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20TH ANNUAL MEETING OF PAKISTAN SOCIETY OF NEUROLOGY; BHURBAN, ISLAMABAD APRIL 2013

DETERMINATION OF RISK FACTORS LEADING TO THE DEVELOPMENT OF STROKE IN DIABETIC PATIENTS

Aftab Leghari, Hazim Brohi

Background: Stroke is a major preventable public health challenge. Diabetic patients constitute a fair percentage of patients with stroke. Numerous epidemiological studies in the United States and Europe suggest that one fifth of stroke patients have diabetes. We evaluate the risk factors leading to stroke in Diabetic patients.

Methods: A Cross sectional observational study was used that involved all confirmed diabetic patients for month's period from Jan to March 2012 at Jinnah medical College Hospital Karachi. Stroke Risk score card was used to identify the risk factors. Data was collected in the form of a questionnaire, Sugars and non fasting cholesterol was checked by test strips in clinic. The results were divided in three categories mid, moderate and severe factors to find out the most contributing factors.

Results: Out of the 167 sample size collected, 90 patients were confirmed to have diabetes, measured by Hb1ac levels ≥ 7 . Range of age was between 32-73 years. Eleven patients had low risk, while 77 had moderate risk. 2 had high risk. So our results showed that majority of diabetic patients are at moderate to high risk for stroke, approximately 97%. Analysis of risk factors showed that, raised BMI and lack of exercise were most contributing factors.

Conclusion: Diabetes were associated with moderate to high risk for stroke in 97% patients. Analysis of risk factors showed that raised BMI and lack of exercise were high contributing factors

ELECTROPHYSIOLOGICAL SUBTYPES OF GULLIAN BARRIE SYNDROME

Syed Arslan Haider, Adnan Mahmood, Athar Javed, M.Naeem

Introduction: Guillain-Barre syndrome (GBS) is an autoimmune polyradiculoneuropathy subdivided in demyelinating and axonal types. Electrophysiology plays a determinant role in Guillain-Barre syndrome (GBS) diagnosis, classification of the subtypes and in establishing prognosis.

Objective: To determine the frequency of different

electrophysiological subtypes in GBS.

Methods: This was a retrospective cross sectional study carried at department of neurology, mayo hospital, Lahore from Jan 01, 2012 to Feb 28, 2013. Patients fulfilling the Ashbury and Cornblath's clinical diagnostic criteria for GBS were included in the study. Exclusion criteria included all cases with asymmetric motor weakness, bladder or bowel dysfunction at onset, distinct sensory level suggestive of transverse myelitis. Data was entered in predesigned Performa and analyzed through SPSS 20.0.

Results: The study included 65 patients with mean age of $31.5 \text{ years} \pm 14.5$ (median 29 years); 41 (63%) males and 24 (36.9%) females, with a ratio M:F 1.7:1. Preceding history of infection was seen in 38 (58.5%). Patients with acute motor sensory axonal neuropathy (AMSAN) were 27 (41.5%), acute inflammatory demyelinating polyneuropathy (AIDP) 26 (40%), acute motor axonal neuropathy (AMAN) 06 (9.2%), pure sensory 06 (9.2%).

Conclusion: AMSAN was found to be the predominant electrophysiological subtype in our study followed by AIDP. AMAN and pure sensory subtypes were least common.

SMALL STEPS TAKEN BY MAN (IN PAST) THAT (BECAME) GIANT LEAPS FOR MANKIND (IN FUTURE)

Syed Ahmad Asif Assistant Professor & Consultant Neurologist, Liaquat National Hospital & Medical College, Karachi.

This is the story of evolution of Neurology. It is a timeline discussing important milestones in the field. First documentation of a neurological case was made by Sumarians, who showed, in a relief, a paraplegic lion with arrows in its back. Since then there have been so many 'first steps' that changed the face of neurology. In the pre-world war II era the neurologist and psychiatrist were making a combined move but after World War II, both the fields emerge as separate disciplines. Late Professor Munawwar Hayat is the Father of Neurology in Pakistan who first introduced it as a subject in 1960s. College of Physicians and Surgeons Pakistan first recognized Neurology, in 1989. Later it took 4 years for the first locally trained Fellow in Neurology, to emerge.

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CEREBRAL VENOUS THROMBOSIS IN ASIA

Mohammad Wasay MD, FRCP, FAAN Aga Khan University, Karachi

CVT is a well known but poorly reported entity. The largest data base of these patients included 624 patients. Most of the studies and registries related to CVT are reported from European countries. No large multi-center or multi-national data base or registry has been reported from Asian countries. CVT is not uncommon in Asia especially in south Asian subcontinent including India, Pakistan and Bangladesh. Pangayara

reported from India that CVT accounted for half of young stroke and 40% for stroke in woman. Review of CVT cases from Asian countries is suggestive of differences in risk factors profile and outcome in these patients as compared to European studies. Largest cohort of CVT patients from Europe (n=624) reported that 50% of these cases were related to OCP pills, 6% were due to pregnancy and 14% were secondary to puerperium. A study of 182 adult patients with CVT from USA reported 7% due to pregnancy and puerperium and 5% related to OCP use. A study from Pakistan (n=109) patients with CVT reported that 17% were due to pregnancy and puerperium and 5% related to OCP use. Cantu from Mexico reported 59% cases due to Pregnancy/ puerperium. Most patients with CVT do not undergo an extensive work up to identify cause of CVT. Outcome of these patients is also different from western countries.

CHEWED TOBACCO AS A RISK FACTOR OF STROKE

Rabel Khalid, Maimoona Siddiqui, Nadia Mehboob, Division of Neurology, Shifa International Hospital, Islamabad, Pakistan

Background: Stroke is the leading cause of long term disability worldwide. Besides the established stroke risk factors some potential risk factors related to lifestyle and dietary habits as huqqa smoking, use of oil and "ghee" and orally consumed forms of tobacco, exist in Pakistan but no data is available about the prevalence of the use of these products or their association with stroke.

