ACNS Annual Meeting & Courses
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Speaker Abstracts

Presidential Lecture: History of the American Clinical Neurophysiology Society's Furthering of the Understanding of Status Epilepticus

Frank W. Drislane, MD

Soon after the founding of ACNS (1946), its leaders had made major advances in understanding status epilepticus (SE). First president, Herbert Jasper, helped elucidate the pathologic anatomy underlying seizures and studied EEG use. Gibbs and Davis helped establish the correlation between clinical seizures and EEG patterns. Fourth president Schwab detailed the electroclinical manifestations of "status epilepticus in petit mal." Niedermeyer (author, "the EEG bible" *Electroencephalography*<) described spike wave stupor. Related phenomena "triphasic" waves and PLEDs were initially described by ACNS presidents Bickford and Chatrian. Decades later, ACNS members are enriching our understanding of complicated EEG phenomena and describing more precisely their relationship to status. Modern methods enable SE's detection through imaging techniques, invasive physiologic recordings, etc. Currently, ACNS neurologists study SE's basic mechanisms and, in larger consortia, investigate how different forms of EEG monitoring can diagnose SE in critically ill patients; differentiate its many forms and determine their relative clinical importance; and evaluate and assist treatment. In 2014, SE, a fruitful area of investigation for generations of ACNS leaders, is a broadening field of intellectual achievement for members of the ACNS, an "ancient" but vigorous society.

2014 Robert S. Schwab Award & Lecture: The Excitable Axon

David Burke, MD, MSc

Studies of axonal excitability complement nerve conduction studies. The latter provide information about the number of conduction axons and the speed of conduction between the stimulating and recording sites. The former provide insight into the biophysical processes that underlie action potential generation at the stimulating site. In excitability studies, the stimulus is set to produce a submaximal potential, and the current is varied by computer to keep the size of the evoked potential constant. From these measurements, it may be possible to infer whether membrane potential and different voltage-dependent processes are normal, particularly if the recorded changes can be reproduced in a mathematical model of human axons.

The greatest value of these studies lies in understanding the mechanisms of disease in patients with generalised peripheral nerve or anterior horn cell disease. However changes have also been found in peripheral nerve axons in diseases of the central nervous system (genetic: EA1, EA2, BFNE, GEFS+ epilepsy, and acquired: stroke, multiple sclerosis and spinal cord injury). Here, the studies may well provide insight into the effects of a mutated ion channel and/or the adaptive changes that occur in motoneurons to compensate for the removal of inputs.

2013 Pierre Gloor Award & Lecture: Thalamocortical Dysrhythmia (TCD): Function and Dysfunction

Rodolfo Llinas, MD, PhD

The functional significance of the brain's thalamocortical activity will be discussed. Such activity will be addressed as the product of intrinsic electrical properties of neurons and the networks they weave. How this recurrent activity engenders motricity and cognition, as well as in the genesis of several neuropsychiatric ailments, will be discussed.

Intraoperative Neurophysiological Monitoring During Skull Base Surgeries

Facial Nerve and Auditory Nerve Monitoring during CP angle Tumor Removal

Aatif Husain, MD

Cerebellopontine angle (CPA) tumor surgeries can injure the facial and auditory nerves. Monitoring these cranial nerves during surgery is important to reduce the morbidity of the procedure. The facial nerve is monitored with needle electrodes in various facial muscles. Brainstem auditory evoked potentials are used to monitor in integrity of the auditory nerve. In this presentation, the methods and interpretation principles for facial and auditory nerve monitoring during CPA tumor surgery will be discussed. The value and shortcomings of monitoring will be highlighted.

Somatosensory Evoked Potentials and Arm Positioning Related Changes During Skull Base Surgeries

Ronald Emerson, MD

The utility of somatosensory evoked potential monitoring during skull base surgery will be discussed, with respect to both detection of patient malposition as well as surgical injury.

Pediatric EMG in the Molecular Era

Current Practice and Temporal Trends of Pediatric EMG in the New Millennium

Ioannis Karakis, MD, MSc

Extraordinary breakthroughs in genetics and imaging have generated questions about the sustainability of more traditional diagnostic modalities in neuromuscular evaluations, such as electromyography (EMG). This is particularly the case for children, where technical difficulties, interpretational challenges, and poor tolerability are more prominent. By analyzing data obtained from 2100 EMG studies performed over 11 years in a single tertiary referral center, this presentation offers important insights into current trends in referral and diagnostic patterns in the field of pediatric EMG. We conclude that EMG continues to play a pivotal role in the diagnosis of neuromuscular disorders in childhood, although its practice paradigm is shifting.

Chronic Inflammatory Demyelinating Polyneuropathies in Childhood

Hugh McMillan, MD, MSc, FRCPC, FAAN

Childhood chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a treatable autoimmune disease affecting peripheral nerves and nerve roots. Diagnostic criteria (clinical presentation, electrodiagnostic and biochemical test results) are used to diagnose CIDP and to differentiate it from a wide range of inherited and metabolic diseases. Although nerve biopsy is no longer an essential criteria for diagnosis of childhood CIDP it still has a role in select cases. There are an increasing number of cases that are being recognized where patients meet electrodiagnostic criteria for CIDP only to be recognized at the time to have nerve biopsy and/or genetic evidence of an inherited polyneuropathy. This observation raises concern that some treatment-refractory cases of CIDP could in fact represent other disease entities. This presentation will: 1) briefly summarize the clinical presentation of childhood CIDP including major differences between the disease presentation in children and adults; 2) review the diagnostic criteria of childhood CIDP; 3) review treatment efficacy and long-term outcome of childhood CIDP and; 4) review challenging cases where inflammatory and genetic overlap has been described.

Myotonia in Childhood

Peter Kang, MD

Clinical or electrophysiologic myotonia is a distinctive phenomenon at any age, including childhood and adolescence. Clinical myotonia can often be detected in school age children and adolescents. It may be more difficult to observe in infants, but some newborns with channelopathies such as paramyotonia congenita may display eyelid myotonia when they sneeze. In children and adolescents, electrophysiologic myotonia has been associated with channelopathies, myotonic dystrophy, congenital myopathies, and glycogen storage diseases. In addition, the presence or absence of myopathic motor units on needle examination can help distinguish further among these disease categories. Channelopathies such as myotonia congenita, paramyotonia congenita, and hyperkalemic periodic paralysis tend not to be associated with myopathic motor units. Myopathic motor units may or may not be seen in cases of myotonic dystrophy, congenital myopathies, and glycogen storage disorders. The needle examination is sometimes less comprehensive in children compared to adults, thus when electrophysiologic myotonia is not found, such results should be interpreted in light of the number of muscles examined and the degree of clinical suspicion. Thus, it is worth examining pediatric patients for clinical and electrophysiologic myotonia in the proper clinical contexts, as this finding can be very helpful in the diagnostic evaluation when detected.

Pediatric Neuromuscular Junction Disorders and the use of Stimulated SFEMG

Matthew Pitt, MD, FRCP

This presentation begins with a discussion of the different techniques available for analysis of the neuromuscular junction. It passes from nerve conduction studies and routine EMG to repetitive nerve stimulation before focusing on single fibre electromyography (SFEMG) and in particular stimulation SFEMG. The technique is demonstrated. There are problems with the analysis of the results, in particular the selection of potentials, which can be used. With the difficulties of identifying suitable potentials, a proposal is put forward that distinction between normal and abnormal responses is perhaps all that can accurately be achieved. Specificity is another problem that is considered. In contrast to the situation in adult patients where the differential diagnosis for myasthenia is relatively restricted the situation in children is much less easy with a wide range of clinical presentations being observed in myasthenic syndromes. To this end an investigative strategy using stimulation single fibre EMG is described, which allows differentiation between the common conditions entering the differential diagnosis such as bulbar palsy as well as myopathy with disordered neuromuscular junction function. Finally it is recommended that the technique should be called stimulation jitter analysis with concentric needle electrodes or StimJACE.

