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Correlation of Radiologically Abnormal Sella with Small Pituitary Adenomas in an Unselected Autopsy Series

N.B. REWCASTLE, G. WORTZMAN, R. HOLGATE, K. KOVACS, G. BURROU
University of Toronto

Great stress is paid to the demonstration of the asymmetrical sella in the diagnosis of microadenomas of the pituitary gland by computed tomography. This correlation with surgical findings is reported, it is evident that the incidence of sella asymmetry exceeds the incidence of adenoma.

In an attempt to clarify this relationship, the sphenoid bone containing the pituitary gland was removed en bloc in 120 unselected autopsies. Tomography was first performed in frontal and lateral projections. Following fixation and decalcification, parasagittal step sectioning was performed at 1 mm intervals.

Thirty-two glands (26%) contained adenomas ranging in size up to 6 mm maximum diameter. Five glands contained multiple adenomas, maximum three. Only 9 of 23 (39%) were prolactin adenomas as shown by the immunostaining technique. Of 7 cases in which serum prolactin levels were above normal, an adenoma was present in only one.

Tomograms were reported by two observers together, then independently, and were found to be positive and compatible with a microadenoma in 27 cases. Correlation with pathological findings showed there to be 20 false positives and 24 false negatives. In 7 of these, the adenoma and pathology were both negative. The x-ray findings corresponded to the site of the adenoma in 5. In 2 of these the adenoma was too small to be observed in histology.

We conclude that in the asymptomatic microadenoma, sella tomography is neither specific nor sensitive.

Brain Stem Glioma: Ablative Omission of Radiography with Rapidly Progressive Course

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Durham, N.C.

We describe two children, ages 7 and 16, with malignant brain stem gliomas who presented with the abrupt onset of right hemiplegia requiring intubation entreatment within 15 and 23 days from onset. The initial studies included computed tomography, radio-isotope scanning, three- vessel angiography, and surgery failed to reveal the location of the lesion. The brain stem and glioblastoma multiforme was found at biopsy. Neither child responded to radiotherapy nor chemotherapy.

The sudden onset may be the result of hemorrhage, infarction or necrosis within the malignant tumor. The one previous case report of abrupt onset of motor hemiplegia due to malignant brain stem glioma was in a 60 year old woman. This report demonstrates that brain stem gliomas in childhood can also present suddenly with hemiplegia as a result of a malignant tumor with rapid deterioration to complete bulbary paralysis.

Cerebral Cysticercosis in Canada: A Report on Three Cases from Montreal

PAUL A. HWANG, Z. AL-KHAN, E.J. ARPIN, M. AUBE, G. CHONG
Montreal, P.Q.

Parasitological and meningeval cerebral cysticercosis were diagnosed in two Haitian and one Italian male immigrants residing in Montreal. The presenting problem was either a seizure disorder (partial or generalized) or headache or both. Examination of the cerebrospinal fluid revealed hypodense intracranial pressure and on x-ray typical calcified subcuneate nodules.

CT scans showed numerous hypodense cisticular lesions, which enhanced moderately with contrast infusion, in the basal cistern and brain parenchyma. Hydrocephalus in two patients required ventricular shunting. Occlusion of the left middle cerebral artery in one patient resulted in right hemisphere and aphasia. Despite recent fossa decompression and removal of several cysts around the brain stem the patient died of respiratory failure. The clinical management included anticonvulsants, corticosteroids, and ventricular shunting. It is apparent from their case histories, negative stool samples for Taenia solium ova and absence of endemic taeniasis in Canada that in all three cases the cerebral infections went on to duration and import.

In immiggrant cerebral cysticercosis should be included among differential diagnoses in patients with headache and or without signs of raised intracranial pressure. Although a definitive diagnosis of cysticercosis is based on histological features of the larva, a CT scan may be characteristic and suggestive of cystercicosis.

Chemotherapy of Pediatric Brainstem Tumors

DORCAS S. FULTON, VICTOR A LEVIN AND CHARLES B. WILSON
San Francisco, Ca.

Twenty-eight children with brainstem tumors diagnosed by clinical and radiographic examination were treated with chemotherapy. Pathological diagnosis was made in 9 patients. All patients were treated with radiation therapy. Six patients received chemotheraphy immediately following radiation therapy. Two patients received radiation therapy alone following diagnosis and later, at the time of tumor progression, gigned by chemotheraphy. The mean time to progression following radiation therapy in the latter group of patients was 9 months. The median survival of all 28 patients from diagnosis was 14 months-19 months for those receiving cytoxan and vincristine alone or in combination (n = 11). 11 weeks for those receiving BCNU alone or in combination (n = 9), 14.5 weeks for those receiving a nitrosourea with cell cycle specific agents (n = 6), and 11 weeks for those receiving a nitrosourea plus procarbazine (n = 11). Median survival, from onset of chemotheraphy, was 17 weeks for those receiving CCNU alone or in combination, 14 weeks for those receiving BCNU alone or in combination, 30 weeks for those receiving a nitrosourea with cell cycle specific agents, and 125 weeks for those receiving a nitrosourea plus procarbazine. The results indicate no therapeutic benefit for adjuvant chemotherapy following radiation therapy. While we were able to document response of some brainstem tumors to chemotherapy at the time of tumor progression, we found the results of an examination of the current data are not rewarding. Chemotherapy agents are needed for treatment of this rapidly progressive disease.

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The Neurological Manifestations of Rhabdomyosarcoma in Childhood

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Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma and the fifth most prevalent malignancy in childhood. Case reporting stress that tumor may present with a specific neurological syndrome when arising from the head and neck. The incidence of neurological presentation is unknown.

We report 103 cases of RMS diagnosed at the Hospital for Sick Children, Toronto, between 1935-79. Sixteen patients presented with neurological manifestations. Four groups were identified.

Sixty-six percent of patients with tumor arising from the Middle Ear Cleft presented with neurological signs. A VII nerve palsy was the most common finding occurring in 8.10 cases. In association with hearing loss in 3. Three cases presented with muscle weakness and hemiplegia. Sixteen patients presented with cranial nerve palsy and optic atrophy in one.

