SPECIAL ARTICLE

Electrodiagnosis in Clinical Neurology - The Challenge for Pediatric Neurology in Indonesia

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Introduction

In most cases the diagnosis in neurology is based upon clinical manifestations, anatomical abnormalities, physiological disturbances and biochemical investigations. Clinical manifestations can be investigated by history of illness and neurological examinations. Neurophysiological examination (electrodiagnosis) can provide information on physiological abnormalities of the central nervous system (CNS). In recent years computer tomography (CT) and magnetic resonance imaging (MRI) have made it possible to visualize the morphologic anatomy of the CNS in detail and to diagnose the lesion.[1]

Infants and children are often poor historians of neurological symptoms, and the clinical examination can be difficult, relying significantly on observation made by the child's parents and attending physicians. The lack of information makes electrodiagnostic examination such as electroencephalography (EEG), evoked potentials (EPs), and electromyography (EMG), much more important as an adjuvant to the clinical examination.

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This paper will describe the principles of electrodiagnostic studies and the clinical utility of electrodiagnosis in pediatrics.

Principles of Electrodiagnostic Studies

1. Electroencephalography

There is now considerable evidence from experiments on animals to suggest that the rhythmic activity has a cortical origin, being derived from the postsynaptic potentials of cortical neurons. These neurons are triggered by neural activity in subcortical structure or by horizontally oriented cortex cells. Many studies suggest that it is the thalamus that serves as pacemaker of the cortical rhythms. Rhythmic activity can arise in any or all of the thalamic nuclei, can spread from one nucleus to another and it is imposed on the cortex via the thalamocortical projections, that are recorded with the electroencephalograph (EEG).[2]

Recording can show the background rhythms (e.g. alpha, mu rhythm) and paroxysmal activities such as localized slow activity or epileptic discharges. Background activities are not less important than paroxysmal activity, because it can be used to evaluate the maturity or abnormality of the brain. Van Huffelen[3] studied quantitative electroencephalography in cerebral ischemia and he found that a lower peak frequency of the alpha rhythm and an increase in delta activity indicates the ischemic side in patients with cerebral ischemia.

Digital EEG techniques have evolved gradually over the past 30 year. This equipment can acquire more channels of data than usually are displayed on paper EEGs. It is common to record from all 21 of the ordinary 10-20 electrode locations. Digital analysis techniques provide the opportunity to select the optimal montage whichever montage is desired or even several different montages and a variety of filters. An interesting segment of the record also can be replayed on expanded scale to look at the timing and potential distribution of spike and sharp waves in greater detail.[4]

Digital EEG or electroencephalographic brain mapping can be applied specifically to construct topographic maps of cortical electrical activity. In this technique, EEG features are plotted providing a small map of the cortical potentials. Voltages or other features are displayed using contour lines or color coding to identify similarly valued regions. Thus, it can help localize scalp regions, with excess or decreased amounts of an EEG feature.[4] Koszer and colleagues found that the dynamics of spike activity, including correlation, are better visualized by using the computerized technique than by visual interpretation.

2. Evoked potentials

An EP is an electrical manifestation of the brain in response to an external stimulus. The source of the potentials is not completely established. The electric generators may be discharging neuronal pools, or may be electrical waves passing through nerve tracts. Far-field potentials can also be recorded by scalp electrodes.[6] Most EPs cannot be seen in routine EEG recordings. This is because their low amplitudes (0,1 - 20 uV) and their admixture with normal background activity and various artifacts that together are from twenty to several hundred microvolts in amplitude.[7]

Sensory stimuli, after entering the cord pass through the sensory afferent via the dorsal root, ascend the cord. A synapse occurs in the nucleus cuneatus (for the upper extremity) or the nucleus gracilis (for lower extremity). Fibers then pass through the brainstem (the lemniscal system) and again synapse in the thalamus. Finally, the impulse travels to the sensory cortex via third-order neurons. Because of this, somatosensory evoked potentials (SEP) can be used to study pathology in the spinal cord, brainstem, thalamus, or the sensory cortex.[6,7]