Wide Bandwidth Electrophysiology and Epilepsy Biomarkers

International Electrophysiology Database and Collaborative Research

Brian Litt, MD

The International Electrophysiology Database (http://ieeg.org) is a free, cloud computing-based resource sponsored by the NIH/ NINDS. "The Portal" can be run from any web browser, it contains easy to use tools for viewing, annotating, analyzing and selectively sharing data, including multi-scale time series, imaging and meta-data. The portal is set up to share actual algorithms and analysis code. Data can be selected and streamed to any computer for analysis from the portal, or distributed on Amazon's Elastic Computing Cloud (EC2), for big analysis jobs. Each experiment is given a unique identifier linking data with processing algorithms that allows users to repeat and validate any experiment once it has been "published." The portal makes multi-center studies, data collection and analysis extremely easy, as well as sharing educational materials such as teaching sets of EEG, evoked potentials, ICU monitoring, device recordings and intracranial data. The portal allows users to establish consensus/ gold standard training and testing sets for evaluating software to interpret human and animal electrophysiology. In this lecture I will familiarize the audience the International Electrophysiology Portal, its capabilities and discuss the role of "Big Neural Data" in Translational and Basic neurophysiology research.

Origin of Pathological and Physiological Oscillations

William Stacey, MD PhD

For over a decade, researchers have evaluated the role of High Frequency Oscillations (HFOs) in the brain. Originally described in normal activity, HFOs were later found to have a strong correlation to epileptic tissue, and subsequent efforts have concentrated on the characteristics of this relationship. The origin of these oscillations is a complex problem, as there is evidence for a number of independent network processes. More importantly, there is great interest in identifying a difference between 'normal' versus 'abnormal' HFOs. Early work suggested stratifying by peak frequency, as HFOs > 250 Hz appeared to be more specific to epilepsy. However, more recent work has shown that the distinction is much more complicated. This talk will discuss the current theories on the generation of physiological and pathological HFOs, as well as the challenges we face and potential benefits of using HFOs as a biomarker of epilepsy.

Botulinum Toxin: Mechanism of Action and Ultrasound vs. EMG Guidance

The Clinical Neuropharmacology and Neurophysiology of Botulinum Toxin

Francis Walker, MD

The basic pharmacology of botulinum neurotoxins is well delineated in that the toxins prevent the release of synaptic acetycholine from terminal nerve endings at the neuromuscular junction. The drug interferes with the action of docking proteins on the neuronal membrane that allow the synaptic vesicle to fuse with the membrane and release their content. The clinical neurophysiology and pharmacology of the drug relates to its time course of action following intramuscular injections, the degree to which it spreads or is absorbed into the bloodstream, and its duration of action. Currently available toxins, although dosed differently in terms of units, seems to have similar biologic effects at equipotent doses. Studies with injections into small muscles show that the vast majority of the drug is active at the injected site, that diffusion is generally robust within a muscle, and that the reduction in compound muscle action potential amplitude that follows injections seems to begin before and last long after clinical effects are appreciated in patients with dystonia and spasticity. The cause of these differences remain unclear. Optimal use of these agents is likely enhanced by clear understanding of their biological properties.

Ultrasound Guidance of Botulinum Toxin Therapy

Katharine Alter, MD

Botulinum Toxins (BoNT) are exotoxins of CLostridium Botulinum, an obligate anerobic bacterim. BoNT are known to be highly toxic but in minute quantities can provide significant relief to patients with a wide variety of clinical problems. the pharmacology of BoNTs and the principles of chemodernvation therapy will be reviewed. When performing chemodenervation procedures, accurately targeting the structures to be injected is important for efficacy as well as to reduce potential risks of the procedures. Conventional guidance techniques including palaption/anatomic landmarks, EMG and Electrical stimulation have recognized limitations. B-mode ultrsound is increasing recognized as a more accurate imaging technique. This presentation will review ultrasound technology and scanning techniques required for performing ultrasound guided chemodenervation procedures. A review of ultrasound imaging of muscles commonly targeted for botulinum toxin injections or nerve blocks will also be presented.

Ischemia Monitoring in Critical Care: EEG Trend Analysis to Detect Development of and Recovery From Cerebral Ischemia

cEEG Monitoring in Subarachnoid Hemorrhage: Current Practices and Limitations

Nicolas Gaspard, MD, PhD

Several studies have demonstrated the feasibility and utility of CEEG monitoring for the detection of delayed cerebral ischemia. Many obstacles however prevent its application in routine practice. During our presentation, we will discuss new methods to automatize

quantitative analysis, account for confounders and identify false alarms and ultimately develop a robust continuous real-time monitoring solution.

Evolution of EEG Patterns after Global Cerebral Ischemia

Michael Van Putten, MD

In patients with postanoxic encephalopathy a rich phenomenology of EEG patterns is observed, ranging from iso-electric recordings, burst-suppression patterns or generalized periodic discharges to diffuse slowing, reflecting the extent of initial hypoxic injury and neural repair. These processes are well captured by continuous EEG monitoring, that typically evolve over time and have a strong predictive value for the neurological outcome. I will shortly review the differential sensitivity of neuronal processes and network activity to oxygen/glucose deprivation, and present examples of the evolution of EEG patterns, and their correlation with predicting poor and good outcome, based on our prospective cohort of postanoxic patients (n>150) treated in the ICU after cardiac arrest.

Emerging Methods for cEEG Ischemia Detection in Subarachnoid Hemorrhage

Eric Rosenthal, MD

Continuous EEG monitoring (cEEG) in the intensive care unit has emerged as a tool for detecting delayed ischemic neurologic decline (DIND) following subarachnoid hemorrhage. Its use as a continuous, noninvasive, and multiregional biomarker of end-tissue health is complementary to other tools focusing on uni-regional, temporally discrete, or inferred measures of blood flow. However, clinical practice in real time is labor-intensive, and the benefit of cEEG-guided management on clinical outcomes remains unexplored. Prospective clinical experience with SAH ischemia monitoring will be detailed; including 1) the impact of seizure and DIND rates on reading and reporting standards, 2) variations in practice among institutions; and 3) reasonable expectations of accuracy in practice compared with other methodologies, including timeliness, authority, and novelty. A proposed clinical guideline for the use of cEEG monitoring and reporting in clinical practice will be discussed as well as requirements for clinical trials assessing its benefit.

The EEG of Cerebral Ischemia

Brandon Foreman, MD and Jan Claassen, MD, PhD

cEEG for seizure detection is well established; the use of cEEG to detect early ischemic changes has a solid basis in physiology, but has been underutilized in clinical practice. The EEG signal normally records voltage oscillations that reflect neocortical circuits in action. This circuit functioning, and therefore the EEG, are tightly coupled to cerebral blood flow (CBF) and energy metabolism. Decreases in CBF produce predictable changes in EEG background rhythms, from the loss of fast oscillations to the emergence of high amplitude irregular or rhythmic slow oscillations, and ultimately to voltage attenuation and background suppression. Quantitative EEG (qEEG) measures have been shown to objectively capture these nearly instantaneous changes to the EEG in response to changes in CBF; several studies have demonstrated the use of qEEG parameters in detecting the slower ischemic changes associated with delayed cerebral ischemia after subarachnoid hemorrhage. We will review the pathophysiological processes that create changes to cEEG/qEEG during ischemia and discuss important groundwork that has paved the way for current efforts in using qEEG to detect ischemic injury before it becomes irreversible.

The Creation of Evidence Based Medicine in Intraoperative Monitoring

Grading of Evidence and the Creation of Trials

David Gloss, MD, MPH&TM

In 2011, the Institute of Medicine published new standards for Clinical Practice Guidelines. The Guideline Development Subcommittee of the American Academy of Neurology (AAN) accepted these standards, and developed a new process for creating AAN guidelines. Previously, it was a two step process. 1. Creation of a systematic review based solely on AAN grading risk of bias in papers. 2. Development of recommendations based on the strength of evidence. This process has put intraoperative monitoring in a difficult position, since by AAN grading, most papers in intraoperative monitoring are considered low quality. The new process has four basic steps. 1. Slightly modified systematic review, 2. outcomes are abstracted from the papers, and a AAN-specific modified GRADE process is performed on them, 3. A modified Delphi process is performed on the resulting outcomes, looking at other factors such as variation in preferences, financial burden, magnitude of harm and beneft. With this new process in place, it may be time to propose a new guideline, using this new process. A new guideline may allow for more comprehensive community standards.

