old was positive but his maternal titre was negative. Repetitive nerve stimulation and stimulated jitter with concentric needle confirmed a neuromuscular junction disorder. A provisional diagnosis of AMG was made. However, he responded poorly to pyridostigmine and immunotherapy requiring ventilatory support. A repeated anti-AChR antibody at 13 months old remained positive. In view of the unfavorable clinical progress and an early onset phenotype, the possibility of CMS was explored. Pyridostigmine treatment was then withheld, and salbutamol was introduced with a remarkable response at 14 months. CMS genetic panel showed a compound heterozygous COLQ gene mutation.
Conclusion: False positive anti-AChR antibody in CMS has never been reported in the literature. This highlights CMS should be considered in the early onset phenotype rather than AMG.

P2-23-10

Clinical and Electrophysiological study of 29 cases of Dermatomyositis

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Objectives: To analyze the clinical and electromyography of 29 patients with dermatomyositis, and to explore the diagnostic value and clinical significance of electromyography (EMG) in this disease.

Methods: Twenty-nine patients were retrospectively collected, the data including clinical and electromyographic characteristics.

Results: The first symptom statistics were as follows in the group: skin changes in 12 patients (41.4%), myasthenia in 4 patients (13.8%), rash companied with fever in 4 patients (13.8%), fever alone in 3 patients (10.3%), rash companied with myalgia in 2 patients (6.9%),and rash companied with myasthenia in 2 patients, and myalgia in 2 patients (6.9%). MUAP showed myogenic damage found in 18 patients (61.7%) and the probable muscular damage found in 4 patients (13.8%), the neurogenic damage found in 1 patient (3.4%), The overall abnormal rate of electromyography was in 23 patients (79.3%).

Conclusion: EMG showed that myogenic MUAP of dermatomyositis was characteristic and it's a valuable method for early diagnosis.

P2-24-01

Time From Onset to EMG Examination in ALS Patients in Indonesia: A Survey by ALS Indonesia Foundation

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Background. EMG is required for diagnosis of ALS. Several factors may contribute to diagnostic difficulties delaying treatment for optimizing patient's QoL. We want to investigate the time from onset to EMG examination of patients with ALS/PALS in the ALS Indonesia Foundation.

Methods. A survey was performed using structured questionnaires.

Results. There were 41 PALS registered from July 2015-2018 (27 males, 14 females) and 33 patients resided in Java Island. Adult-onset ALS (onset at the age of 40 years/above) was shown in 28 patients, 12 patients had young-onset ALS, 1 patient was diagnosed < 20 years old/juvenile-onset. In this survey, data was obtained from 31 PALS (20 males, 11 females). Time from onset to EMG examination was > 12 months in 16 PALS, 10 PALS required 6-12 months, < 6 months in 5 PALS. Multiple EMG were done in 16 PALS (2 PALS had > 3 times EMG examinations). There were 16 PALS who spent hours of travelling to the nearest EMG facility with 8 hours being the longest transport time.

Conclusions: The time from onset to EMG examination in the majority of the PALS was > 12 months. Difficulty of finding near-by EMG facility is described. Further study is needed to see other attributing factors.

P2-24-02

Vestibular evoked myogenic potentials (VEMPs) and clinical study in patients with amyotrophic lateral sclerosis

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Background The objective of this article is to evaluate the diagnostic value of vestibular evoked myogenic potentials (VEMPs) in the assessment of brainstem function integrity in patients with amyotrophic lateral sclerosis (ALS).
Methods: This was a prospective case-control study including 30 definite or probable ALS patients divided into two groups (with or without brainstem involvement) and 30 healthy controls. Cervical (c-), masseter (m-) and ocular VEMP (o-VEMP) measurements were performed in all the participants.
Results The c-VEMP mean p13 and n23 were significantly prolonged in the ALS patients. The interside peak differences in p13 and n23 of c-VEMP and in n10 and p15 of o-VEMP were significantly prolonged. The rates of alteration in c-VEMP, m-VEMP and o-VEMP in the ALS patients were 67%, 40%, and 45%, respectively. The ALS patients with brainstem involvement had a significantly higher percentage of VEMP abnormalities than the patients without brainstem involvement ($p = 0.027$).
Conclusions C-VEMP is a sensitive tool to detect lower brainstem involvement. Impairments in o-VEMP and m-VEMP indicate involvement of the upper brainstem. The use of combined VEMPs may provide useful insights into the pathophysiological mechanism of ALS