Two patients with tumor arising from the retropositional area presented with cranial nerve palsy. One patient with RMS arising from the neck presented with Horner's syndrome.

There were 10 patients with orbital tumors and all presented with protrusus rather than any specific neurological manifesta-

It is important to early diagnosis of this highly malignant tumor will be stressed and the rapid neurologic progression illustrated by the case material.

Intracranial Pressure Monitoring

D.L. MORRISON AND F.B. MAROUN
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I.C.P. monitoring has been in clinical use for over 20 years and is now advanced for management of serious head trauma and diffuse encephalopathies such as Reye's Syndrome.

This display outlines the pathophiology of raised I.C.P. the principal drugs, means of measuring I.C.P. The technique, as used at Memorial University of Newfoundland is demonstrated along with some interesting features of I.C.P. monitoring.

Intracranial Pressure Monitoring in Head Injury: A Practical Experience

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Montreal, Que.

In 24 adult patients with severe head injury, the intracranial pressure (ICP) was monitored for one to 5 days with a ventricular canula. Recording of the ICP was technically satisfactory in all but two instances. Intracranial hematomata were removed from 9 patients. Efforts were made to maintain the ICP below 20 mm Hg. by controlled ventilation. Small doses of Magnacrin and in 9 instances, barbiturate coma. The EEG is a practical method of monitoring barbiturate dosage. In selected cases concentrations of barbiturates in CSF and serum were measured and correlated with ICP and EEG. Removal of small quantities of CSF were sometimes necessary and effective. The prognosis was related to the initial level of consciousness and to the ICP. All patients with sustained ICP of greater than 40 mm Hg. developed fixed dilated pupils and died. When the ICP was less than 40 mm Hg., an increase of outcome between ICP and EEG was detected in this series. Eleven patients were ventriculostomized and one asclated. Five of them died and 3 remained in a persistent vegetative state. Two of the remaining patients (Glasgow coma sum 5. 6. 7) died. Elevated ICP in the major cause of death in head injuries. When ICP is monitored, excessive intracranial hypertension can usually be prevented. Because of possible complications of inserting a ventricular canula and inducing barbiturate coma, these techniques are presently reserved for patients with severe head injury. Indications for the use of these methods are still difficult to define precisely.

A Practical Flow Chart for the Management of Patients with Head Injuries

MICHAEL L. SCHWARTZ
Toronto, Ontario

The flow chart is a well-recognized tool that is useful in the preparation of computer programs. Its utility for this purpose re- sides in the fact that the computer proceeds from operation along a path that is selected from a multitude of possible paths by the application of a simple test at each point where the choice of an operation must be made. Since the state of a patient with a head injury is determined by the presence or absence of very few events, since there is only a small number of diagnostic and therapeutical procedures that may be selected and since the choice of one path may be reduced to a few questions that demand a yes or no answer, it has proved possible to construct a relatively simple flow chart that may be used in the management of a patient with cranioencephal trauma. The flow chart has been used in the retrospective analysis of the hospital course of 30 patients with head injury admitted to the Montreal General Hospital and a diagno- sis of multiple trauma. The method has proved useful in identi- fying delays in the application of treatment and deviations from the expected course. Since the chart embodies treatment proce- dures it serves to teach correct decision-making. Finally, even if completely hands one must consider the specific psychological to be described. It is anticipated that it will prove useful in the analysis of a prospective study on which we are embarking.
EEG ACTIVITY DURING BARBITURATE ADMINISTRATION OF HYPOXIC BRAIN INJURIES IN ADULTS

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Vancouver, B.C.

This report describes the results of EEG monitoring during the administration of high doses of barbiturate (Thiopental) in seven patients (ages 17-71 years) who had suffered severe hypoxic brain injury. The patients varied from those who survived, one died a cardiac death on day three, and three appeared to proceed to "brain death. Thiopental was administered as a bolus intravenous blood levels attained ranging from 20 to 150 mg/L in the different patients, assayed by a specific high-pressure liquid chromatographic procedure. EEG recording included a prebarbiturate baseline, and monitoring for the duration of the treatment, and for up to 72 hours after completion, with daily or weekly EEGs carried on for the duration of the patient's hospital stay. In some patients due to technical difficulties data were not complete.

From this experience there are some interesting findings which appear to be important for the interpretation of the EEG in these difficult clinical situations. In the patients who survived for more than a few days, including the two who returned to very nearly normal neurological function, the EEG depression (which usually only attained a burst suppression pattern, rather than an isoelectric trace) was only maintained during the initial phase of the barbiturate administration. After approximately 12-14 hours, despite constant blood levels the EEG gradually became continuous and showed some activity in the Alpha frequency range, or in the patients who did the best in the Beta frequency range. Failure of this return of activity within 24 hours (even in the presence of continued intraventricular barbiturate and maintained blood levels) was uniformly associated with a bad outcome. The EEG patterns associated with the burst suppression blood levels in the individual patients will be considered.

The EEG appears to have an important role for monitoring brain function during barbiturate administration, but it is suggested that in any future studies, the patient should have both the EEG and serum thiopental levels monitored, as these two parameters appear to have different implications for assessment of cerebral function.

ANTICOAGULANT ASSOCIATED INTRACRANIAL HEMORRHAGE

DAVID R. MACDONALD AND CHARLES F. BOLTON
London, Ontario

The contribution of anticoagulation was assessed in a retrospective study of 389 patients who suffered non-traumatic intracranial hemorrhage during a 5 1/2 year period. Twenty-six patients had anticoagulant associated intracranial hemorrhage. Seventeen had intracerebral hemorrhage; 8 had subdural hematoma, and 2 had subarachnoid hemorrhage. 1 had an intracerebral hemorrhage two months after a subdural hematoma. Twenty were receiving warfarin, 3 heparin, and 3 both. Indications for anticoagulation included pulmonary embolism in 11, valvular heart disease in 10, stroke in 2, and other thrombotic disorders in 3. The duration of anticoagulation ranged from 1 day to 13 years. Nineteen patients had hyperension (BP > 160/90 mmHg), 8 had a PT > 1.5 (PTT > 2.5 control), and 6 had previous cerebral infarction. Seventeen had two or more risk factors. The prognosis was poor: intracerebral hemorrhage 19/17 died, 3/17 had major neurological deficit: subdural hematoma - 2/8 died, ½ major deficit: subarachnoid hemorrhage - ½ died.