Visual stimuli, after passing through the optic media, are processed by the outer layer of the retina. Fibers then pass through the optic nerve, and after decussation through the optic tracts. They synapse in the lateral geniculate bodies. From there, the geniculocalcarina radiation carries the impulse to the visual (occipital) cortex. With the use of visual evoked potentials (VEP), problems in the optic nerve, chiasm, geniculocalcarine radiation, or occipital lobes can be studied .[6]

Auditory stimuli are sensed by the eighth nerve and relayed to the midbrain. There are synapses with nuclei in the pontomedullary region (cohlear nucleus, superior olivary nucleus). Subsequently, the stimuli pass through the upper pons (via the lateral lemniscus and its nucleus) and low midbrain (inferior colliculus) to synapse in the thalamus. Because of this, brainstem auditory evoked potential (BAEP) can be used to evaluate the eighth nerve, the brainstem, and from the pons to the midbrain.[6,7]

3. Electromyography

Electromyography (EMG) studies diseases that might affect the anterior horn cells, nerve roots, peripheral nerves, neuromuscular junctions, or muscles, So that it can be a valuable asset in the diagnosis and follow up of neuromuscular diseases. There are three types of tests: (1) nerve conduction velocity; (2) needle EMG; and (3) the repetitive stimulation test.

Nerve conduction velocity is an expression of the physiological or pathophysiological state of the nerves. Nerve conduction velocity studies:[8] (a) motor nerve conduction; (b) sensory nerve conduction, and (c) mixed nerve conduction.

The needle electromyographic examination of skeletal muscle usually consists of

four steps:[8,9] (1) insertional activity when a needle electrode is introduced into the muscle; (2) spontaneous activity is evaluated at rest; (3) motor unit potentials are recorded with mild voluntary contraction of the muscle; (4) recruitment and interference pattern are assessed at maximal muscle contraction.

The result of nerve conduction and needle electromyography studies can be closely correlated to the structural abnormalities of the nerve. Based on histologic changes in the nerve and the nature of conduction abnormalities, axonal degeneration and segmental demyelination can be identified. [9]

The function of the neuromuscular junction can be indirectly assessed by repetitive stimulation of motor nerve with a recording electrode over the appropriate muscle. The muscle action potentials characteristically show a decremental response to repetitive stimulation at low rates and to a lesser extent at higher rates. This test is indicated and most helpful in myasthenia gravis, myasthenic syndrome (the Eaton-Lambert syndrome) and botulism.[8,9] Single fiber electromyography (SFEMG) is a recording method of single muscle fiber potentials in voluntarily contracting human muscle with use the single fiber needle. This method is useful as supplement to conventional electromyography:[9] (1) measurement of fiber density, the number of single fiber action potentials within the recording radius of the electrode; (2) determination of electromyographic jitters, the variability of the interpotential interval between two or more single muscle fibers belonging to the same motor unit.

Oey and coworkers[10] demonstrated that SFEMG technique showed a better relation with the clinical diagnosis of ocular myasthenia gravis than repetitive nerve stimulation test and infrared reflection oculography.

Clinical Utility of Electrodiagnosis in Pediatrics

1. Digital EEG and brain mapping

The EEG is an important tool in the evaluation of an infant or child with symptoms referable to the central nervous system. The EEG can provide unique information in the assessment of newborns about:[11] (1) diagnosis and therapy of seizures; (2) evaluation of infants with compromised cerebral function due to primary neurologic disorders (e.g., cerebral infarction) and systemic disease; (3) determination of conceptional age; (4) identification of specific neurologic entities, such as congenital malformation, herpes simplex encephalitis and metabolic encephalopathies; (5) determination of prognosis and long-term neurologic outcome.

In children EEG is used to:[11] (1) confirm clinical suspicion or the location of a particular lesion, (2) follow the progression of disease and the response to therapy, (3)

investigate the possibility that the episodic behavioral or motor phenomena are epileptic in nature, or (4) provide data that will help the clinician classify a seizure disorder and develop rational therapy.