The Level of Evidence in Intraoperative Monitoring

Jay Varma, MD

In the first portion of the session, we review the studies used to create the evidence-based guideline for intraoperative spinal monitoring with somatosensory and transcranial electrical motor evoked potentials which was updated last year by the American Academy of Neurology. This will serve as the background for the subsequent talks which cover the methods by which trials are created and the creation of community standards and guidelines.

Creation of Intraoperative Monitoring Guidelines

Marc Nuwer, MD, PhD

Creating formal assessments involves several steps. A Panel of Experts devise one or several specific research questions. A literature search is based on relevant terms. Abstracts are screened for prospectively set inclusion-exclusion criteria. Literature is scored by pre-set criteria to determine evidence table data from all included studies. Studies' class of evidence are graded. The assessment's level of evidence is based on the studies' classes of evidence. The assessment's text addresses the methods, data found, conclusions drawn, and clinical relevance. The draft is reviewed by users, vendors, other specialties, and the sponsoring society's committees and board of directors. After an extended period for all comments, questions, and conflicts to be resolved, the Panel, committee(s) and society's board can endorse the new assessment. Setting the initial research questions well is a key to a good assessment. Problems include how to weigh animal studies, ethical issues about possible studies, heterogeneity of techniques and clinical applications, and lack of well designed class one studies. Recommendations are based on evidence, which can be constraining. Other comments can be considered in a discussion or commentary section, which may go beyond the evidence presented. Evidence-based Guidelines for clinicians are based on the outcomes of these assessments.

Advanced Practice Technologists in the New World of Continuous Neurophysiological Monitoring

Eduction, Knowledge, and Credentials for Advanced Practice Technologists

Joshua Ehrenberg, BS, R EEG T, CNIM

Over the course of many years, the field of neurophysiology has developed significantly. With the growth in LTVM, ICU cEEG, and intraoperative monitoring, there has been a need for advanced practice technlogists, and in many cases, these roles have been defined at the facility level. The ASET scope of practice has attempted to encompass this change. In instances where this has been implemented, and a differentiation has been made between procedural and advanced practice technologists, the qualifications have typically been subjective. There is a clear difference in some places between procedural technologists, who are at varying ranges of technical expertise, and advanced practice with clinical knowledge in non-traditional EEG areas). Due to the increasing need for this advanced practice role, and a clearer definition of procedural technologists versus advanced practice technologists scopes of practice, formal and standardized programs of education were needed. Emory University, in conjunction with staff at other facilities where advanced practice roles are used, developed formal educational routes to advanced practice and the qualifications necessary to enter. Current needs and usual practices will be discussed including proposed advanced practice requirements, the need for formal facility scope of practice differentiation, and the standardization of 'advanced practice fellowship' programs.

Advanced Practice in the OR

Brett Netherton, MS, FASNM, CNIM

As the neurodiagnostic specialty of IONM evolves, a correlate number of highly skilled IONM Technologists evolve with the field. The role of the Advanced IONM Technologist (A-IONM tech) is explored by presenting the results of questionnaire responses by multiple A-IONM Techs. The A-IONM Tech routinely utilizes many different skillsets, tools, and relationships with others on the IONM team to accomplish their scope of practice. The goal of this presentation is to highlight the levels of achievement of modern A-IONM Techs. What are the challenges in their scope of practice? How are their skillsets evolving to further benefit the IONM team? What new electrodiagnostic and non-electrodiagnostic technologies are becoming part of their scope of practice? What are the challenges in their daily practice from remote versus onsite professional oversight? Attention will be given to differences in experience noted between A-IONM Techs working daily in the same hospital setting compared to those A-IONM Techs practicing between multiple facilities and settings. Additionally, what can we learn from the A-IONM Tech regarding how to recruit and train more like them? And what improvements in the overall IONM team function can the A-IONM Tech identify to further the ultimate goal of improved patient outcomes?

Utilization of Advanced Practice in EEG Monitoring

Sherry Nehamkin, R. EEG/EP T., CNIM, CLTM

The EEG Technologist Reader is an evolving advanced practice role that will help fill a tremendous need in the rapidly growing field of electroneurodiagnostics, especially when considering the rapidly increasing demand for ICU video EEG monitoring.

We propose that when carefully designed protocols are in place, experienced, credentialed technologists with advanced practice training should have the opportunity to be actively involved in the ongoing evaluation of electroclinical studies.

Taking on this new role in the Comprehensive Epilepsy Center, the Technologist Reader is responsible for evaluating long term video EEG files, including monitoring real time activity, and preparing reports for the epileptologists, attending daily patient rounds with the epilepsy team, and teaching health care workers who take care of epilepsy patients and other patients requiring long term video EEG monitoring.

Spikes and Cognition: To Treat or not to Treat?

Electrophysiological Assessment of Frequent Spiking and Non-Convulsive SE Syndromes

Iván Sánchez Fernández, MD

The relationship between interictal epileptiform activity and cognition could be better studied if quantified. While we can measure the cognitive function with standard cognitive tests, and express it numerically, the same does not apply for interictal epileptiform activity. We have no standard method of quantification of the interictal epileptiform activity, which makes comparisons between different patients and different studies challenging. Several reasons make quantifying epileptiform activity difficult: spikes have different morphologies and voltages, they are more or less localized or generalized, they occur mainly during wakefulness or mainly during sleep; should they be quantified equally? A relatively straightforward method of quantification is counting the number of spikes in a given EEG tracing. This can be done as the percentage of seconds occupied by, at least, one spike or as the total number of spikes per unit of time. The latter method has no ceiling effect (can go over 100%) and therefore is able to distinguish between different degrees of severity in EEG tracings with many spikes. Future developments of spike quantification include the automatization of spike counting, and quantification of qualitative features such as voltage or distribution.

Relationship Between Spiking and Cognition

David Loring, PhD

Concerns that interictal spikes may negatively affect cognition have been described since the late 1930s, but they have often been considered primarily of academic interest and without relevance to everyday function. This presentation will highlight findings from relevant studies describing the relationship of interictal discharges to decreased cognitive functioning that extends beyond simple reaction time prolongation. Differences between generalized and focal discharges have been observed in multiple reports, with focal discharges appearing to have more local effects in disrupting cognitive performance. The relationship between discharges and cognition may also be bidirectional in which decreased discharge frequency is present during task performance, with increased discharge frequency during periods of rest. Evidence also suggests that the disruptive effects of interictal discharges extends to the hippocampus, with a negative effect on Sternberg task short term memory performance recently has been described. Interictal discharge effects likely contribute to "noise" in the clinical characterization of epilepsy surgery patients and their outcomes. In addition to differences in cognitive effects associated with generalized vs. focal discharges, initial evidence suggests potential differential benefit following successful treatment of interictal spikes, although conclusive evidence is currently lacking.

Should we Treat Spikes?

Kevin Chapman, MD

Interictal spikes have been associated with cognitive impairment. This presentation will discuss which patients with interictal spikes should be considered for treatment. We will also review the available data studying the effect of anticonvulsant medications on interictal spikes.

Spasticity - What is it and What is the Electrophysiology

Spasticity - An Historical and Clinical Conundrum

Morris Fisher, MD

Spasticity is one of the most commonly used terms to describe neurological abnormalities. Spasticity per se, however, has no clear definition or any definite anatomical or pathophysiological correlates. By frequently giving the sense of knowledge where none exists, the use of the term spasticity can hinder clinical evaluations and limit the potential for meaningful therapy. This symposium will discuss spasticity in its historical context as well specific investigational approaches, including electrophysiological, designed to better characterize patients with upper motor neuron dysfunction. The topic is important if one is to understand these patients as well as provide a basis for improved management.