P2-24-03

Safety and Efficacy of Edaravone in Delaying Functional Decline in Amyotrophic Lateral Sclerosis: A Meta-Analysis

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Background: Novel therapies are being sought based from the molecular discoveries on the pathogenesis of neuronal cell death in ALS. This metaanalysis aims to determine the safety and efficacy of edaravone in delaying functional decline among patients with ALS.
Methods: A meta-analysis of randomized controlled trials was done which included patients 19 years old and above, diagnosed with ALS within 3 years based from the revised El Escorial criteria, and with no compromise respiratory functions.
Results: Three studies included had a total of 368 ALS patients. Patients were given either 60 mg of intravenous edaravone or a matching placebo. Incidence of both non-serious (RR1.00, CI 0.93,1.08) and serious adverse events (RR0.66, CI 0.39,1.12) are not statistically different between groups. Mortality also did not differ (RR1.79, CI 0.39,8.23). All studies are homogenous (I2=0%). Mean change in ALSFRS-R scores at 24 weeks is higher in the placebo group (MD1.6, CI 1.44,1.76) reflecting more functional deterioration. However, the three studies exhibited heterogeneity (I2=99%). Sensitivity analysis was done but still yielded substantial heterogeneity (I2=61%-94%).
Conclusion: The limited evidence shows that edaravone is safe to use and patients treated with edaravone has less functional decline at 24 weeks compared to those receiving standard of care.

P2-24-04

Does high voltage electrical injury cause spinal motor neuron disease?

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Background and Purpose: There are many reports suggesting that amyotrophic lateral sclerosis (ALS) is associated with prior electrical injury. These studies suggest that electrical injury may induce chronic cell death mechanisms such as autophagy. However, the existence of an association between ALS and electrical injury is controversial. Our objective was to examine the potential correlation of high voltage electrical injury with evidence of spinal motor neuron disease in human patients.

Methods: We reviewed electromyography data from patients admitted to our hospital for high voltage (22,900 V) electrical injury from 2009 to 2011. To avoid confounding factors due to direct effects of electrical muscle injury, only electromyography data from muscles proximal to the electrical injury site were used for analysis.

Results: A total of 346 patients were treated in our hospital for electrical injury during the specified period. Data from 68 extremities of 56 patients were selected. We found no evidence of acute and chronic denervation in proximal muscles.

Conclusion: We thus conclude that high voltage electrical injury does not promote spinal motor neuron disease in adjacent motor circuits.

CIDP – Diagnostic and Therapeutic Challenges

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Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a clinically heterogeneous, acquired immunemediated disorder characterized by progressive proximal and distal weakness, often accompanied by sensory deficits. CIDP is regarded as an autoimmune disease involving cellular and humoral immunity, although the precise targets of this immune response remain uncertain.

In clinical practice, heterogeneity of the variant forms has complicated accurate early diagnosis and choice of appropriate therapy. To improve the management of CIDP, diagnostic criteria have been proposed for both clinical and research purposes by consensus of experts from the EFNS/PNS. These include a combination of clinical, electrodiagnostic and laboratory parameters, and are reported to have high sensitivity (81%) and specificity (96%), with some recognized limitations.

Treatment is aimed at stopping the inflammatory response to prevent further demyelination and secondary axonal injury. First-line treatment options, namely corticosteroids, intravenous immunoglobulin and plasmapheresis are effective in 60-80% patients. Around 25% patients unresponsive to first line therapies will respond to an alternate immunosuppressive therapy, but no Level 1 evidence exists to guide choice of agent. There are no clearly defined predictive factors for response to therapy in CIDP nor biomarkers for disease activity. Some variant forms of CIDP have a different disease course and response to treatment.

This talk will outline the clinical CIDP phenotypes, review diagnostic criteria, and present the evidence related to treatment of CIDP. Therapeutic dilemmas including choice of first line therapy, administration paradigms, duration and monitoring of treatment and validated outcome scores, will be discussed.