Avoiding the pitfalls of pediatric EEG interpretation includes the recognition of such normal EEG features during wakefulness as posterior slow waves, mu rhythm, and lambda waves. In addition, the understanding of age-dependent characteristics of EEG state changes is essential.[12]

Digital EEG with brain mapping studies are more sensitive than routine EEG for detecting abnormalities in patients with epilepsy and damage to the cerebral hemispheres from cerebrovascular disease. Careful analysis of the field of epileptiform spikes can shed light on the location of the cortical generator, and may contribute prognostic information about the patient's seizure disorder.[1]

Van der Meij et al[13] studied sequential EEG mapping in rolandic epilepsy and they found a specific sequence of changes of potential fields . This sequence started with a bipolar field, with the negative pole in the frontal region and the positive pole in the centro-temporal region. This bipolar field changed to a unipolar or bipolar field, with a negative potential field in the centro-temporal region and, a simultaneous positive potential field in the frontal region, morphologically represented by the prominent rolandic spike.

2. Evoked potentials

The clinical utility of evoked potentials is based on their ability:[7] (1) to demonstrate abnormal sensory system function when the history and or neurological examination are equivocal; (2) to reveal the presence of clinically unsuspected malfunction in a sensory system when demyelinating disease is suspected; (3) to help define the anatomic distribution of a disease process; and (4) to monitor changes objectively over time in a patient's status epilepticus.

2.1. Somatosensory evoked potentials

Somatosensory evoked potential testing can provide a noninvasive, objective method of evaluating the central and peripheral nervous system, and can also generate information about the maturation of the human afferent sensory system. This testing is particularly useful because the clinical sensory neurologic examination is often difficult and unreliable in infants and young children.[14]

In pediatrics, evoked potentials must be interpreted with an understanding of the maturational changes which affect normative data.[7] Clinical utility of SEP studies in children has been shown in:[14,15] (1) neurodegenerative disorders such as leukodys-

trophy, Freidreich's ataxia, hereditary motor sensory neuropathy, because disorders affecting gray and white matter cause abnormal responses; (2) encephalopathies; (3) perinatal asphyxia; (4) compressive lesions of the braistem and spinal cord; (5) structural lesions such as myelodysplasia; (6) peripheral nerve lesions such as brachial plexus injuries; (8) juvenile diabetes mellitus; (9) comatose children for prognostic value.

2.2. Brainstem auditory evoked potentials

The BAEP is used to test:[7] (1) the peripheral hearing apparatus in conductive and sensorineural hearing disorders and (2) the brainstem auditory tracts in CNS disorders. The BAEP is usually abnormal with brainstem tumors, almost always abnormal with the leukodystrophies, and often abnormal with degenerative diseases that affect white matter. It is a valuable part of the neurologic work up in children presenting with degenerative processes, encephalopathy, ataxia or coma.[16]

In patients in whom the extent of brain damage has not been fully determined, the BAEP can demonstrate whether the brainstem has been affected significantly. It is important to realize, however, that the BAEP is resistant to many neurologic insults and can be normal in a child who is far from normal neurologically.[16] Hearing impairment during the first few years of life results in auditory deprivation at a time when the infant experiences the extensive sensory stimulation that is necessary for the normal development of speech and language. All newborn infants who are considered 'at risk' for hearing impairment should have their hearing assessed:[17] (1) family history of congenital childhood hearing loss; (2) congenital perinatal infection (e.g. TORCH); (3) anatomic malformations involving the head or neck; (4) birth weight <1500 g; (5) hyperbilirubinemia; (6) ototoxic medications; (7) bacterial meningitis; (8) severe depression at birth; (9) mechanical ventilation for more than 10 days; (10) stigmata of syndromes known to include sensory neural hearing loss (e.g. Waardenburg or Usher's syndrome).