Investigative Approaches to Characterizing Spasticity

W. Zev Rymer, MD, PhD

Recently, there has been an increase in our knowledge concerning ways in which spinal motoneurons become more excitable after lesions of the neuraxis. Central to this knowledge is the discovery that spinal motoneurons display "voltage-gated conductances" – active sodium and calcium conductances that are switched on by ongoing membrane depolarization. Most importantly, these conductances are sensitive to the presence of neuromodulators such as serotonin or norepinephrine; these are released by brainstem pathways. Disturbances of excitability, such as spasticity may be linked, in part, to disruptions of these voltage-gated conductances. In complete spinal cord injury, spinal motoneurons lose their dependence on ambient serotonin, and respond as if serotonin receptors are highly active, giving rise to exaggerated excitability - this is responsible for hyperactive stretch reflexes as well as spasms. In hemispheric brain injury, such as follows stroke, serotonin release may be increased, augmenting voltage gated conductances in spinal motoneurons, giving rise to exaggerated responses to muscle stretch. These various mechanisms can change reflex threshold by

depolarizing motoneurons –as detected by clinical tests such as the Tardieu. Changes in reflex gain may also be manifested in an exaggerated response to tendon tap. We will describe ways to separate these two contributing mechanisms

Ion Channel and F-Wave Findings in Patients with Storkes

Cliff Klein, PhD

Weakness, muscle hypertonia, and spasticity contribute to poor hand function after stroke. These impairments may be attributed in part to altered excitability and behavior of spinal motoneurons. Spasticity may reflect increased excitatory synaptic input to the motoneurons and/or an increased intrinsic motoneuron excitability. Regardless of the origin, spastic paretic motoneurons may reside in a more depolarized (excitatory) state (Katz and Rymer, 1989). Axonal ion channels, membrane pumps, and axonal geometric properties play pivotal roles in axon function, and these components are derived in part from synthetic activities of the motoneuron. Hence, it is reasonable to assume that peripheral axonal properties may change when their central source of impulse activity and metabolic support is disrupted. This talk will provide an introduction to nerve excitability testing using threshold tracking (Bostock et al. 1998) and present some data using this technique in people with chronic stroke. Nerve excitability testing can indirectly assess excitability properties (ie., ion channels, membrane potential) of peripheral nerve fibers in-vivo, and by extension the motoneurons. Hence, the method may provide important insights into the processes underlying neuronal plasticity associated with disease and injury.

Seizures, SUDEP and Autonomic Nervous System

Epilepsy and Autonomous Nervous System

Ekrem Kutluay, MD

Epilepsy is one of the most common neurological disorders. It is known that people with epilepsy have higher mortality rate than general population. Seizures, especially if they are poorly controlled, may have effects on autonomic nervous system. These effects can be in the form of cardiovascular, respiratory, sexual or gastrointestinal. However, cardiovascular and respiratory changes during epileptic seizures are “hot topic” now considering their possible role in SUDEP. Several studies revealed different forms of cardiovascular changes associated with seizures. Ictal tachycardia is probably the most common one reported in 80% or more seizures. Ictal bradycardia and asystole is significantly less common but consequences are more threatening. Brain regions involved in autonomic control include limbic structures (amygdala, orbitofrontal cortex, insula and cingulate), hypothalamus, periaqueductal gray matter and autonomic medullary centers. Also, some cortical stimulation studies revealed possible lateralized representations with bradycardia elicited by left insular stimulation and tachycardia by right. Seizures are also associated with respiratory depression and apnea. Studies revealed prolonged ictal apnea and significant drop in oxygen saturation even in patients whose seizures did not generalize. Neurophysiological tests also revealed altered autonomic regulation of the cardiovascular system in patients with epilepsy during interictal state.

Neurophysiological Testing of Autonomous Nervous System

Safwan Jaradeh, MD

The Autonomic Nervous System (ANS) is responsible for homeostasis. It is divided into central and peripheral components, also known as pre-ganglionic and post-ganglionic. It is responsible for the innervation of multiple organs. It has 3 major components: Parasympathetic, Sympathetic and Enteric. Cardiovagal function testing consists mainly of Heart rate response to deep breathing (HRDB) and the Valsalva ratio (VR). HRDB variability also forms the basis for high frequency power spectral analysis. Adrenergic function consists mainly of BP responses to the Valsalva maneuver and the BP responses to Head Upright Tilt. HRDB variability also forms the basis for low frequency power spectral analysis. Microneurography measures muscle sympathetic nerve activity. Sudomotor testing evaluates sympathetic cholinergic function. It consists of Sympathetic (galvanic) skin response (Electrodremalactivity), Thermoregulatory sweat test, Resting sweat output, Quantitative sudomotor axon reflex test and Silastic sweat imprint method. Autonomic testing is important in the evaluation of suspected Autonomic dysfunction, Small fiber neuropathies (painful), Syncope, Orthostatic intolerance (hypotension, tachycardia), Sweating disorders, GI or GU motility disorders, Extrapyramidal and cerebellar disorders, dizziness with dysautonomia, vasomotor rhinitis, GERD, and sleep apnea. The value of testing as it relates to epilepsy will be reviewed.

Amplitude-integrated EEG in Neonates: When is it Used and When is it Useful?

Principles of aEEG

James John Riviello, MD

Continuous EEG (CEEG) is an essential tool for ICU monitoring. Digital trending analysis (DTA) compliments CEEG, using mathematical signal transformations based on amplitude, frequency, or power, displayed on a compressed time scale. DTA is useful to visualize long-term trends, especially seizures, EEG background, and sleep-wake cycling. aEEG modifies EEG data, filtering frequencies less than 2Hz and greater than 15Hz, rectifying and smoothing the signal, has a linear display for 0-10μV and logarithmic display for 10-100μV), on a time-compressed display at 6cm/hour. The leads are placed over the central (C3, C4) or parietal (P3, P4)

regions. Frontal lead placement has more muscle artifact. Limitations include: limited electrodes, low amplitude or short duration seizures may be obscured by time compression; artifact contamination with "raw EEG" data needed to identify artifact; interpreter experience. aEEG machines now use dual channel recordings (C3,P3 and C4, P4), and display the "raw EEG" channel. CEEG is the gold standard for neonatal seizure detection and quantification and should be used whenever available. aEEG may be a useful complimentary tool when CEEG is not available, but if seizures are suspected on aEEG, CEEG should begin as soon as possible to confirm the diagnosis.

Interpretation of aEEG
Courtney Wusthoff, MD
Dr. Wusthoff will provide an introduction/refresher overview of how to interpret amplitude-integrated EEG. A tutorial will be given on the two most widely used background scoring systems for aEEG in term neonates. Patterns suggestive of seizures will be presented. Unique findings in preterms and special populations will be considered. These principles will be applied to example cases, leading to discussion of how neonatologists interpret aEEG in practice.

Current Applications of aEEG in the NICU
Tammy Tsuchida, MD PhD
Amplitude integrated EEG (aEEG) is being used more frequently in Neonatal Intensive Care Units (NICUs). aEEG enables the NICU to detect subclinical seizures, so that neonatal seizures can be identified and appropriately managed. In addition, there is literature to support the use of aEEG as a tool for prognosis after hypoxic ischemic injury. However, there are limitations to aEEG. In addition, the type of aEEG recording can affect the sensitivity of aEEG for seizures and background abnormalities. This talk will present the strengths and weaknesses of aEEG and optimal recording parameters.

Impact of aEEG Use in the NICU
Cecil Hahn, MD, MPH
Amplitude integrated EEG (aEEG) monitoring of newborn infants, originally pioneered in Europe, has now become commonplace in neontatal intensive care units (NICUs) throughout North America. In most NICUs, aEEG monitoring is applied by bedside caregivers and interpreted by neonatologists, with varying input from clinical neurophysiologists. This presentation will review the available evidence on the impact of aEEG use on clinical practice in the NICU, including the diagnosis of seizures, the use of antiepileptic drugs, the use of neuroimaging, and the frequency of neurology consultations. I will discuss how the availability of aEEG use has influenced the pattern of conventional EEG use, and reflect on the complementary roles of aEEG and continuous EEG monitoring.