Anticoagulants appear to be an important cause of intracranial hemorrhage. The risk of hemorrhage is greater when the patient is hypertensive, has had a cerebral infarct, or is receiving excess anticoagulation. It is not clear how the risk of hemorrhage in our series is greater than in previous reports, probably due to improved detection by computerized tomography brain scans.

CEREBRAL EDEMA IN ACETAMINOPHEN OVERDOSE

W.J. BECKER, R. SUTHERLAND, P.J. MULLER
Calgary, Alberta

A 15 year old girl was admitted after taking 30 grams of acetaminophen 4 hours before admission. At admission she was comatose with decorticate posturing and fixed dilated pupils. 72 hours after ingestion Acetaminophen levels were 16.5 mg/dl 48 hours after admission she became delirious. 60 hours after admission she was comatose with decorticate posturing. Head C.T. scan was normal. Total bilirubin was 5.4 mg/dl, SGPT was 2631 (N=33). 72 hours after admission she was hyperventilating, and her right pupil became fixed and dilated. There had been no hypoxia.

A right temporal craniectomy was done. On opening the dura, edematous brain under pressure issued forth. Her pupils again became equal and reactive. 12 hours later both pupils became fixed and dilated, with death 24 hours post surgery.

At autopsy, cerebral edema was present, with gyral flattening. The term "cerebral edema" is used to describe the increased brain and CSF volume due to an increased intracranial pressure. Cerebral edema is characterized by an increase in the brain and CSF volume due to an increase in the interstitial fluid pressure. The term "cerebral edema" is used to describe the increased brain and CSF volume due to an increased intracranial pressure. Cerebral edema is caused by an increase in the interstitial fluid pressure. The term "cerebral edema" is used to describe the increased brain and CSF volume due to an increase in the interstitial fluid pressure. The term "cerebral edema" is used to describe the increased brain and CSF volume due to an increase in the interstitial fluid pressure.
INTRACRANIAL VENOUS THROMBOSIS OCCURRING IN EARLY PREGNANCY

A. GUBERMAN, D.A. GUZMAN AND V. MONTPETIT
Ottawa General Hospital

Intracranial venous thrombosis in pregnancy classically occurs in the postpartum period and only rarely in early pregnancy. We wish to report two patients with extensive intracranial venous thrombosis at 6 weeks and 4 months gestation.

A 32-year-old, unaware of her pregnancy, presented with severe headache and vomiting (without dehydration or electrolyte imbalance) followed by Wernicke's aphasia. Ten days later she developed generalized seizures, a right hemiplegia, left leg weakness and coma. Computerized cranial tomography initially showed a left temporal-pontine hemorrhagic lesion surrounded by edema and later extensive edema with focal hemorrhagic perifocal edema. This was followed by Wernicke's aphasia. Ten days later she was transferred to our hospital. The outcome of cases in early pregnancy is less favourable than those presenting postpartum and treatment remains symptomatic.

"ASSESSMENT OF EXPERIMENTAL MICROVASCULAR SURGERY BY SCANNING ELECTRON MICROSCOPY"

G.E. OUAJKINE, G. MOHR AND J. HARDY
Notre-Dames Hospital and University of Montreal

This poster presents the surgical evaluation by scanning electron microscopy (S.E.M.) of various microvascular operations performed in rabbits and rats (9 longitudinal sutures, 11 venous patches, 40 end-to-end anastomoses and 40 end-to-side anastomoses). Vessels were irrigated with heparine solution and various temporary microclips were used. Different needles and threads were compared, among which 10.0, 11.0 and 12.0 nylon threads mounted on various needles (140, 100, 70 and 50 in diameter). The animals were kept alive for varying lengths of time (from 2 hours to 6 weeks). The patency rate of our experiments was slightly above 90.

The S.E.M. permitted to demonstrate that all varieties of microclips showed alterations of the normal endothelial folds all over the surgical area. The HEFETZ clip showed complete disruption of the endothelial surface, the SCOVILLE clip showed an important flattening of the endothelial folds with prominent cell nuclei, the more recent ACLA and KEES microclips demonstrated the least traumatic modifications. The best suture material to minimize the endothelium lacerations appeared to be 10.0 nylon threads. Continuous suturing technique appeared more traumatic and thrombogenic than the interrupted stitches technique.

MOYA-MOYA DISEASE: A CLINICAL PATHOLOGICAL CORRELATION

D.L. MACGREGOR AND L. BECKER
The Hospital for Sick Children, Toronto

Moya-moya disease (bilateral occlusion of the internal carotid arteries at the level of the carotid siphon, often involving the circle of Willis and main cerebral arteries, and sometimes with a collateral system of fine vessels in the region of the basal ganglia and transcerebrally) was initially observed in Japan in 1956. More than 500 Japanese cases have now been described and recently more than 100 cases have been reported from North America and Europe.

The juvenile type (with onset age 4-6 years) usually presents as repeated attacks of acute hemiplegia and unconsciousness. The clinical and radiological aspects of both the juvenile and adult types are well documented, however, few pathological reports are found in the world literature. We present the clinical, radiological and pathological findings of a case of Moya-moya disease in a 4 1/2 year old Caucasian boy who died 4 days following the onset of an acute left hemiparesis; the diagnosis having been made 2 days prior to death by cerebral angiography. This case represents the first description of detailed pathology in the circle of Willis in Moya-moya disease in a child (showing eccentric production of intimal thickening of the internal elastic lamina and chronic inflammatory cells in the adventitia, particularly in the occluded areas). Immunofluorescent stains (IgG, IgA, IgM, C3 and fibrinogen) were negative.