Objective: To determine the frequency of use of chewed or smokeless tobacco in patients with ischemic stroke
Methods: This is a cross sectional study conducted in division of neurology, Shifa International hospital, Islamabad. All patients with ischemic stroke from June 2012 to February 2013 were enrolled in the study. A data collection form was filled and relevant information regarding risk factors was gathered. Data was analyzed using SPSS version 16.

Results: One hundred and seventy seven patients with ischemic stroke were enrolled, out of them 65% were male. Mean age was 62 years. Total smokers were 68 (38.4%). Out of these, 64 (94.1%) smoked cigarette and only 4 patients (5.9%) smoked huqqa. Chewed tobacco consumers were only 11 out of 177 (6.2%). Among them, the percentage of naswar addicts was 5.1%, while supari and paan addicts were 0.6% each. Among cigarette smokers, 32 (50%) had large vessel and 30 (46.8%) had small vessel disease. Out of 11 chewed tobacco consumers, 6 (54.5%) had large vessel while 5 (45.4%) had small vessel disease.

Conclusions:

Frequency of chewed tobacco was found in only 6.2% patients in our cohort. Further population based studies are needed to understand the association of chewed tobacco with stroke.

MS AND NMO IN PAKISTAN

*Mohammad Wasay MD, FRCP, FAAN
Aga Khan University, Karachi*

Multiple sclerosis is not uncommon in Asia. Japan, Israel and Middle Eastern countries have the highest prevalence of MS among Asian countries. Patients with MS in Asia share many similarities with the disease pattern seen in Western countries. Optic spinal MS, a distinct clinical and immunological entity is more common in Asia especially Far Eastern Asia. There are notable racial and geographical differences among various regions of Asia. MS in South and western Asia have more resemblance to Western type MS as compared to Asian type MS.

We describe retrospective data from the largest series from Pakistan of patients with multiple sclerosis (n=142). Mean age of onset was 27 years with a female to male ratio of 1.45:1. The disease onset was poly symptomatic in 75% patients. Motor weakness was the most common onset symptom (70%), followed by sensory symptoms (45%). Optico-spinal type of MS was seen in a very small proportion of patients (3%). The course was relapsing-remitting in 81%, primary progressive in 21% and secondary progressive in 4% patients. Almost three fourths of the patients were moderately (45%) or severely (31%) disabled at the time of evaluation. Two-thirds of patients with severe disability had the disease duration of only 5.2 years.

There are no studies related to incidental MS in Pakistan. We conducted a study to identify asymptomatic patients with brain MRI lesions suggestive of MS in a low-prevalence area of Pakistan. Brain MRIs for 864 patients were reviewed at Aga Khan University Karachi during an eight month period of 2006 and 2007 to identify patients with lesions suggestive of MS. Six (2 females) patients out of 864 (0.7%) fulfilled brain MRI criteria suggestive of MS. The mean number of MRI lesions (total lesions on T2) were 9 (range 5-14). There has been no previous study from Pakistan regarding genetic predisposition for MS with certain HLA types. Another study was conducted to analyze genetic makeup of MS patients as compared to a control population. This study showed that frequency of certain HLA types in Pakistani MS patients was not different from population.

Two more studies underway to study the rate of progression of disability in a cohort of MS patients. Another study is underway for telephonic validation of EDSS in Pakistani MS population.

PRIONS AND NEUROLOGICAL DISEASE

Saad Shafiqat Professor of Neurology, Aga Khan University, Stadium Road, Karachi 74800, Pakistan

Prions are infectious proteins devoid of DNA or RNA. They are responsible for a range of neurological illnesses in humans and other species.

The ability of prions to cause disease results from their capacity to induce pathological conformational change in the native isoform of the prior protein, PrPC. It is considered an infection because the transmissibility pattern satisfies Koch's postulates. The pathological isoform PrPSc is identical in amino-acid sequence to PrPC, but has a secondary structure rich in β -sheets. Disease-associated mutations have also been described in the PrPC gene that impart excess β structure and result in familial forms of prion disease. The cellular function of normal PrPC is unknown.

The classic human prionosis is Creutzfeldt-Jakob disease (CJD), a late-onset, rapidly progressive multi-domain dementia with prominent myoclonus that exists in sporadic, familial, iatrogenic, and variant forms. The classic non-human prionosis is scrapie, a lethal ataxia of sheep. Prion infection is also recognized in cats, deer, elk, mink, and especially in cows (so-called "mad cow disease"). Other human prionoses include kuru, Gerstmann-Strausler-Schienker disease, and fatal familial insomnia - all rare but distinct neurological syndromes. Variant CJD has public health significance as all known cases are traceable to contaminated beef exposure during the 1980s in Britain.

The discovery of prions represents a major intellectual advance in which separate lines of investigation starting from scrapie and CJD converged in a series of ground-breaking experiments, resulting in two Nobel Prizes (for Carlton Gajusek and Stanley Prusiner) and scientific confirmation of an unprecedented form of disease pathogenesis primarily afflicting the nervous system.

SPECTRUM OF CNS TUBERCULOSIS IN PATIENTS PRESENTING IN A TERTIARY CARE HOSPITAL

Dr. Zahid Abbas, Dr. Adnan Javed Leghari, Dr. Athar Javed, Dr. Muhammad Naeem Kasuri, Dr. Mohsin Zaheer.
Department of Neurology, Mayo Hospital, Lahore.