EEG as a Basic Neuroscience and Psychology Research Tool

ERPs from Basic to Clinical Research: Toward Biomarkers
Greg Hajcak, PhD
This talk will focus on early electrocortical activity that has been shown to index response monitoring: the error-related negativity (ERN) is a negative deflection in the response-locked ERP that is maximal just 50 milliseconds following an erroneous response at midline frontal recording sites. Both source-localization and intracerebral recordings suggest that the ERN is generated in the anterior cingulate cortex (ACC)—a brain region that integrates information about negative emotion, pain, and cognitive control. An increased ERN has consistently been found in anxious patients—even after successful treatment. Anxiety, but not comorbid anxiety/depression, is characterized by an increased ERN, and this relationship emerges early in development. Unaffected first-degree relatives of anxious individuals also have an increased ERN. In addition to genetic mechanisms, I'll describe learning-related differences may underlie the association between ERN and anxiety. In The ERN demonstrates excellent test-retest reliability, and appears to be a trait-like biomarker of certain forms of anxiety.

Use of EEG and MEG in Studying Oscillatory in Normal and Pathological Brain States
Michael Gandal, MD, PhD
Autism and schizophrenia are neurodevelopmental disorders characterized by deficits in social and cognitive function. The lack of efficacy of current medications for these impairments represents a significant obstacle for the treatment of these disorders. Developing novel therapeutics to target these symptoms requires appropriate neural biomarkers that can be investigated in model organisms, be used to track treatment response, and provide insight into pathophysiological disease mechanisms. A growing body of evidence indicates that neural oscillations in the gamma frequency range (30-80 Hz) are disturbed in autism autism schizophrenia. Gamma synchrony has been shown to mediate a host of sensory and cognitive functions, including perceptual encoding, selective attention, salience, and working memory. This talk will demonstrate that autism and schizophrenia are characterized by an elevation in baseline cortical gamma synchrony ('noise') coupled with reduced stimulus-evoked GBRs ('signal'). This endophenotype can be recapitulated by acute or chronic NMDA-receptor hypofunction on pyramidal neurons in mice. Such signal-to-noise deficits are tied to changes in excitatory/inhibitory balance within glutamatergic afferents and can be reversed pharamacologically by boosting tonic inhibition. These

data demonstrate a clinically relevant, highly translatable biomarker for preclinical therapeutic development across a host of disorders that share common endophenotypes.

Development of Biomarkers and Clinical Tests from Basic EEG Research

Joshua Ewen, MD

Clinicians who interpret clinical tests and those who order them must understand precisely what the test results mean--and with what level of confidence those results inform the care of the patient. Tests may be based on measurements whose relationship to the mechanism of disorder are clearly understood (such as presented by the previous two speakers) or on more obscure measurements; either way, a number of experimental design and statistical considerations must be taken into account when evaluating a diagnostic test. The goal of this talk is to review a number of key aspects of diagnostic test/biomarker development and evaluation, as they relate to EEG-based methods.

Intraoperative Neurophysiologic Monuitoring During Functional Neurosurgery

Mark Stecker, MD, PhD; *Peter Dempsey, MD*; *Jay Shils, MD, PhD*

The data obtained during the neurophysiologic acquisition process is needed to plan the next surgical steps (i.e. what would the recommendation be given a recording with minimal STN and a significant portion of thalamic activity - also, what role does the state of the patient and the specific disease have on the decision process). Also, how can the OR data be used in the post-operative management of the patient? These concepts will play a large role in the interactive elements of this session.

The neurophysiologists interpretive skills and how they convey information affects how the surgeon uses that data in their decision process. IONM data interpretation may cause the surgeon; (1) to stop the surgery; (2) modify their initial plan; (3) revise something they have done. Any of these changes are potentially beneficial or detrimental to the patient. Thus, communication and trust are critical factors in the surgeon-neurophysiologists relationship in the operating room and understanding these are key element to beneficial patient outcome.

This session will present a short theoretical basis for functional surgical approaches to treatment of movement disorders. The rest of the session will present case examples with an interactive discussions, involving all three speakers and the audience.

Extending Critical Care EEG Monitoring to Community-Based Practice

Mixing General Neurology and ICU Monitoring

Jennifer Jones, DO

Abstract for Mixing general neurology and ICU monitoring.

This portion of the talk will include a review of issues relating to incorporating ICU EEG monitoring into a general neurology practice. Specifically, the topics to be covered include (1) what are the different models of employment and reimbursement for ICU monitoring (2) what are time management considerations in a general neurology practice including how to cover weekends and nights (3) how often and in what manner to communicate updated reports with ICU team (4) what is necessary to train EEG technologists to best utilize the technology?

Starting an ICU Monitoring Program in a Community Hospital

Evan Fertig, MD

Traditionally, ICU monitoring has been performed in intensive care units based in academic, tertiary medical centers. As private-practice based epilepsy programs grow, many groups are starting ICU monitoring programs in smaller, community-based hospitals, where there is a significant unmet need. This talk will focus on strategies to implement a successful program. Topics will include education of professional staff, development of protocols, potential pitfalls and solutions, and incorporation of clinical research into practice. Ample time will be provided to allow for audience members to provide input about their local experiences.

Interesting Spinal Cord Tumor Cases: A Discussion by Some Experts

Interesting Spinal Cord Tumor Cases: A Discussion by Some Experts

Francesco Sala, MD, PhD; David Gloss, MD; Mirela Simon, MD; Eva Ritzl, MD

Introduction: Intramedullary spinal cord tumors (ISCT) still represent a challenge for neurosurgeons. In the past 30 years, about 550 ISCTs have been operated on at the Institute of Neurosurgery in Verona. Since 2000, all ISCT surgeries have been performed with the aid of intraoperative neurophysiological monitoring (IONM). Material and Methods: We reviewed our experience in 240 ISCTs operated on with an IONM protocol including somatosensory evoked potentials (SEPs), transcranial muscle motor evoked potentials (mMEPs), spinal MEPs (D-wave) and the bulbocavernosus reflex (BCR). Selected cases are presented for discussion.

Results and Conclusions: 1) The vast majority of patients with ISCT present some degree of transient neurological worsening early after surgery, in spite of the use of IONM. 2) The neuro-oncological aspects of ISCT surgery are relevant and the IONM professional

must include these in establishing warning IONM criteria. 3) The goals of IONM should be tailored to different ISCT pathology (ependymomas, astrocytomas, hemangioblastoma,…). 4) The main factor limiting the extent of resection of ISCTs remains the cleavage plane between tumor and spinal cord, yet IONM continue to play an essential role in this surgery.

Semiology of Status Epilepticus in Adults

Semiology of Status Epilepticus in the Responsive Patient

Frank Drislane, MD

Nonconvulsive status epilepticus (NCSE) produces a remarkable variety of altered behavior -- and prominent 'non-behavior.' This session will concentrate on the signs and symptoms of NCSE in patients who retain some, if limited, responsiveness. Before integrating EEG evidence, it is worthwhile to record the semiology precisely, to help avoid confusion between the seizure and the epilepsy syndrome. Some NCSE patients have seemingly global dysfunction, with immobility and staring. This can be of a primary generalized (absence) nature or focal-onset NCSE (often considered complex partial SE). The EEG, however, can be misleading as to the nature of the seizures. Simple and complex automatisms can occur in both focal-onset and generalized NCSE; careful description of the semiology may help define the syndrome. "Hypermotor" seizures may also occur without complete unresponsiveness. Finally, some NCSE appears quite focal or regional in character, with perceptual and cognitive deficits very similar to those seen in patients with fixed focal lesions such as strokes and tumors. Failure to recognize that such semiology can result from NCSE can lead to major misdiagnosis. Among these cases are aphasia, amnesia, hemianopia, and neglect syndromes. Difficulties in diagnosis will be detailed.