CLINICAL APPLICATIONS OF INHALATIONAL XENON 133 FOR THE STUDY OF REGIONAL CEREBRAL BLOOD FLOW

B. WEIR AND D. MENON
Edmonton, Alberta

Our experience with aneurysms, arteriovenous malformations and carotid occlusive disease as studied by the above method will be presented with pictorial presentation of illustrative cases and tabular summaries of our overall experience. We think material demonstrates a useful method of gaining information in these neuro-vascular problems.

CEREBRAL BLOOD FLOW AND ARTERIAL VASOSPASM IN PATIENTS WITH SUBARACHNOID HEMORRHAGE

K. MEGURO, G.G. FERGUSON, J.K. FAPPAR, R. CAROUTHERS
London, Ontario

Over a ten month period, 73 CBF studies in 25 patients with subarachnoid hemorrhage have been performed using the 133-Xenon inhalation technique and a 32-probe multi-detector system. A quantitative estimate of the degree of cerebral arterial vasospasm in each case was made from measurements of the diameter of the major cerebral arteries from relevant angiograms in each case.

Mean CBF in patients who were Grades 1, 2 and 3 on the Barrelet scale were 37.6 ± 10 mg/min, 35.7 ± 10 mg/min and 32.6 ± 9 mg/min, respectively. The difference between the Grade 1 and Grade 3 groups is statistically significant (p < 0.01). Good correlation between CBF and clinical grade was seen in 73 studies and poor correlation in 18 of 73 studies. Of these 18 studies, eight showed poor clinical grade with relative preservation of flow. All of these patients had evidence of cerebral infarction. Ten of the 18 studies showed relatively low flows in patients still intact neurologically. Mean CBF in patients without vasospasm was 39.6 ± 6 mg/min, while flow was 31 ± 6 mg/min in those patients with spasm (p < 0.01).

The results demonstrate that CBF is significantly reduced in patients with poor clinical grade and that vasospasm produces a significant reduction in hemispheric flows. We believe that CBF measurements add a new dimension to the assessment of patients with subarachnoid hemorrhage which may be helpful in the management of such cases.

MYOCARDIAL LESIONS IN ACUTE STROKE: A PATHOLOGICAL STUDY

A. KOLIN, J.W. NORRIS, V.C. HACHINSKI
Toronto, Ontario

We have previously documented an increase incidence of cardiac arrhythmias, elevated serum cardiac enzymes and ischemic EEG changes in acute stroke patients as compared to controls. We have also noted a rise in plasma catecholamines in acute stroke patients which may be related to the occasional findings of local myocardial lesions in these patients who die acutely.

We have systematically studied cardiac sections in 70 consecutive autopsies, using histochemical methods sensitive to myocardial damage. No damage was seen in 5 cases who died instantaneously from violent deaths. However, in 13 of 34 hearts from patients with cerebral lesions, myocardial damage was seen throughout the whole thickness of the left ventricle. These cases included intracerebral hemorrhage and infarction. Cardiac arrhythmias, elevated serum cardiac enzymes and ischaemic EEG changes occurred in 26 patients dying of systemic illnesses only 8 showed similar transmural enzymatic changes. We suggest that elevated plasma catecholamines may be related to the areas of focal myocardial damage seen in these patients.

COMPUTED MAPPING OF ELECTROENCEPHALOGRAM (CME) IN CEREBRAL INFARCTION; COMPARATIVE STUDY WITH CT AND REGIONAL CEREBRAL BLOOD FLOW

K. NAGATA, G. ARAKI, M. MIZUKAMI AND T. KAWASE
Mihara Memorial Hospital, Japan

Computed mapping of electroencephalogram (CME) is a newly developed microcomputer system to display equipotential maps of surface root of average power spectrum over each frequency band on color television. This new device was employed in examination of 20 apasies due to cerebral infarction in comparison with CT and regional cerebral blood flow (rCBF) study. High voltage foc in slow wave bands and asymmetry of alpha distribution were regarded as functional lesions on CME. In 12 patients, high voltage foc in slow wave bands were corresponded to both clinical symptoms and CT findings, and in 8 out of 12 patients the lesions on CME were also corresponded to the ischemic lesions in rCBF measurement. Six patients showed asymmetries of alpha distribution which reflected the clinical symptoms. In three patients with motor aphasia, CME demonstrated the lesions in advance of the appearance of low density on CT. Two patients with TIA of aphasic symptoms had hemispheric asymmetry of alpha distribution in spite of no abnormal finding on CT until 6 months after the episodes. Comparing with conventional angiography, the diagnostic sensitivity of functional lesions topographically and objectively though the source of the data is same as conventional EEG.

MONITORING REQUIREMENTS DURING CAROTID ENDARTERECTOMY

MICHAEL BOYD, A.R. WATTS, M.W. BOWERING
Regina, Saskatchewan

Concern for the possibility of shunts causing emboli or ischaemia led to a progression of intra-operative monitoring proce-
dures during arterial clamping without shutting. Initially, only ca-
rroted artery stump pressure less than 50 torr guided shut
ntervention. Subsequently, a two lead EEG oscilloscope display
other abnormalities of possible ischemia. With the addition
of multi-channel EEG recording the prime indication for shutting
became the appearance of EEG ischemic changes. Lastly, ma-
nulation of the systemic arterial pressure (SAP) with drugs, per
cent inspiratory oxygen (FiO₂) and arterial CO₂ pressure (PaCO₂)
by ventilation has been used to control ischemia prior to main-
terior incision. Throughout all cases the occlusion time was
kept to a minimum with the mean being fifteen
utes.

The low mortality and morbidity achieved with these moni-
toring procedures has raised many questions as to what is the
optimal technique to employ. Does routine shutting with its
risk of embolisation or occlusion in fact provide adequate intra-cran-
al blood flow during surgery? On the other hand just how much
monitoring is required to provide sufficient information for relia-
table prediction of intra-cranial ischemia?

Our results suggest that in most situations continuous multi-
channel EEG monitoring provides enough data to allow suffi-
cient intervention with either SAP, FiO₂, PaCO₂ manipulation and or shutting.