Hearing is assessed using 30 dB nHL clicks presented at a rate of 60 per second, 1000 responses are averaged. Any infant not showing responses at 30 dB is retested at age 3 to 5 months. Many of these infants have by that time developed normal threshold. Screening at 3 months is more accurate than screening in the neonatal period, because transient neonatal conductive losses may have resolved.[16]

BAEP are used to test the brain stem auditory tract in CNS disoders[7,16] such as: ototoxic drugs, cerebellopontin angle tumors or tumors in the posterior fossa, multiple sclerosis, degenerative diseases, meningitis, hydrocephalus, Arnold Chiari malformation, Guillain-Barre syndrome, epilepsy, metabolic disorders, mental retardation, and brain death.

2.3. Visual evoked potentials

Flash VEPs provide a noninvasive means of evaluating the integrity of the sensory pathways of very young infant. They have been used to (1) evaluate the effect of intraventricular hemorrhages on cortical maturation and mental development, and (2) to evaluate the integrity of shunts used to control increased intracranial pressure in posthemorrhagic hydrocephalus.[18]

Placzek and coworkers[19] recorded flash VEPs from premature infants, in comparison with neurologically normal children without IVH, to neurologically abnormal infants with grade III IVH, and found a delayed onset of the first major positive wave in the severely ill children. The results of several author's investigations,[20,21] have demonstrated a significant relationship between ventricular size, intracranial pressure and evoked potential parameters and consequently the usefulness of the flash VEP examination in the assessment of patients with hydrocephalus .

Pattern VEPs can be used for the detection of amblyopia, in patients with refractive error, occulomotor disorders, such as nystagmus, cortical blindness, and delayed visual maturation.[18] Moskowitz and Sokol[22] reported pattern VEPs in children ranging in age 1 month to 5 years in response to large and small checks. Qualitative analysis of the VEP wave form showed that the first major positive component, P_1, is consistently present at all ages, while the frequency of occurrence of later positive component more variable..

3. Electromyography

Electromyography in children is used to study peripheral neuropathy, radiculopathy, myopathy, myasthenia gravis, Bell's palsy, myotonic disorders, polymyositis, motor neuron diseases, peroneal nerve palsy, heredodegenerative diseases. Diagnostic possibilities exist in peripheral neuropathies at varying ages of onset:[8]

- Infancy: congenital sensory neuropathy, hypomyelinative neuropathy, Dejerine-Sottas neuropathy, Riley-Day syndrome, infantile axonal dystrophy, Krabbe's disease.
- One - three years of age: giant axonal neuropathy, metachromatic leukodystrophy, ataxia teleangiectasia, Bassen-Kornzweig disease.
- Late childhood (5-15 years of age): Friedreich's ataxia, Refsum's disease, Charcot-Marie Tooth disease, Roussy-Levy syndrome.
- All ages: Guillain-Barre' syndrome, subacute demyelinating neuropathy, chronic relapsing neuropathy, Tick paralysis, metabolic and toxic neuropathies.

Eng et al[23] studied electromyography in obstetrical brachial plexus palsy with conservative management with serial examinations. They found that electromyography was exceedingly useful in delineating the extent of involvement and recovery; while a correlation exists between clinical and electrodiagnosis findings.

Clinical Neurophysiology at The University Hospital Utrecht

Like many other hospital, the University Hospital Utrecht is confronted with a complete range of patients with neurological complaints. The diversity of complaints and clinical pictures are of real importance for medical training provided by the Division of Clinical Neurosciences. This Division consists of three medical areas : neurology, neurosurgery and clinical neurophysiology. The University Hospital Utrecht is a center for cerebrovascular disorders, neuromuscular diseases and the single center for epilepsy surgery in the Netherlands. The Department of Clinical Neurophysiology contributes to the studies in these fields.