Correlation of Semiology and EEG Pattern in Comatose Patients

Peter Kaplan, MD, FRCP

There is an extensive literature on EEG patterns in encephalopathy and comatose patients. EEG patterns range from reactive or unreactive patterns of theta, theta/delta and delta; alpha and spindle coma tracings; and periodic waves in the form of triphasic waves. Various constellations of epileptiform activity occur that span the interictal-ictal continuum, with patterns of periodic discharges appearing focally, bilaterally, asynchronously and synchronously. Isolated seizures may appear focally or in a generalized pattern and when in the form of status epilepticusasmay may be continous or exhibit discrete seizures. Semiology during unresponsiveness varies from the minimally unreactive patient to the deeply comatose, the latter with either intact or variably absent brainstem reflexes, motor responses to pain and myoclonus. Clinical - EEG corrleations in the context of etiology have prognostic import as well as providing a yardstick for management of coma and seizures or status epilepticus in the intensive care setting.

Intraoperative Neuromonitoring Below the Belt

Bulbocavernosus Reflex; Anal Sphincter and Cremaster Electromyography; Pelvic Autonomic Monitoring

Stanley Skinner, MD

The great advantage of the BCR is its ability to detect low sacral segmental conduction block at both the peripheral somatic (afferent or efferent) and central levels. Because parasympathetic fibers are associated with sacral roots of the cauda equina, BCR affords surrogate testing of major bowel, bladder, and sexual vegetative functions. Obviously, for surgery within (or affecting) the lower pelvic periphery, sacral root/plexus, cauda equina, and conus medullaris… BCR implementation should be very strongly considered. Extracellular recordings within the corpora (perivaginal and penile erectile tissue) may be tentatively termed "electromyography." Although intriguing animal research has been published, rigorous but flexible thinking will be required 1) to exclude possible artifacts during slow wave recording (< 2 Hz) and 2) to accept the realities of unsummated responses in the OR. Near field bipolar recording by electrodes that are shielded from skin activity and can reject other artifactual common mode signals will be required. The promised benefit of an electrophysiological approach lies in the recording of extracellular field potentials without waiting for a fickle end organ physical change (like strain gauges around the penis). Electromyographic recordings should be equally applicable in both sexes.

Pudendal Nerve and Sacral Root Evoked Potentials

Matthew Eccher, MD, MSPH

Originally described in 1982, scalp SSEP responses can be recorded following stimulation of multiple different pudendal-nerve- and sacral root-supplied structures. The resulting P40 response is usually highest amplitude at Cz. Responses are generally easy to resolve and therefore should be of equivalent ease to follow for neurophysiologic intraoperative monitoring versus lower limb peripheral nerve SSEPs (e.g., tibial or fibular [peroneal] nerves), but sizeable reports of pudendal SSEP monitoring are few. Direct orthodromic sensory nerve action potential (SNAP) recording from the cauda equina in response to single such sacral stimuli has been reported of utility for preserving roots that participate in urinary control during dorsal rhizotomy procedures for spasticity. Technical application of both techniques is quite straightforward. As in most areas of NIOM, there are no well-constructed historical-control series informing use of these techniques, and certainly no clinical trials. Given the socially devastating consequences of urinary and anal continence

disturbances, and a fairly high rate of functional postoperative disturbances when sacral roots are manipulated, this field begs more active clinical investigation.

Neurophysiology of Complex Spinal Cord Untethering

Francesco Sala, .MD

Introduction: Intraoperative neurophysiologic monitoring (IOM) is nowadays extensively used to minimize neurological morbidity in tethered cord surgery. We present a 10-year clinical experience using a multimodal IOM approach in 47 patients with a tethered cord secondary to a variety of spinal dysraphisms. **Material and Methods:** Neurophysiological mapping of the cauda equina was performed through direct rootlet stimulation and bilateral recording from segmental target muscles in the lower limbs, and anal sphincters. Monitoring techniques included somatosensory evoked potentials (SEPs), transcranial motor evoked potentials (MEPs) from limb muscles and anal sphincters, and the bulbocavernosus reflex (BCR). **Results:** Neurophysiological mapping was successful in all cases. In six patients an unexpected muscle response was evoked by stimulating tissue macroscopically considered as not functional. The monitorability rate was 84% for SEPs, 97% for limb muscle MEPs, 74% for anal sphincter MEPs, and 59% for the BCR. SEPs, MEPs and BCR remained stable during surgery in all patients but one. This child showed worsening of a lower limb paresis and urinary/fecal incontinence, which completely recovered over 8 and 5 weeks respectively. **Conclusion:** Combining mapping and monitoring IOM techniques allow to safely address tethered cord surgery

Special Interest Group: Critical Care EEG Monitoring and Outcomes: Do We Have Enough Data?

cEEG in the Adult ICU (Seizure Indications)

Jeffrey Kennedy, MD

The past few decades have witnessed a revolution within the fields of epilepsy and intensive care medicine through a significant increase and interest in continuous EEG monitoring (cEEG) in adult intensive care units. Advancements in technology leading to digital recording and remote review have allowed practical and feasible cEEG to be performed. Currently, the major application of cEEG in ICU is primarily to diagnose paroxysmal events, detect nonconvulsive seizures, and nonconvulsive status epilepticus (NCSE). cEEG monitoring has also revealed further understanding of associations of NCS and NCSE with several pathological processes commonly encountered in the ICU setting as well as identified several EEG patterns, whose clinical significance remains yet to be elucidated.

Update of Current Practices

Jay Gavvala, MD

The use of continuous EEG (cEEG) monitoring in the ICU has increased dramatically and is frequently used in a variety of clinical settings. Unlike other procedures or imaging modalities, at present time, there are no practice parameters or clinical guidelines to ensure a minimal standard for the completion and reporting of cEEG monitoring in addition to basic requirements for all staff involved. In a recent survey aimed to describe current practice among neurophysiologists and neurointensivists, practices are similar among neuro-intensivists and neurophysiologists regarding primary indications for utilization of cEEG. However, there is wide variability regarding recommended duration of monitoring and secondary indications for cEEG. Among neurophysiologists, there was large practice variability in the technical aspects of cEEG including frequency of review and use of EEG technologists in screening cEEG. These results have demonstrated that while use of cEEG has become more frequent with greater agreement for primary indications of cEEG monitoring, there is large practice variability in both use for indications other than seizures as well as cEEG reporting frequency, utilization of technologists processing time and other aspects of the practice of cEEG monitoring. Further large prospective multicenter studies are needed to clarify the optimal utilization of cEEG.

cEEG in the Neonatal ICU

Courtney Wusthoff, MD

The arguments for and against cEEG in the Neonatal ICU will be discussed, citing currently available evidence supporting each side of the debate. This session will critically evaluate the data regarding how cEEG monitoring for neonatal seizures and background predicts and impacts ultimate clinical outcomes. Priorities for future research will be considered by the group.

cEEG in the Pediatric ICU

Cecil Hahn, MD, MPH

In this presenation, I will review the current practice of continuous EEG monitoring in North American pediatric ICUs, summarize the available epidemiological data on the prevalence and risk factors for seizures among critically ill children, and discuss our current knowlege of the relationship between electrograhpic seizures and outcomes. I will highlight the gaps in knowlege regarding outcomes, and discuss research strategies to address these gaps.

EMG SIG

Conduction Block is Essential for diagnosis of MMN

Michael Cartwright, MD, MS

Multifocal motor neuropathy (MMN) is a rare condition with a prevalence of 1 to 2 per 100,000 individuals. It affects men more often than women and typically starts with slowly progressive asymmetric upper extremity weakness without sensory involvement. Differentiating MMN from motor neuron disease is critical, because MMN responds to immune modulating treatment, typically intravenous immunoglobulin. Conduction block is considered the electrodiagnostic hallmark of MMN, but there is a lack of consensus as to whether conduction block is essential for the diagnosis of MMN. This presentation will review the data supporting and refuting the need to demonstrate conduction block prior to diagnosing and treating MMN.