PROXIMAL NERVE CONDUCTION IN IDIOPATHIC CHRONIC RELAPSING POLYNEUROPATHY
VERA BRIL, JOHN G. HUMPHREY
Toronto, Ontario

F waves are useful in the assessment of proximal motor nerve function in the Guillain-Barre syndrome. Sixteen patients with idiopathic chronic relapsing polyneuropathy were examined to determine the value of the F wave measurements in this specific group. Eight of the sixteen had varying degrees of motor weakness and sensory axonatia, and the remainder were clinically normal when last seen.

Fifteen patients had abnormal proximal motor nerve conduction, as measured by F waves. In the eight clinically normal or mildly abnormal, the distal motor nerve conduction was found, but marked slowing in proximal conduction was present. In the eight patients who were abnormal clinically, the distal motor nerve conduction was abnormal in six, and the proximal conduction was abnormal in seven. Four of these had greater proximal than distal slowing. One patient out of sixteen had normal conduction in spite of a severe sensory axonatia.

These results confirm the diffusely distributed lesions in idiopathic chronic relapsing polyneuropathy, with a greater proximal involvement. F wave determinations show definite abnormalities in proximal nerve function in the Guillain-Barre syndrome. Sixteen patients with idiopathic chronic relapsing polyneuropathy were examined to determine the value of the F wave measurements in this specific group. Eight of the sixteen had varying degrees of motor weakness and sensory axonatia, and the remainder were clinically normal when last seen.

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suggesting indirectly that Alzheimer’s disease might also have a neurological basis. Although M.S. is the most common CNS disease in young adults, relatively little has been learned about the nature and extent of neurological deficits associated with cerebral demyelination. The neuropathological literature on cerebral demyelination and the clinical epidemiological and neuropsychological literature on neurological deficits is reviewed. It is proposed that neurological dysfunction is associated with overall functional disability but not with disease duration or extent of disease involvement in different systems. Fifty-five sequential referrals to a Neuropsychology Service from a Regional M.S. Clinic who have defined M.S. as determined by a Neurologist were examined on a battery of psychological tests. Significant differences between groups formed by level of disability on a number of cognitive tests confirmed the tendency for overall functional disability. Evidences for why neurological function in new-onset M.S. patients is impaired is given by examination of a subset of patients who submitted to CTT scans. Suggestions are made as to the etiology of this aberration of development.

AN AUTOSOMAL RECESSIVE SYNDROME OF MENTAL RETARDATION, SEIZURES IN INFANCY, AND PROGRESSIVE MULTISYSTEM DEGENERATION

E. ANDERMANN, A. EISEN, T. KIRKHAM, F. ANDERMANN, S. CARPENTER, G. KARPATI AND A. BARBEAU
Montreal Neurological Hospital

In the course of a systematic investigation of different forms of spina bifida disease as part of the Quebec Cooperative Study on Friedreich’s Ataxia, a French-Canadian family was ascertained in which 5 out of 6 siblings, offspring of unrelated parents of normal intelligence, were affected. Two other siblings died of congenital malformations in infancy. Four first cousins of the proband died of congenital malformations in infancy. Four first cousins of the proband died of congenital malformations in infancy. Four first cousins of the proband died of congenital malformations in infancy. Four first cousins of the proband died of congenital malformations in infancy. Four first cousins of the proband died of congenital malformations in infancy.

The five affected siblings, ranging in age from 18 to 27 years, had delayed developmental milestones. Recurrent vomiting and seizures predominantly by fever were present in all in the first year, and remitted spontaneously. By the mid-teens, a combinatorial derivative of pyridoxine, caloric intake, and ketonuria were present. When the spinal cord injury in incomplete, alterations in potential may be post-traumatic.

THE CORTICAL SOMATOSENSORY EVOKED POTENTIAL IN ACUTE SPINAL CORD INJURIES

SONJA ZIGANOW, D.W. ROWED
Toronto, Ontario

The cortical somatosensory evoked potential (SEP) is used predominantly as a useful adjunct in the diagnosis of acute spinal cord injuries. The SEP is absent in patients with complete motor and sensory loss below the level of spinal cord injury. When the spinal cord injuries in potential may be elicited from stimulation of a nerve entering the cord below the level of spinal cord injury. Our results have shown that the presence of such potentials within the first week after injury and progressive normalization of the wave form are sensitive early indicators of favorable prognosis.

In particular, we have found and statistically demonstrated that the better the SEP shortly after injury, the shorter the time interval to walking with a cane and to incomplete motor function that is useful for the determination of the SEP positive peak at about 28 msec discriminates well between patients who ultimately show neurological recovery and those who do not.

having observed the SEP’s in well over 100 spinal cord injured patients we conclude that this technique has prognostic utility because recovery of the SEP can precede major clinical improvement.

CEREBROSPINAL FLUID (CSF) CATALASE IN MULTIPLE SCLEROSIS

W. SHEREMATA, A. SAZANT AND E. PARRIS
University of Miami

Phagocytes are commonly present in central nervous system (CNS) lesions in many diseases; thus implicating free radical production in the pathologic process. Tissue damage by these cells may be produced by the toxic free radical superoxide, a molecular form of oxygen bearing and extra electron. The enzyme superoxide dismutase (SOD) removes superoxide, but its toxic reaction product — H2O2 must be removed by catalase. Low levels of catalase could lead to increased tissue damage or increased levels might lower induction by high SOD activity. As CSF would probably reflect such activity, therefore, we examined fluid from 25 multiple sclerosis and 25 other patients with CNS disease. The timed disc rotation technique developed by Gagnon was utilized with bovine liver catalase (Pharmacia) standards. Fresh 3% H2O2 (Pharmacia) was stabilized with EDTA. All test specimens were examined concomitantly with standard dilutions. Multiple Sclerse CSF gave a mean of 18 ± 4 U/d, while normals gave a mean of 11 ± 2 U/d. Chronic deenerative CNS disease gave a mean of 9 ± 2 U/d. Patients in exacerbation gave values significantly greater than those with chronic progressive or stable disease. Multi ple sclerosis does not appear to be due to abnormally small amounts of catalase.