Introduction: The involvement of central nervous system

(CNS) is an important and serious manifestation of extrapulmonary tuberculosis (TB). It occurs in approximately 10% of all tuberculous patients. TB of the CNS can have varied manifestations of which the most commonly seen are: Intra-cranial manifestations such as; tuberculous meningitis (TBM), tuberculous encephalopathy, and space-occupying lesions called tuberculomas; Spinal form are tuberculous spondylitis with or without compressive myelopathy, tuberculous arachnoiditis (myeloradiculopathy) and spinal tuberculomas.

Objective: To determine the spectrum of CNS tuberculosis in patients presenting in a tertiary care hospital in Lahore.

Methods: This was a retrospective cross sectional study carried out at neurology department Mayo hospital Lahore from 1st January 2012 to 31st December 2012. Medical records were reviewed. History, clinical examination and investigations including CSF examination and neuroimaging were entered on specified Performa. Patients fulfilling the criteria for various manifestation of CNS tuberculosis were included in the study. The data was entered and analyzed using SPSS v20.

Results: A total of 72 patients with various manifestations of CNS tuberculosis were identified and included in the study. Mean age of presentation was 35.51 ± 16.735 . Out of the 72 patients, 55.6% (n=40) were male and 44.4% (n=32) were females. TBM alone was the most common manifestation representing 41.7% (n=30) of total patients, TBM with Tuberculoma 11.1% (n=8), tuberculoma brain either single or multiple were presents alone in 8.3% (n=6). TBM with Infarction 6.9% (n=5) and TBM with tuberculous arachnoiditis 5.6% (n=4). In spinal tuberculosis, all the patients in our series were of compressive myelopathy. Tuberculous Spondylitis with or without abscess was seen in 26.4% (n=19). Pulmonary tuberculosis (PTB) was present in 19.4% (n=14) of total patients. Interestingly all our patients suffering from PTB had TBM as a neurological manifestation.

Conclusion: Tuberculous meningitis is the most common form of intracranial tuberculosis and tuberculous spondylitis with or without abscess is the most common form of spinal tuberculosis. While all patients with history of active or old pulmonary tuberculosis presenting with CNS involvement had tuberculous meningitis only.

IMMEDIATE SYMPTOMATIC RELIEF AFTER MICROVASCULAR DECOMPRESSION FOR HEMIFACIAL SPASM: A CASE SERIES

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Department of Neurosurgery, Postgraduate Medical Institute, Lady
Reading Hospital Peshawar, Pakistan.

Objective: To assess the surgical outcome of microvascular decompression (MVD) for Hemifacial spasm (HFS).

Material and Methods: This descriptive study was conducted in Neurosurgery Department of PGMI, Lady Reading Hospital Peshawar from January 2011 to June 2011. A total of 4 patients with HFS who underwent MVD via a retrosigmoidcraniectomy are presented. The root exit zone (REZ) of the facial nerve was exposed, the offending vessels were identified intra-operatively and procedure was performed using surgical prosthesis. All the data was collected by using a Performa and all the patients were followed up for a minimum period of 4 weeks.

Results: Out of 4, 3 were males and 1 was female. We had follow-up of all patients for up to 4 weeks. Three patients had complete symptomatic relief within 24 hour of surgery, while in 1 patient relief observed after 2 days and minor problem in hearing (sensorineural hearing loss) occurred, which improved after 4 weeks. All of them were satisfied with the procedure. A causative vessel was found on the root exit zone of all the patients, 3 had anterior inferior cerebellar artery and 1 had basilar artery. There was no mortality in the series.

Conclusion: All the patients of hemifacial spasm got symptomatic relief with MVD, but minor complication (partial hearing loss) occurred in single patient.

SWALLOW THERAPY FOR DYSPHAGIA

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Introduction: Swallowing disorder is a difficulty moving food from mouth to stomach. Swallowing disorders occurs in all age groups, from newborn to the elderly. It may be temporary and permanent. 90% patients after any cerebrovascular accident encounter to some instance of swallowing disorder. The most effected stages are oral and pharyngeal stage of swallow. To deal with dysphagia is one of the important roles of speech language therapist. Basically swallow therapy includes techniques for reducing any delay in triggering the oral and pharyngeal swallow and improving pharyngeal transit time.

Thus, the term swallowing refers to the entire act of deglutition from placement of food in the mouth through the oral, pharyngeal and esophageal stages of swallow until the material enters the stomach.

Case presentation: A 57 years old male patient came to rehabilitation unit with the history of pontine infarct 15 days back with the complain of slurring of speech and dysphagia. He was fed by NG tube.

MRI Findings: Acute Pontine Infarct

Barium Swallow: right pyriform fossa pooling

Dysphagia Assessment: A detail assessment of swallowing revealed:

Delayed bolus transit time, No anterior and posterior propulsion, Food residue after eating, Gurgly and wet voice quality, The patient has pharyngeal dysphagia with all diet and liquids level. Frequency of therapy was decided four times a week.

Treatment plan: Therapy plan was consist on three important techniques oral motor stimulation, postural maneuvers and diet modifications.

Result: With the help of these techniques he will able to take partial oral feed within one week. After 2 weeks we were able to take regular food but feel a little difficulty in thin liquid diet. The therapist decides to modify the thin liquid into level two which is shows no difficulty. And at this stage the NG tube were removed. It shows that the speech therapy really works with the patients who have difficulty in swallowing.

ELECTROPHYSIOLOGICAL PATTERNS IN DIABETIC NEUROPATHY

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Department of Neurology, King Edward Medical University/Mayo Hospital, Lahore.

Background: Neuropathies are characterized by a progressive loss of nerve fiber function and is caused by a number of diseases. 'Diabetes Mellitus (DM)' remains one of the most common causes of peripheral neuropathy worldwide. Diagnosis of 'Diabetic Peripheral Neuropathy (DPN)' is based both on clinical and electrophysiological basis.