Skin Biopsy is the Best Diagnostic Test for Assessment of Small Fiber Neuropathy

Morris Fisher, MD

Small fiver neuropathies are common. They are clinically important and frequently associated in an isolated fashion with prominent pain. The clinical findings in SFN may be minimal and the usual electrodiagnostic studies unrevealing even if other electodiagnostic studies of autonomic dysfunction may be abnormal. Skin biopsies with evaluation of intraepidermal fiber densities (IEFD) are a well-established, safe, and readily available technique for defining SFN. The clinical features, evaluation, and differential diagnosis of SFN will be discussed. The arguments for skin biopsies with IENF density analyses being the best diagnostic test will be presented.

Ultrasound Improves Sensitivity in the Diagnosis of Entrapment neuropathy

Francis Walker, MD

Neuromuscular ultrasound is a recent addition to many EMG laboratories that provides complementary anatomic, and to some extent, physiologic information regarding entrapment neuropathies. Determining the value of this additional information is not as straightforward as it might seem because there is no gold standard for the diagnosis of nerve entrapment. It is tempting to use electrodiagnostic criteria as the gold standard, but then no test could be shown to add to sensitivity to electrodiagnosis. Clinical criteria for entrapment neuropathies are problematic in that they are not particularly sensitive nor specific. In practice, ultrasound sometimes is positive in patients with classical clinical features of the disease when electrodiagnostic studies are normal; the reverse is also true. As ultrasound techniques have improved, there has been an improvement in the accuracy in diagnosing carpal tunnel syndrome, and in some studies, ultrasound shows superior sensitivity. For ulnar neuropathy at the elbow, ultrasound offers some advantages over electrodiagnostic testing, particularly when there is ulnar nerve subluxation. Clearly, ultrasound adds information not routinely available by electrodiagnostic studies, particularly in terms of pathology in adjacent structures, anatomic variants, and dynamic aspects of nerve movement.

Intraoperative Neurophysiologic Monitoring SIG

Welcome and Introduction

Jaime Lopez, MD

The goal of the IOM-SIG is to provide a venue to practitioners of intraoperative neurophysiologic monitoring where they can discuss matters of importance. This is done via formal lectures and in an open and informal manner moderated by the IOM-SIG Director.

Cervical surgery and C5 palsy: IONM or not to IONM? And How? Evidence-Based?

Viet Nguyen, MD

Two studies, both from the past few months, highlight the risk of C-5 root palsy in cervical spine surgery.
One study examined 58 cervical spondylolisthesis and/or kyphosis cases. 24 involved laminoplasty with posterior fusion (lateral mass instrumentation); 6 experienced a C5 palsy (all six included fusion at the C4-5 level). 34 involved laminoplasty without fusion; only 1 experienced a C5 palsy ($p < 0.05$). No patients underwent foraminotomy. No patients received intraoperative neurophysiological monitoring (IONM). Another study examined 235 cases involving the C4-5 level, ranging from single-level discectomy to combined anterior-posterior fusion. Of the 12 cases with C5 root injury, 5 were immediately postoperative and had transcranial electrical motor evoked potential (tceMEP) changes, but only one had significant spontaneous electromyography (spEMG) activity. The other 7 had delayed-onset deltoid weakness, and neither tceMEP or spEMG was effective in predicting those cases. These studies imply that (1) C5 palsy can occur in cervical spine surgery, particularly cases involving corpectomy and/or fusion at C4-5, (2) IONM can detect acute, but not delayed-onset, C-5 nerve root injury, and (3) tceMEP changes are more senstive than spEMG at detecting this injury.

Diagnostic Advances in ALS

Ultrasound in ALS

Michael Cartwright, MD, MS

The diagnosis of amyotrophic lateral sclerosis (ALS) is based upon the appropriate history and physical examination, but electrodiagnostic testing, imaging, serum studies, and sometimes cerebrospinal fluid analyses are used to support the diagnosis. Over the past decade the use of diagnostic neuromuscular ultrasound has increased for a variety of conditions, including ALS, with several studies describing the typical findings in the muscles and nerves of those with ALS. This presentation will review the use of neuromuscular ultrasound to detect fasciculations, to assess muscle size and echotexture, to evaluate diaphragm thickness and mobility, and to describe the typical findings in peripheral nerves of individuals with ALS. In addition, incorporation of neuromuscular ultrasound into the diagnostic evaluation of ALS will be described, with a focus on practical experience and logistics. While further research in this field is needed to refine the technique, the necessary ultrasound devices and data are already available and neurologists have begun incorporating neuromuscular ultrasound into their routine evaluation of suspected ALS.

Electrodiagnosis in ALS

Devon Rubin, MD

Amyotrophic lateral sclerosis (ALS) is a progressive disorder of motor neurons of unknown etiology. Establishing an accurate diagnosis of ALS requires a combination of clinical findings, electrodiagnostic confirmation, and exclusion of other mimicking disorders. Electrodiagnostic studies are useful to provide supportive evidence of ALS, may help to provide prognostic information, and are used to identify other disorders that may clinically mimic ALS. A review of the typical features on nerve conduction studies and needle EMG will be presented and clinical examples of atypical findings to indicate alternative diagnoses will be demonstrated.

Fast-Train Cortical and Subcortical Stimulation for Motor Mapping and Monitoring

Intracranial MEP Stimulation and Recording - Practical Setup, Pearls and Pitfalls

Tyson Hale, AuD

Intracranial mapping during brain tumor resection may involve a mix of neurophysiologic modalities such as electroencephalography (EEG), somatosensory evoked potentials (SSEPs), and myogenic evoked potentials (MEPs). Hospitals with in-house IOM teams and independent IOM companies have specific protocols for brain mapping procedures that may vary from site to site. Additionally, surgeons and neurophysiologists may decide adjustments be made for a given mapping protocol before, or even within a case dependent on factors such as surgical approach, location of lesion, etc. This lecture will focus on Geisinger Health System's approach to brain mapping studies, utilizing intracranial MEP testing with regard to initial planning, setup, supplies, modalities, and parameters for stimulus and recording. Ultimately, mapping success for each individual patient is achieved through willing neurosurgeons, so it is very important to educate and plan with them before each mapping case is performed.

Cortical and Subcortical MEP Stimulation

Stuart Hoffman, DO

Despite ongoing advances in neuronavigation imaging tools to aid in tumor resection, neurophysiologic mapping and monitoring of motor pathways remains the gold standard when supratentorial tumors are located near the motor strip and subcortical motor pathways. Intraoperative neurophysiologic mapping and monitoring of these pathways aids the neurosurgeon by improving the odds of obtaining a total resection of the tumor while avoiding a new post-operative deficit. This talk will focus on the fast-train monopolar electrical stimulation technique for mapping and monitoring cortical and subcortical motor pathways during neurosurgical resection of tumors near these important pathways. The history of neurophysiologic mapping and monitoring of motor pathways will be briefly reviewed to put the current practice in context. Pertinent neuroanatomy and neurophysiology regarding these techniques will be highlighted and the basic neurophysiologic set up will be presented. Other techniques, including 60 Hz bipolar stimulation will be reviewed and contrasted with fast-train techniques. Intraoperative alert criteria, subcortical intensity-distance mapping data, and the pitfalls and limitations of these neurophysiologic techniques will be presented.

DCS-MEP for Epilepsy Surgery: Current Evidence and Questions

Matthew Eccher, MD, MSPH

In the years since the first 1993 report of fast-train monophasic direct cortical stimulation, there have been several small reports regarding this technique in the epilepsy population. The clear advantage appears to be that there is a lower rate of induced seizures with this technique compared with biphasic 50-to-60 Hz long train stimulation. There are several comparative disadvantages, however, including inapplicability of this technique for sensory or language mapping, absence of FDA-approved equipment for this technique, and, perhaps most challenging, an absence of good head-to-head comparison of testing results between this technique and the older one. What literature there is in this area will be reviewed, some illustrative cases discussed, and next research steps for the field suggested.