LISSENCHEPHALY: THREE CASES DIAGNOSED BY CT SCAN

S. HORIZWITZ, B. ROSENBLATT AND G. O’GORMAN
Montreal Children’s Hospital

Lisencephaly is a rare congenital malformation of the brain whereby it never develops convolutional markings, remaining smooth as in a twenty week fetus.

Up until 1978 about thirty cases were reported in the literature, all diagnosed at autopsy with retrospective clinical analysis. Pathologic findings included various degrees of pachygyria or agenesis, with associated cerebral anomalies. Microscopically, heterotopic grey matter was found in the subependymal zone which gave rise to “clefts” or “bands” (Grunert). Experimental studies of abnormal migration of neuroblasts in fetal life. Several studies show convincing evidence for a genetic origin of this defect, whereas others present isolated examples.

We recently observed three patients with the lisencephaly syndrome. However, through the use of the CT scanning we were able to make the diagnosis in life, and thus have the opportunity of monitoring the course as it evolves. We wish to discuss these children in detail with reference to the pathological and clinical features that have been described in the literature, and to show the CT scans. These patients illustrate the spectrum of syndromes ranging from pachygyria to agya giving fuel for speculation as to the etiology of this aberration of development.

HEREDITARY DEMYELINATING INFANTILE NEUROPATHY WITH UNUSUAL CLINICAL AND PATHOLOGICAL FINDINGS

A.F.G. HAHN, J.J. GILBERT, G. HINTON
London, Ontario

A disease expressed as a primary Schwann cell abnormality akin to but different from Dejerine-Sottas neuropathy is present.

S.B. age 12, showed nystagmus, slow motor development and progressive gait and limb ataxia since early infancy. He developed pes cavus and progressive distal lower limb weakness. Reflexes became absent; vibration sense was decreased distally and proprioceptive maneuvers were ob-
served. Peripheral nerves were not enlarge. Progressive partial external ophthalmoplegia was noted and mild sensory neural hearing loss was documented. No evoked motor or sensory ac-
action potentials were recordable with surface electrodes at age 3.

Needle electrode study showed no active denervation, record action potentials were small and dispersed, estimated MNVC 3.5 mV, and SSEP protein 77 mg/mL.

Father, age 42, had slow motor development, pes cavus and slowly progressive distal atrophy and weakness. MNVC was 15 mV/sec in the upper limbs and unrecordable in the lower limbs; sensory potentials were absent.

Sural nerve biopsy showed teased fibers to be totally or par-
dially demyelinated along 80% of their lengths. Oligodendroglia of myelin lamellae were noted in every fiber scattered at random. Onion bulbs were formed by concentrically arranged double layered basement membranes. Axonal loss was minimal.

An abnormality of axon-Schwann cell interaction and a failure of myelination appears to form the basis of this disorder.

SPLANCHIC NERVE SWELLING IN MULTIPLE SCLEROSIS
THOMAS E. FEASBY, GEORGE C. EBERS
DONALD W. PATY, ALLAN J. FOX
London, Ontario

In subacute progressive spinal paraparesis, myelography is commonly used to differentiate inflammatory myelitis from external cord compression and intrinsic cord neoplasms. In 3 women, aged 35, 37 and 49, subacute spinal cord syndromes with paraparesis and a sensory level evolved over 10 to 40 days. Myelography was performed because of the clinical suspicion of spinal cord compression. Widening of the cord, similar to that seen in intramedullary tumor, was seen in the upper thoracic cord in 2 patients and in the mid-cervical cord in 1. However, CSF protein electrophoresis showed oligoclonal IgG banding in each case. This was consistent with the subsequent clinical course, confirming a diagnosis of multiple sclerosis (MS) in all 3 patients. Repeat myelography after 21 days in one case showed a reduction in cord swelling.

Two of our 3 patients with water-soluble contrast medium which may facilitate demonstration of minor cord swelling associated with MS which might otherwise be missed on myelography. These results demonstrate that spinal cord enlargement may be seen in MS. This observation is of special importance in evaluating myelographic findings in suspected spinal cord gliosis. It underlines the importance of alternate diagnostic techniques, particularly the CSF protein electropho-
resis, for a correct diagnosis and avoidance of surgical interven-
tion in demyelinating myopathies.

FAVOURABLE RESULTS WITH THE SYRINGO-SUBARACHNOID SHUNT FOR THE TREATMENT OF SYRINGOMYLIA
KOTO MEGURO, CH.T. TATOR, D.W. ROWED
Toronto, Ontario

There is still considerable controversy about the indications for surgery and the method of surgical treatment of syringomyelia. During the past six years, 19 patients with syringomyelia were treated by a syringo-subarachnoid shunt. The principal indications for this procedure were significant and continuing neurological deterioration during the preceding six months and absent or minimal evidence of tarsal ectopia. There were 14 patients with diastrophic syringomyelia, three with post-traumatic syringomyelia, one with spinal arachnoiditis and one with a spinal arachnoitis cyst. The operations were performed with an operating microscope and attention was inserted into the syrinx through a posterior midline myelotomy in most instances.

The average follow-up was four years. A favourable result was obtained in 14 of 19 patients (73%), including an excellent result with improvement of neurological deficit in 11 patients and a good result in 3. A good result was associated in three patients. There was a poor result with further progression in five patients. A short duration of preoperative symptoms was usually a good prognostic sign. Four patients with a history of less than six months all had excellent results. Eleven patients had only the shunt procedure, and all had good or excellent results. Eight patients had other surgical procedures, before, accompanying, or after the shunt, and three had favourable results.

Thus, syringo-subarachnoid shunt is an effective modality of treatment for syringomyelia particularly if no major tonsillar herniation is present.

AN EVALUATION OF SILASTIC SHEATHING IN NEUROSURGERY OF THE ULNAR NERVE
B.G. BENITO, D.E. PRESTON AND V. DA SILVA
Ottawa Civic Hospital

The variety of procedures designed to relieve entrapment of the ulnar nerve at the elbow, attests to their deficiencies. This study was designed to evaluate the effect of silastic sheath on the ulnar nerve with silastic, neuromyelitis, as a means of minimizing the effects of recurrent scar formation.