Aim: To study the frequency of various 'Electrophysiological Patterns' in patients with symptomatic 'DPN'. Study design: It was a cross sectional study based on non-probability purposive sampling technique.

Venue & duration of study: Study was conducted at the Department of Neurology, Mayo Hospital, Lahore, over a period of one year (from March, 2012 to February, 2013).

Methodology: One hundred and eighty eight patients (n=188) of diabetes mellitus (Type 1 or 2) with symptomatic neuropathy, both male and females, were selected from amongst those visiting the hospital outpatient department. Patients with neuropathies due to hereditary cause, renal or thyroid disease, alcoholism or toxin / drugs, those with nutritional deficiencies, malignancy or an autoimmune disorder were excluded from the study. After taking consent, patients were subjected to electrophysiological studies. Analysis of data was done using SPSS-16 (Independent-Samples T-Test).

Results: We collected 188 patients with diabetic peripheral neuropathy. Out of these, there were 88 (46.81%) male cases and 100 (53.29%) were female. Mean age of sample was 52.69 yrs. (+ 9.91). Male gender had a mean age of 53.20 yrs. (+ 10.13) while it was less in females [52.25 yrs. (+ 9.75)]. Age of the sample had no significant association ($p < 0.498$) with the electrophysiological patterns.

Regarding frequency of various electrophysiological patterns in DPN, 'sensory-motor mixed polyneuropathy' (100 cases i.e. 53.2%) was the commonest electrophysiological pattern. Among these 100 cases, 94/100(94%) had a predominantly axonal & 06/100 cases (6%) had a predominantly demyelinating type of sensory-motor mixed polyneuropathy. Second commonest electrophysiological pattern (41 cases i.e. 21.8%) was that of 'sensory axonal polyneuropathy'. CTS was placed at third position (22 cases i.e.11.7%). A total of 16 diabetics (8.5%) had a normal electrophysiology. These four subgroups constituted more than 95% of the entire electrophysiological profile. Rest of the profile depicted minor electrophysiological patterns of neuropathy like 'sensory motor axonal' (4 cases i.e. 2.1%); 'motor axonal pattern' was seen in 3 cases (1.6%) & 'radiculopathy' seen in 2 diabetics (1.1%). Considering the electrophysiological patterns with reference to gender, 53/100 male (53%) & 47/100 females (47%) had the commonest neuropathic pattern i.e. sensory-motor mixed polyneuropathy. Second commonest electrophysiological pattern i.e. sensory axonal polyneuropathy was exhibited by 16/41 male (39.03%) & 25/41 females (60.97%). CTS was more frequent in females (21/22 i.e.95%) than in male gender (1/22 i.e. 4.55%). There exists a strong

association of CTS with female gender ($p < 0.017$). However, while considering the entire sample, statistics do not yield any significant relationship ($p < 0.087$) between gender & rest of the entire electrophysiological profile.

In the study sample, electrophysiological patterns had no statistically significant relationship with either mean 'age of onset of DM' [42.07 yrs. (+ 11.10); ($p < 0.808$)], nor with 'age of onset of DPN' [50.18 yrs. (+ 10.27); ($p < 0.889$)]. Furthermore, relationship between mean 'duration of DM' [10.65 yrs. (+ 6.87); ($p < 0.150$)] and the electrophysiological spectrum was also insignificant. Importantly, there existed a statistically significant relationship between 'duration of DPN symptoms' [2.38 yrs. (+ 2.59)] and the electrophysiological profile ($p < 0.048$).

Conclusion: DM is more prevalent in female gender. Mean age of female gender in the sample is less than male. This implies an early 'onset of DM' in female than in male gender. Commonest electrophysiological pattern seen in diabetics is 'sensory motor mixed (predominantly axonal) polyneuropathy. Second commonest pattern is 'sensory axonal polyneuropathy'. 'Duration of symptoms of DPN' is positively associated with the presence of various electrophysiological patterns. No electrophysiological pattern has statistically significant relationship with age of onset & duration of DM or onset age of DPN. Even patients symptomatic of DPN can have normal electrophysiology.

SPECTRUM OF ANTERIOR HORN CELL DISORDERS IN PATIENTS PRESENTING TO A TERTIARY CARE HOSPITAL

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Aim: To review the spectrum and epidemiological characteristics of patients diagnosed as suffering from Anterior Horn cell disorders.

Methods: This was a retrospective cross sectional study carried out at Neurophysiology lab of King Edward Medical University/Mayo hospital Lahore during one year from 1st January 2012 to 31st December, 2012. The record of all the patients undergoing Electromyography & nerve conduction studies during the last one year was reviewed. Patients fulfilling criteria to be diagnosed as suffering from anterior horn cell disorders were selected and their clinical & Electrophysiological record reviewed. These were further subdivided into those suffering from Motor Neuron

though rare but constitute less than 5% of hospitalized patients with equal male and female involvement. CIS remained most common disorder followed by MS. ADEM in adults is rare.

SPONTANEOUS THORACIC SPINAL EPIDURAL HEMATOMA IN THE POST-PARTUM PERIOD

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Spontaneous spinal epidural hematoma (SSEH) is a rare cause of neurological emergency in pregnancy and postpartum period. This complication has previously been published very rarely in the postpartum period. We report the development of spinal epidural hematoma resulting in acute onset paraplegia in a 30 year-old woman after prolonged, assisted vaginal delivery. There was no history of any predisposing factors like coagulopathy, anticoagulation, trauma, vascular anomaly, vasculitis and iatrogenic manipulations such as spinal/epidural anesthesia. Magnetic resonance imaging revealed changes consistent with an epidural hematoma extending from D2-D4. An emergency spinal decompression and laminectomy was done, epidural hematoma was evacuated. Due to delayed recognition and intervention, her neurological functions did not improve significantly.