DCS-MEP and CST Motor Threshold: Utility for Cerebral Hemispheric Lesion Surgery

Kathleen Seidel, MD

Mapping and monitoring are believed to provide an early warning sign to avoid damage to the corticospinal tract (CST) during brain surgery. We compared subcortical monopolar short train stimulation thresholds (MT) with direct cortical stimulation (DCS) motor evoked potential (MEP) monitoring signal abnormalities in 100 patients. At 3 months, 5 patients had a postoperative new motor deficit (lowest mapping MT 20mA, 13mA, 6mA, 3mA, 1mA). All showed DCS-MEP alterations (2 sudden irreversible threshold increases and 3 sudden irreversible MEP losses). Of these cases, 2 had vascular lesions (MT 20mA, 13mA) and 3 had mechanical CST damage (MT 1mA, 3mA and 6mA; in the latter two resection was continued after mapping and DCS-MEP alterations occurred thereafter). Mapping should primarily guide tumor resection adjacent to the CST. DCS-MEP is a useful predictor of deficits, but its value is limited because signal alterations were reversible in only approximately 60% of cases and irreversibility is a post-hoc definition. The true safe mapping MT is lower than previously thought. We postulate a mapping MT below 1 mA where irreversible DCS-MEP changes and motor deficits regularly occur. The limited spatial and temporal coverage of contemporary mapping may increase error and may contribute to false higher MTs.

Stereo Electroencephalography

Why Stereo Electroencephalography?

Samden Lhatoo, MD

Epilepsy surgery is often the most effective intervention, whether curative or palliative, in the treatment of refractory focal epilepsy. There is emerging evidence that the epilepsy surgery landscape in the industrialized world is changing such that straightforward temporal lobectomy cases are diminishing in relative proportion to extratemporal, MRI negative and previously failed surgery patients. Increasing case complexity is likely to require increased usage of intracranial electrode studies. Whereas the subdural grid electrodes approach has been the preferred methodology in many centers worldwide, the stereotactic electroencephalography (SEEG) approach is gaining traction, particularly in cases with putative epileptogenic zones inaccessible to grids and strips. These areas may lie deep in basal and mesial brain areas but also in sulcal depths over the dorsolateral brain convexities. SEEG thus provides a 3 dimensional perspective that with the appropriate surgical hypothesis, encompasses necessary and previously untargetable brain regions. Since resective surgery has such potential for benifit, SEEG is an investigational modality that should complement traditional approaches and provide intractable epilepsy patients with surgical options otherwise unavailable to them.

Surgical Approach: Concept and Technique

Jorge Alvaro Gonzalez-Martinez, MD, PhD

In many centers in North America, invasive monitoring for medically intractable focal epilepsy is performed using subdural grids/strips, combined or not with depth electrodes. Most recently, the stereo-electroencephalography (SEEG) methodology has been applied in patients with difficult to localize seizures when a deep focus is suspected or a more wide spread epileptic network is responsible for the generation and early propagation of the epileptic activity. The SEEG method provides a 3 dimensional map of the epileptic network in association with minimal morbidity. In this presentation, the authors will discuss the principles behind the SEEG methodology, surgical indications and as variances in surgical techniques.

Pediatric Stereo EEG: Challenges and Opportunities

Jonathan Miller, MD

Focal epilepsy in infancy and early childhood often is not associated with abnormalities on imaging because incomplete myelination interferes with the recognition of cortical dysplasia on MRI. Even when a lesion is present, MRI and video-EEG sometimes fail to adequately localize the epileptogenic zone. In these cases, invasive monitoring can be helpful to determine whether resection is feasible and to define the margins of resection. Stereo-EEG (SEEG) involves placement of multiple intracerebral electrodes to sample brain tissue directly. When thoughtfully planned, the individual contacts become “voxels” that can be used to plan the subsequent resection. Very young children present unique challenges related to the ability of the skull to accommodate anchoring devices and the tolerance of young children to prolonged invasive monitoring in general. Also, unlike widespread coverage with a subdural grid, each individual SEEG electrode samples only a single dimension, which can lead to a “tunnel vision” effect unless the arrangement of electrodes is carefully tailored according to a good working hypothesis defined by a multidisciplinary approach. When appropriately applied, SEEG can help to identify focal epilepsy in a majority of pediatric epilepsy patients with minimal morbidity.

Clinical Neurophysiology Trials in the Neurointensive Care Unit: Focus on TRENdS

TRENdS: From Conception to Implementation

Aatif Husain, MD

The Treatment of Recurrent Nonconvulsive Seizures (TRENdS) study is the first study of its kind in which the efficacy of two antiepileptic drugs (AEDs) is being compared for the treatment of electrographic seizures using continuous EEG monitoring. This is an industry sponsored, investigator initiated study (IIS). The challenges and rewards of designing and implementing a multicenter, randomized IIS will be discussed.

Participating in a Clinical Trial

Jong Woo Lee, MD, PhD; *M. Brandon Westover, MD PhD*

There are far too few evidence-based treatments for patients with critical neurological illness, including the treatment of nonconvulsive seizures. Clinical trials represent the best and most important method available to address these questions. This symposium will provide an overview of designing, implementing, and executing a clinical neurophysiology based trial. We will focus on the multicenter trial "Intravenous Lacosamide Compared with Fosphenytoin in the Treatment of Patients with Frequent Nonconvulsive Seizure (TRENdS)." Issues and challenges specific to neurocritical care patients will be addressed.

Next Steps in Clinical Neurophysiology Trials

Suzette LaRoche, MD

Over the past decade, there has been considerable increase in the utilization of continuous EEG monitoring in critically ill patients, particularly in patients with acute brain injury. However, in the age of rising healthcare costs, administrators demand evidence of "return on investment" prior to funding evolving technologies. Unfortunately, it remains unclear exactly what influence EEG monitoring for detection and treatment of secondary injuries such as seizures and ischemia has on outcome. Retrospective studies have shown that electrographic seizures are common, yet prospective data on how treatment of seizures affects outcome measures such as length of stay and functional recovery is lacking. Nonetheless, findings on EEG frequently result in treatment changes and can have a large impact on clinical decision making. Therefore, the challenge is to devise clinical trials that might provide better outcome data but also strive to provide more efficient and cost-effective EEG monitoring. This session will discuss the next steps that are necessary to design and implement neurophysiology trials that would answer these critical clinical questions.

Crashing the Cultures of the Sole MEG or EEG Source Modeling: Inseparable, Not Only Complementary

Anto Bagic, MD, PhD; *Richard Burgess, MD, PhD*; *John Ebersole, MD*

Even after the optimal combined acquisition of MEG and EEG for identification of epileptic foci, it is the prevailing practice in clinical MEG to perform source localization only of MEG, while source localization of EEG is usually an exception rather than the rule. Overall in fact, despite the recommendation and customary inclusion of simultaneous EEG during MEG, different centers use EEG quite variably, from being completely ignored, to being a pointer to "interesting" corresponding MEG discharges for source localization, to being used to corroborate authenticity and relevance of MEG discharges, and finally on rare occasions to being combined with MEG for electromagnetic source imaging. Furthermore, software to import and localize EEG and MEG may not be available from the MEG instrument manufacturer. Thus, while we profess the complementary nature of MEG and EEG, it is evident that this complementarity is exploited quite incompletely. To highlight the critical issues regarding EEG and MEG source localization, we will have an expert discussion of this issue in a classic debate format:

Neonatal and Pediatric EEG: Patterns of Epileptic Encephalopathies Across the Age Range

Evolution Over Time in Neonatal Epileptic Encephalopathies

Courtney Wusthoff, MD

Dr. Wusthoff will consider EEG patterns in the neonatal epileptic encephalopathies. Illustrations of specific findings will be presented, including those in KCNQ2 encephalopathy, Ohtahara syndrome, and congenital hemimegalencephaly. Typical evolution of EEG findings in the neonatal period and infancy will be considered, particularly in relationship to etiology and outcome. This session will include a focus on debunking myths about EEG findings in specific diseases.