A total of 43 procedures were performed on 39 patients, four of which were re-explorations. All cases were graded according to a weighted scoring system utilizing the clinical evaluation of sensory and motor function, but with a major emphasis on electromyographic testing. The patients were then re-assessed using the same parameters, from one to five years after surgery.

Thirty-four (79%) were improved, 6 (14%) remained unchanged and 3 (7%) had deteriorated. Two of the latter were re-explored and new adhesions were found at the ends of the silastic sheath. Most of those who remained unchanged, suffered from an advanced neuromyelitis or had a metabolic predisposition towards entrapment.

It is concluded that silastic, without transposition, combined with silastic sheathing of the nerve, compares favourably with other techniques for primary entrapment.

256 HZ VIBRATION IN THE CARPAL TUNNEL SYNDROME
J. DALTON AND D.N. PRESTON
Ottawa Civic Hospital

To investigate the observation that testing of 256 Hz vibration sense is valuable in assessment of peripheral nerve dysfunc-
tion, we analyzed 120 consecutive patients referred to an electrophysiological laboratory with carpal tunnel syndrome. The clinical signs were evaluated and compared with distal sensory and motor latencies of the median and ulnar nerves. Vibration sense was tested with a 256 Hz tuning fork and considered abnormal if vibration, sensed by the examiner through the patient’s finger, was not sensed by the patient, in patients under age 65. In patients over 65, a selective impairment of vibration in the first three digits of an involved hand was considered abnormal.

Impaired vibration in the first three digits was present in 75% of the whole group. Impairment of vibration was the most common sensory defect noted clinically and exceeded impairments of touch, pain, and 2-point discrimination. No patient had motor signs without sensory findings. Only 16% were normal on clinical examination. The impaired vibration group had a mean distal sensory latency in the involved median nerve of 5.5 - 1.7 milliseconds, compared to 4.6 - 0.9 milliseconds for the others (P < 0.005).

It is concluded that corticospinal and anterolateral concepts, careful 256 Hz vibration testing is useful in the evaluation of a suspected carpal tunnel syndrome.

THE RELATIONSHIP BETWEEN FOCAL PENICILLIN SPIKES AND SPINDLES IN CEREBRAL ISOLE CATS
R.S. MCCLACHLAN, M. KAIBARA AND J.P. GRIVIN
University Hospital, London, Ontario

Gloor and others have suggested that generalized spike dis-
charges induced by Penicillin are closely related to spindles. We studied the relationship between focal cortical spikes induced by topical Penicillin and spontaneous spindles of the Caroie Isobe preparation in cats. A significant association between spikes and spindles occurred in 23 out of 27 foci. Immediately after es-
tablishment of the focus, spikes were independent of or poorly associated with spindles, but the relationship between the two wave forms gradually increased with time. In 11 foci, after a lat-
ter period of 1 to 40 minutes, spikes occurred only during a spindle, i.e. 100% association. In the other cases, spikes occurred after establishment of the epileptic focus.

The morphologic relationship of the focal spike discharge to the spindle was consistent. A new spindling will be presented. These findings will be discussed with respect to the thalamocortical interaction in the production of local Peni-
cillin spikes.
PERIPHERAL NEUROPATHIES IN CHILDHOOD

H.G. DUNN
University of British Columbia

In children, electrophysiological studies are required more often to aid in the diagnosis of polymyopathies than in the investigation of isolated cases. This is particularly so in the extremity function tests. The extremity function test showed 126 children with polymyopathies as compared to only 35 with mononeuropathies due to trauma or entrapment (excluding facial palsies). The former may be grouped as follows:

- Genetic
  - hypotrophic type of Charcot-Marie-Tooth disease
  - Friedreich's ataxia
  - Krabbe's leucodystrophy
  - metachromatic leucodystrophy
  - adrenoleucodystrophy
  - hereditary sensory neuropathies
- Others, e.g. muscular dystrophies
- Metabolic
  - hypothyroidism
- Inflammatory
  - Guillain-Barré syndrome
- Cryogenic
  - chronic sensori-motor neuropathy
- Nutritional
  - coeliac disease
- Others, e.g. dysthyroid disease
- Sural nerve biopsy.

The clinical seizure forms in 30 children with a temporal lobe epilepsy were analyzed. Nineteen had a typical clinical pattern of focal motor seizures. Seven had a more complex pattern of seizures, with either secondary generalization or a more diffuse onset. In these seven children, the seizures were often difficult to classify due to the presence of other clinical features. The results suggest that temporal lobe epilepsy is a heterogeneous disorder, with a wide range of clinical presentations. The differences in seizure patterns may reflect underlying differences in the underlying pathology of the temporal lobe.

DERMATOLYMPHIC ALTERATIONS ARE A SUBSTITUTE INDICATOR OF ANTICONVULSANT EFFECT ON THE FETUS

E. ANDERMAN, A. SHERWIN, F. ANDERMAN, AND L. DANSKY
Montreal Neurological Hospital

Gestational exposure to anticonvulsant medication has been associated with a higher risk of major congenital malformations and mental retardation. This study investigated the pattern of dermal ridge formation in children of mothers treated with anticonvulsants during pregnancy. The results suggest that dermal ridge patterning, which is laid down between the 13th and 18th week of gestation, may serve as a sensitive indicator of drug-induced dysmorphogenesis early in development.

Surgical treatment of intractable seizures is continuously developing. Some newer drug treatments include stereotactic surgery, which involves the precise and safe placement of electrodes in the brain to monitor and control seizures. The use of functional magnetic resonance imaging (fMRI) and positron emission tomography (PET) scans is also being explored to localize the seizure focus.