Keywords: epidural hematoma, postpartum period, acute onset paraplegia, assisted vaginal delivery, spinal decompression and laminectomy.

CHRONIC RELAPSING INFLAMMATORY OPTIC NEUROPATHY (CRION)

*Muhammad Athar Javed
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Introduction: The newly recognized entity named 'Chronic Relapsing Inflammatory Optic Neuropathy (CRION)' is a form of inflammatory optic neuropathy which is frequently bilateral and often painful and is characterized by relapses and remissions. Neuroimaging and cerebrospinal fluid examination for oligoclonal band is normal. The syndrome behaves in a way which is typical of the condition known as granulomatous optic neuropathy without systemic evidence of sarcoidosis.

Objective: To study the clinical features and response to treatment in patients with recurrent optic neuritis consistent with diagnosis of chronic relapsing inflammatory optic neuropathy (CRION).

Materials and Methods: This is retrospective cross

sectional study carried out at department of Neurology, King Edward Medical University, and Lahore. Patients with history of recurrent episodes of subacute loss of vision along with pain consistent with optic neuritis (unilateral or bilateral) were included in the study. Those with evidence of an acquired demyelinating disorder, systemic collagen vascular and granulomatous disease were not included in the study. The response to various treatments was also analyzed.

Results: A total of 4 patients were identified. All were females with mean age at presentation 35.50+8.10. Mean age at the onset of first episode was 28+10.92. Mean duration of illness was 7.25 + 4.57 yrs. The number of episodes varied from 3-6 (mean 4+1.41 episodes). Mean episodes of right optic neuritis were 2.25+.95 and left side 1.50+ 0.57. One patient had one episodes of simultaneous bilateral optic neuritis.

All patients experienced severe pain with loss of vision to finger counting at less than one meter. MRI brain, detailed vasculitic profile, ACE levels, X-ray chest were normal and CSF for oligoclonal band was negative in all patients. All patients received pulse therapy with methylprednisolone 1 gm daily for three days followed by two weeks oral taper. Most patients improved after first episodes but there was partial recovery after the second episodes. Only one patient had complete loss of vision in one of her eye. Two patients received long term oral steroids with azathioprine after the third and sixth episodes with prevention of further relapses. One patient received beta interferon after the 3rd episode with complete remission so far. One patient received cyclosporine and oral steroids with remission for 6 years but there was subsequent relapse after immunotherapy was discontinued.

Conclusion: Chronic relapsing inflammatory optic neuropathy (CRION) should be considered in patients with history recurrent optic neuritis without evidence of acquired demyelinating disorder, systemic collagen vascular or granulomatous disease.

ROLE OF IMAGING IN DIAGNOSTIC WORKUP OF ACUTE CEREBROVASCULAR ACCIDENTS.

*Dr. Tahir Malik
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Stroke is a leading cause of mortality and morbidity in the developed world. The goals of an imaging evaluation for acute stroke are to establish a diagnosis as early as possible and to obtain accurate information about the intracranial vasculature and brain perfusion for guidance in selecting the appropriate therapy. A

comprehensive evaluation may be performed with a combination of computed tomography (CT) or magnetic resonance (MR) imaging techniques. Unenhanced CT can be performed quickly, can help identify early signs of stroke, and can help rule out hemorrhage. CT angiography and CT perfusion imaging, respectively, can depict intravascular thrombi and salvageable tissue indicated by a penumbra. These examinations are easy to perform on most helical CT scanners and are increasingly used in stroke imaging protocols to decide whether intervention is necessary. While acute infarcts may be seen early on conventional MR images, diffusion-weighted MR imaging is more sensitive for detection of hyperacute ischemia. Gradient-echo MR sequences can be helpful for detecting a hemorrhage. The status of neck and intracranial vessels can be evaluated with MR angiography, and a mismatch between findings on diffusion and perfusion MR images may be used to predict the presence of a penumbra. The information obtained by combining various imaging techniques may help differentiate patients who do not need intravenous or intra-arterial therapy from those who do, and may alter clinical outcomes.

UPDATE ON TIZANIDINE FOR MUSCLE SPASTICITY AND EMERGING INDICATIONS.

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Background: Tizanidine hydrochloride, an α_2 -adrenergic receptor agonist, is a widely used medication for the treatment of muscle spasticity. Clinical studies have supported its use in the management of spasticity caused by multiple sclerosis (MS), acquired brain injury or spinal cord injury. It has also been shown to be clinically effective in the management of pain syndromes, such as: myofascial pain, lower back pain and trigeminal neuralgia.

This review summarizes the recent findings on the clinical application of tizanidine. Objective: Our objective was to review and summarize the medical literature regarding the evidence for the usefulness of tizanidine in the management of spasticity and in pain syndromes such as myofascial pain.

Methods: We reviewed the current medical and pharmacology literature through various internet literature searches. This information was then synthesized and presented in paragraph and table form.

Results: conclusion: Tizanidine hydrochloride is a very useful medication in

patients suffering from spasticity caused by MS, acquired brain injury or spinal cord injury. It can also be helpful in patients suffering from chronic neck and/or lower back pain who have a myofascial component to their pain. Doses should be started at low dose and gradually titrated to effect.

SKIN RASH WITH ANTIEPILEPTIC DRUGS: LOW OCCURRENCE IN PAKISTAN

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Background: It has been an anecdotal observation, that skin rash associated with use of antiepileptic drugs (AEDs) in Pakistan is less than that reported in world literature.

Purpose: To study the occurrence of skin rash with antiepileptic drugs in patients.