Daily activities in this department consist of electroencephalography, cortical EEG during epilepsy surgery, brain mapping, transcranial Doppler (TCD), evoked potentials, and electromyographic examinations. There is advanced neurophysiological equipment such as for digital EEG, paper EEG, brain mapping, evoked potentials, transcranial Doppler, and electromyography. There are several experts for EEG, evoked potentials, and EMG with many technicians. The technicians have attended courses in neurophysiology for 3 1/2 years. One examines approximately per day 12 -15 patients with EEG, 1-2 patients per week with corticography, 2 - 4 patients per day with TCD, in 1-2 patients with EP, and in 8-10 patients with electromyography. In 1995[24] they examined 2250 patients with EEG (35% of all in children); 1845 patients with EMG (5% in children); 430 patients with EP (25% in children); 678 patients with TCD (2% in children). They also performed EEG monitoring on 101 patients undergoing carotid surgery, 40 patients undergoing AICD, 35 patients undergoing WADA test, and 40 patients undergoing arteriography. Several research projects are in progress such as on Guillain-Barre' syndrome, in polyneuropathy, neuralgia paresthetica; and transcranial Doppler in patients with cerebrovascular disorders. Modern imaging and physiological techniques have influenced concepts of cerebral function and permitted studies of anatomical-physiological correlation in patient with focal cerebral lesions. These data show that clinical neurophysiology at the University Hospital Utrecht can contribute to the study of neurological problems and benefit from imaging studies.

Training in neurology for medical doctor's in the Netherlands is 6 years that includes 15 months of clinical neurophysiology. In this training period a doctor must examine 500 EEGs, 250 EMGs, evoked potentials, and transcranial Doppler. At the end of their study, they must present 1 scientific paper and pass a National examination. The doctors that will be clinical neurophysiologist have a training of 27 months in clinical neurophysiology.

Electrodiagnosis in Pediatric Neurology in Indonesia

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ral Hospital in West Indonesia has two departments that perform neurophysiological examinations. The Department of Neurology examines adult patients, whereas the Department of Child Health examines children. They perform EEGs, evoked potentials, and electromyography except SEP in pediatrics.

The pediatric neurology division, which performs neurophysiology in children, has four experts in pediatric neurology with two pediatricians and four technicians. There is equipment such as 1 digital EEG apparatus, 1 paper EEG, 1 EP and 1 EMG machine, but there isn't one clinical neurophysiologist. These technicians haven't attended special courses for neurophysiology, in daily practice they learn from their doctors. Everyday, they perform 4-6 EEGs, 4-6 evoked potentials studies, and 1-3 EMG examinations. There is no formal neurophysiological and technological training in the field of clinical neurophysiology in Indonesia.

In the 10th Indonesian Pediatric Association Congress in Padang, West Sumatra, June 1996 52 papers concerning pediatric neurology were presented. Several diseases were studied such as: meningitis, encephalitis, febrile seizures, epilepsy, cerebral palsy, neuromuscular diseases, hydrocephalus, and tumors of the CNS. These data shows that EEG, SEP, BAEP, VEP and EMG should be in common use in pediatric neurology, but only 7 papers about those examinations were presented. These consists of 4 BAEP, 2 EEG, and 1 electromyography poster. Although EEG is commonly use in diagnostic procedures in children with seizures, unfortunately only 2 papers described the results of EEG examination. Electrodiagnostic papers were presented from 3 (25%) among 12 teaching hospitals in Indonesia. These data shows that electrodiagnostic examination is not in common use in pediatric neurology in Indonesia.

In the future, the incidence of patients with neurological infections may change to developmental retardation, epilepsy, heredodegenerative and neuromuscular diseases may become more important. Prevention with early detection of abnormalities of the CNS will be more prominent than treatment. The challenge from another ASEAN countries with high technology in medicine to improve our diagnostic, treatment and facilities. Electrodiagnostic studies may play an important role in pediatric neurology in Indonesia.

Conclusions and Suggestion

In the future, an increase in knowledge will be expected in health services. To face these coming problems, we must increase our knowledge and experience in electrodiagnostic examination in neurological problems in pediatrics.

Several ways are open to improve the quality of electrodiagnostic procedures such as : (1) training in electrodiagnostic examination for doctors and technicians; (2) improving an electrodiagnostic equipment for digital EEG, EPs and EMG; and (3) providing funds for maintenance the equipment.

The question arises how our teaching hospitals can find the fund for all these improvements. This is a big challenge for pediatric science, especially in child neurology in our country. So, why we don't try answer this challenge from now on.

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