Methods: Retrospective analysis of patients' records at National Epilepsy Centre was done to determine occurrence of skin rash in people with epilepsy taking atleast one AED. Patients with epilepsy registered between April 2009 and September 2011 were included in the study. Results were compared with those reported in published literature.

Results: Of the total 2063 patients, 14 patients (0.68%) reported skin rash; non-serious skin rash in 09, Steven Johnson syndrome in 03, Drug Rash with Eosinophilia and Systemic Symptoms (DRESS) in 01 and Toxic Epidermal Necrolysis (TEN) in 01. The patient with TEN expired. Nine of the 11 patients were on monotherapy whilst 05 on polytherapy. Carbamazepine was found to be most responsible.

Conclusion: AED-associated skin rash in Pakistan is much less (0.68%) than that reported in other countries like USA (14.3%), Norway (14%), Canada (8.5%) and China (3.6%). We hypothesize that this variability may be due to ethnic / racial dermatological features. Further information from other countries' research is required to substantiate or refute the hypothesis.

REVERSIBLE ELECTROPHYSIOLOGICAL ABNORMALITIES IN HYPOKALEMIC PERIODIC PARALYSIS

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Compound muscle action potential (CMAP) amplitude declines during a paralytic attack in patients with hypokalemic periodic paralysis (HPP). However, serial motor nerve conduction studies in HPP have not been

commonly reported.

We report a 32 years old male with HPP, who had severely reduced CMAPs in all motor nerves at presentation. During the episode of generalized weakness the serum potassium level at that time was 2.2 mmol/L. However, the amplitude of CMAPs improved and reached normal levels as the serum potassium concentration (4.7mmol/L) and motor power returned to normal.

During the attack nerve conduction study showed significant reduction in CMAP amplitudes of all motor nerves. However, the distal latencies, conduction velocities and F-wave latencies were within normal limits. Sensory nerve action potential (SNAP) amplitudes and electromyography (EMG) were also normal.

We report multiple episodes of low serum potassium associated with clinically generalized weakness and electrophysiologically low CMAPs.

To our knowledge such repeated episodes of low serum potassium, low power, low CMAPs and everything improving after biochemical correction of serum potassium has not been reported from this area. (Later patient was put on Acetazolamide and has not returned even after 9 months.

NEUROPSYCHOLOGICAL ASPECTS OF CONVERSION DISORDER. A CASE STUDY

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Introduction: Conversion Disorder is a type of somatoform disorder exhibit with voluntary sensory and motor functioning without any medical reasons and is thus referred to as "Pseudoneurological". Symptoms may also include seizures and convulsions. Diagnosis of conversion disorder should be made only after thorough clinical investigations, to rule out an etiological, neurological or general medical condition. These problems generate in the result of patient's life stress and depression. Conversion is considered a psychiatric disorder in the Diagnostic and Statistical Manual of Mental Disorder 4th Edition (DSM-IV). The onset of Conversion Disorder is generally from late childhood to early adulthood rarely before age 10 years or after age 35 years

Case: A 23 years old girl admitted in Neuro Medicine ward referred by neurologist for Neuropsychological

Evaluation. At the time of admission the patient was unconscious and she remained unconscious for 2 days. Initial Neuro Diagnostic (EEG, MRI) work up which reveals no medical cause of unconsciousness. She felt weakness before the fits. She had already past psychiatric history. Her mother reported that when she was 11 years old she scared in a park and got unconscious and from the age of 11 years she was taking anti psychotic drugs.

After detail Neuropsychological Assessment she diagnosed as having Conversion Disorder. Assessment Findings: The assessment finding revealed that the patient needs therapeutic measures for

Depression, Unrealistic Expectations, Negative Self Assessment, Lack of Social Interaction.

Along with medicines she was enrolled for psychotherapy which includes psychodynamic formulation, non confrontational supportive psychotherapy, reassurance and suggestion. Family psychotherapy works a lot. Cognitive Behavioral Therapy for positive thinking and positive life goals. Problem solving therapy changed the areas of her life. Stress management training to overcome stress in range of situations. After the intensive therapy of 6 months now she is able to perform well and teaching at school.

Conclusion: Neuro psychological rehabilitation can change life endeavors of patients with conversion disorder.

PSYCHOSOCIAL FEATURES OF ASPERGER'S DISORDER: A CASE STUDY

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Introduction: Asperger Disorder is one of the type of pervasive development disorder manifested by significant difficulty in the acquisition and use of maintaining eye contact, social skills, pragmatics language along with stereotyped behavior (like repetitive patterns of behavior), and limited range of interest and co-ordinations problems. It differs from other Autism Spectrum Disorder by its relative preservation of linguistic and cognitive development. As per Diagnostic & Statistical Manual of Mental Disorder (4th Edition) "Asperger Disorder is characterized by stereotyped behaviors and interests and by more severely impaired social interaction".*

Case Presentation: 7 years old boy was referred for

the purpose of Neuropsychological Assessment. He came with the complained of week memory, learning disability, shy behavior and hyperactivity with unoccupied play. After a detail assessment, comprehensive observations and assessment batteries he was diagnosed as Asperger's disorder.

His assessment findings revealed following areas for management:

Social Communication, School Academics
Social Interaction, Behavior eruption, Play.

Frequency of therapy was five days a week. Parental counseling which addressed to minimize their expectations to realistic means and assist in setting realistic expectations and to get rid from the idea of over protectiveness. Parent should view him as a unique individual, with potential abilities to make choices of his own were played a vital role to adjust him in the home environment. Multimodal therapy and Cognitive behavior therapy were used to improve his dependency, learning helplessness, poor self image, and judgment. Remedial teaching which include functional academics and curriculum shaping were tremendously made a difference in his over all performance, acceptance and adjustment in mainstream school. Now he is on Parallel play. Peer group therapy sessions, decreased his pragmatic communication barrier.

Conclusion: Neuropsychological remediation can make a difference in Asperger rehabilitation and increase learning abilities.

TOTAL NEUROLOGICAL SURVEY OF PAKISTAN- INTERIM REPORT

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PURPOSE: To assess prevalence of all neurological diseases in Pakistan.

METHOD: It is a door-to-door, urban and rural, epidemiological study in Karachi, Khairpur, Rahim-Yar-Khan and Sialkot. Random block selection of the sample size was made in 2006 using 1% prevalence of epilepsy in Pakistan (1987 survey) and assuming 9% prevalence of migraine in each survey city. The screening questionnaire is the Urdu translation of WHO screening questionnaire for neurological disorders used by Bharucha et al. Demographic information and disease specific information were collected.

RESULTS: Results of Karachi and Khairpur are being presented. Out of a total of 10714 persons screened 1162;10.9% (male 329;6.1%, female 833;15.8%) had neurological disease. The disorders include: migraine-headache 321(3.0%), non-migraine-headache 598(5.6%), Epilepsy 82(0.8%), Stroke 57(0.5%), Parkinson's disease 5(0.05%), dementia 3(0.03%) polyneuropathy 48(0.5%), and other disorders 48(0.5%).
CONCLUSION: Prevalence of neurological diseases in southern Pakistan is lower compared to that in the literature. Prevalence of epilepsy has a little decreased compare to the 1% in 1987 survey.

GLUE SNIFFING NEUROPATHY AND REVIEW OF LITERATURE

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Glue sniffing neuropathy commonly known as n hexane neuropathy. It is well documented that industrial exposure to n-hexane causes neuropathy, however it is less well recognized that inhalation of n-hexane present in the vapors can also cause neuropathy. However such patients are not seen that frequently. The acute worsening also generates differential diagnosis of GBS. Most of literature is reported from west. We report such case for the first time from Saudi Arabia. A 35 year old male presented to us with progressive numbness followed by weakness in both legs since last three weeks. Over next two week he became chair bound and in the beginning of third week he also stated to feel numbness in both the hands and some weakness was also noted in hands. His past history was significant for Carpet cleaning glue sniffing for many years. His exam was significant for distal weakness feet greater than hands, deep tendon reflexes were absent all over. All sensory modalities showed glove and stocking pattern. Nerver conduction velocities showed slowing. His CSF exam was normal. We conclude that n-hexane is neurotoxic when inhaled to excess and, that the neuropathy has characteristic electrophysiological and pathological features.

TRANSFUSION THERAPY AS A SOLE TREATMENT OPTION IN A THALASSEMIA PATIENT WITH ACUTE PARAPLEGIA- A CASE REPORT AND REVIEW OF LITERATURE

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Thalassaemia intermedia is one of the haemoglobinopathies in which extramedullary hematopoiesis occur at various sites in the body as a compensatory mechanism to combat long standing anemia. Spinal haematopoiesis is an extremely rare phenomenon in such patients. Compressive myelopathy

resulting from these pseudo tumors require immediate attention. Various modes of therapy are available. Our patient with Thallassemia intermedia who developed paraplegia due to spinal haematopoiesis was successfully treated with blood transfusion alone.

FITS OR CONVERSION DISORDER. A CASE STUDY

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Introduction: Conversion Disorder is a neurosis which is known as hysteria or malingering. In conversion Disorder, the physical symptoms such as pain, paralysis, fits and coma without any medical cause but in the presence of stress. This is seen that these problems generate in the result of patient's life stress and depression. Conversion is a considered a psychiatric disorder in the Diagnostic and Statistical Manual of Mental Disorder 4th Edition (DSM-IV). The onset of Conversion Disorder is generally from late childhood to early adulthood rarely before age 10 years or after age 35 years

Case: A 23 years old girl admitted in Neuro Medicine ward referred by neurologist for Neuropsychological Evaluation. At the time of admission the patient was unconscious and she remained unconscious for 2 days. Initial Neuro Diagnostic (EEG, MRI) work up which reveals no medical cause of unconsciousness. She felt weakness before the fits. She had already past psychiatric history. Her mother reported that when she was 11 years old she scared in a park and got unconscious and from the age of 11 years she was taking anti psychotic drugs.

After detail Neuropsychological Assessment she diagnosed as having Conversion Disorder. Assessment Findings: The assessment finding revealed that the patient needs therapeutic measures for Depression, Unrealistic Expectations, Negative Self Assessment, Lack of Social Interaction.

Therapeutic Management: Along with medicines she was enrolled for psycho therapy, the frequency of therapy was thrice a week. Parent's counseling works a lot. Cognitive Behavioral Therapy for positive thinking and positive life goals. Problem solving therapy changed the areas of her life. Stress management training to overcome stress in range of situations. After the intensive therapy of 6 months now she is able to perform well

and teaching at school.

CHILDREN WITH HEARING IMPAIRMENT CAN LEARN TO TALK

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Introduction: Cochlear implant is surgically implanted device which serves to bypass the damaged cochlea to directly stimulate the auditory nerve (CN VIII). The ideal candidate for cochlear implant are children from 1 year to 5 years old and have severe to profound hearing loss with cognitive ability and have received minimal to no benefit from traditional amplification. Case: A 2.4 year old child with hearing impairment was referred for speech therapy by audiologist in neurorehabilitation unit. Have cochlear implant surgery done before one month. After internal component healing he was referred for speech therapy. Detail case history revealed significant history of hearing losses in family and have congenital hearing impairment. He was clinically diagnosed at the age of 1.3 years. His BERA shows profound hearing loss bilaterally. Assessment Findings: His assessment finding revealed no auditory skills, his communications mainly consist of signs and gestures, was lip read perfectly, his verbal communication was based on jargon speech. His vocabulary includes mama and papa only. Rely more on visual input. Socially he was not mingled with his age mates. No pragmatics language was present except gestural greeting. Segmental and suprasegmental errors were present. No response in identification testing, analysis of speech reading and synthetic speech reading test. Verbal and nonverbal communication was restricted to his parents only.

Auditory training, Vocabulary building, Increase mean length utterance, Decrease segmental errors. Present Level of Functioning.

Now he can pick almost all auditory stimuli. Comprehend all verbal commands. Communicate in full length sentence with minimal syntactical errors. Enroll in mainstream school and adjust well. Enjoy company of age mates and class fellows. Now he have 65% segmentally and 40% supra segmentally intelligible speech. Now he is 80% intact on identification test, 50% in speech reading analysis and 20% at synthetic speech reading. Although he still face challenges in pragmatics skills but now his physical and verbal pragmatics is within functional limits. His Verbal and

nonverbal communication is still restricted to familiar communication partners.

Conclusion: Speech therapy makes the difference in speech language development of cochlea implant child.

PRAGMATICS LANGUAGE: QUALITATIVE ASPECTS OF LANGUAGE

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Introduction: Language is the innate feature of every new born. Initially crying, cooing, babbling are communications which excel through developmental period. They convert into word spurts and are turned into nouns, and later on, convert into sentence structure. Language has five components: phonology, morphology, syntax, semantics and pragmatics. The important feature of language is pragmatics which is social use of language. Language according to social scenarios and circumstances is the skill which requires adjusting and accommodating in the environment. To deal with the environment linguistically a person has to have verbal, physical and thinking pragmatics. Topic revival, topic maintenance, join group situation, and so on are the key features of pragmatics skills. Mostly patient with language delay may cope up with their expressive and receptive skills but even after language attainment patient have difficulty in using language according to socially accepted manner. They do have vocabulary, syntax formation, sentence understanding but they may fail in physical pragmatics and thinking pragmatics.

Objectives: To find out incidence of pragmatics delay in children with speech and language disorder. To explore which type of pragmatics delay is higher. **Materials and Methods:** Data was retrospectively drawn from out patient department of Liaquat National Hospital and Neuro Clinic and Falij Care of speech language pathology services, the age range was 2.5 years to 10 years. Patients were screened out in the study through case history, observation, formal and informal assessment tools.

Results: Data of 100 patients with the diagnosis of speech and language delay was collected for analysis. 31% patients had verbal pragmatics. 51% patients had physical pragmatics while 18% patients had thinking pragmatics.

Conclusion: The following study shows that thinking

pragmatics is the area which is severely affected in children with language delay.

POST TRANS-SPHENOIDAL PITUITARY SURGERY MENINGITIS PRESENTING AS STROKE

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Introduction: Cerebral vasospasm is a recognized but rare complication of trans-sphenoidal pituitary surgery secondary to intraoperative subarachnoid hemorrhage. It can also be a manifestation of post-operative meningitis. Regardless of the pathology, it can lead to serious complications with irreversible brain injury in the form of stroke.


Case description: We report a case of 70 year old lady who presented with a history of sudden onset of right sided weakness and facial deviation towards left side for 2 hours. She had a history of pituitary surgery in 2008 and a re-do pituitary surgery 20 days back. On examination, higher mental functions were normal with right upper motor neuron type facial palsy and right hemiparesis (power 0/5 in right arm and 2/5 in right leg) with extensor plantar response. MRI brain showed an acute lacunar infarct in right internal capsule with irregularity and beading of both ACA's and MCA's suggestive of cerebral vasospasm or vasculitis. On 2nd day of admission, she developed disorientation and confused speech. CSF analysis was suggestive of partially treated bacterial meningitis (csf finding). She was managed with IV ceftriaxone and vancomycin in meningitic doses. Headache and confusional state improved and GCS was 15/15 at the time of discharge.

Conclusion: Meningitis after transsphenoidal pituitary surgery is extremely rare, and can lead to life altering experiences for the patient in the form of stroke. It often goes unrecognized and high index of suspicion is required to diagnose it as clinical examination, CSF findings and inflammatory markers are not reliable due to prior surgery. Its prompt recognition and adequate treatment is essential to prevent severe ischemic brain damage.

BARDETBIEDL SYNDROME IN A 19 YEARS OLD BOY PRESENTING WITH ATAXIA, BODY ACHES AND GENERALIZED WEAKNESS

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Introduction: Bardetbiedl syndrome is a rare autosomal recessive condition which can manifest with a variety of symptoms. It is characterized by six primary features



and many secondary features. Primary features include obesity, mental retardation, polydactyly, retinal disturbances, renal malformations and hypogenitalism. Secondary features include conductive hearing loss, recurrent otitis media, reactive airway disease, asthma and congenital heart disease among others. We present the case of a 19 years old boy presented to Neurology department at Northwest general hospital, Peshawar.

Case description: A 19 years old boy presented to us with complaints of ataxia, generalized weakness, body aches and nausea. His visual acuity was decreased since childhood. His developmental milestones were delayed and he also had learning disabilities. He was also complaining of polyuria since childhood. He was hexadactyl in all four limbs and was obese for his age and height. A diagnosis of Bardet-biedl syndrome was made and the patient and family were counseled regarding the condition after appropriate management.

Conclusion: Bardet-Biedl syndrome is a rare condition but recent studies show that it may not be as rare as was once thought. Early diagnosis is important to improve the quality of life of affected individuals. There was delay in diagnosis in the case of this patient. Because of the variety of symptoms patients with BBS can present to many specialties. Clinicians need to recognize the symptoms of BBS and try to diagnose it as early as possible. Epidemiological studies on its prevalence in Pakistani population are recommended.