CLINICAL SEIZURE FORMS IN TEMPORAL LOBE EPILEPSY

K. FARRELL
University of British Columbia

The clinical seizure forms of temporal lobe epilepsy are commonly associated with a focal neurological deficit, such as a sensory or motor deficit. These seizures are often difficult to classify due to the presence of other clinical features. The results suggest that temporal lobe epilepsy is a heterogeneous disorder, with a wide range of clinical presentations. The differences in seizure patterns may reflect underlying differences in the underlying pathology of the temporal lobe. The results of this study may provide clinicians with new insights into the classification and management of temporal lobe epilepsy.
THE TREATMENT OF PROGRESSIVE MULTIFOCAL LEUCOENCEPHALOPATHY WITH ANTI-DNA VIRUS AGENTS: A CASE REPORT

D.J. MACFADYEN, B. ROZDILSKY, G.A. STILWELL
Saskatoon, Sask.

The diagnosis of PML, a rare but well known complication in patients suffering from a lymphoma, leukemia or having received immunosuppressive therapy, has been facilitated by the C.T. scan. To date no consistently effective treatment for PML, a papovavirus infection of the central nervous system, is available. Another DNA virus infection, namely herpes simplex encephalitis, will respond to adenine arabinoside (a-Ara-A) therapy and acyclovir, ganciclovir shows promise. Cytosine arabinoside (a-C) and a-Ara-A have been reported as producing variable results, including prolonged remission, when used in the treatment of PML.

A 37 year old man with a 5½ year history of Hodgkin's disease (Stage III B) and for which he had received previous x-ray radiation and chemotherapy presented with a three month history of progressive slurring of speech and incoordination of the right hand. The physical findings were those of a pyramidal type weakness of the right arm plus an oropharyngeal apraxia. A presumptive diagnosis of PML was supported by a distinctive C.T. scan and further substantiated by means of E.M. and histology of biopsy samples.

On each of nine days he received intravenously 30 mg/kg/m of a-Ara-A. There were no significant side effects. Because of a continuing deterioration of his neurological status and beginning 16 days after the completion of his a-Ara-A therapy he received intravenously on each of six days 105 mg/m of Ara-C. SBS originated from the frontal lobe in 29 patients (50%) and from the temporal lobe in 16 (28%). 35 patients (53%) had continuing focal epileptiform abnormality immediately following the GEPS. In each case, this was the focus of onset of seizures.

The SBS appeared as irregular 2-3 hertz spike and wave complexes in 36 patients (64%), regular 3 hertz spike and wave complexes in 14 patients (25%) and sharp and slow wave complexes in 9 (16%).

Since Tuckel and Jasper's description of SBS (1952), there have been few studies of its electrophysiological and clinical correlations.

CLINICAL SIGNIFICANCE

Although a delay in appearance of the major electropositive potential is common, the pattern potential characteristics of clearly defined (P2) s have been termed an aberrant wave form (AWF) occurred in 20% of patients in whom the (P2) peak for at least one eye was clearly definable. The AWFs for at least one eye had definite MS, an additional 33%.

Electrographic and Clinical Correlates of Secondary Bilateral Synchrony (SBS)

N. PILLAY, W.T. BLUME
London, Ontario

This report describes the results in 118 patients in whom the diagnosis of M.S. was being considered or had already been established. At the time of testing 38 were classified as clinically definite, 21 as probable M.S., 30 as possible M.S., and 12 as progressive Multiple Sclerosis (MacDonald-Haliday 1977 criteria) and 33 did not have sufficient signs or symptoms to fit into any of these groups. All patients were tested with pattern-reversal visual evoked potentials (PVEP), somatosensory evoked potentials from right and left median nerve stimulation (SEP), and brainstem auditory evoked potentials (BAEP).

In the total number of patients in one of the diagnostic categories the PVEP was abnormal in the highest percentage (66%). The SEP was abnormal in 52%, the AEP in 43%, the SEP in 35% and one or more responses abnormal in 78% of the total of these patients. In the 32 patients outside the diagnostic classifications there was an ASPA level in both environments. The clinical situation in which they occurred will be considered.

The addition of the AEPs and the SEPs to the PVEP does increase the yield of diagnositically useful information from senso-motor evoked potentials. In this study the PVEP was the single most sensitive indicator of subclinical involvement, with the SEP slightly less effective and the AEP the least sensitive indicator of disseminated demyelination.
in 105 consecutive cases. An internal shunt was used only three times. Of the 105 patients there had a new deficit post-operatively. In every instance the deficit was transient.

In 64 cases continuing intra-operative EEG recordings are available. An internal shunt was not used in any of these patients. One-third (23/64) showed significant EEG changes during cross-clamping. In two patients a minor post-operative deficit occurred; in neither of whom there was an EEG change. In 18 patients measurements of cerebral blood flow (CBF) and intracranial pressures were made. In six patients with a significant EEG change, CBF fell an average of 39% during clamping; while in 12 patients without a significant EEG change, the fall was 17%. In general, changes in stump pressures paralleled the changes in CBF.

Our results demonstrate that major EEG changes, and profound reductions in CBF as low as 24 ml 100 gm in one case) may occur during carotid endarterectomy without any new post-operative deficit occurring. These observations support our view that an internal shunt is rarely, if ever, necessary during carotid endarterectomy.

RETNAL NERVE FIBRE ATROPHY IN COMPRESSION OF THE CHIASM: A PROGNOSTIC SIGN

Owen B. White and James A. Sharpe

Toric, Ontario

Recovery of vision after chiasmal decompression cannot be predicted reliably from the nature of the lesion or the duration or pattern of visual loss. Atrophy of the retinal nerve fibre layer (NFL) can be detected by direct ophthalmoscopy using red-free illumination when the optic disc appears normal. We describe the correlation of visual fields loss and characteristic fundusoculographic signs of diffuse or retinal NFL atrophy before and after treatment of compressive lesions of the optic chiasm. Red-free fundus photography and funduscopy were performed in ten patients who presented with visual field defects detected by kinetic Goldman perimetry and tangent screen examination. Serial red-free fundus photography was performed in the NFL were correlated over follow-up periods of one to five years (mean 2.2 years) after surgery.

Degeneration of the nasal temprory retinalNFL was present in seven of twenty eyes. Seven eyes showed diffuse atrophy of optic axons on the retina. Six eyes showed NFL atrophy. Field loss persisted in eyes with hemiplegic or diffuse atrophy, but incomplete recovery of acuity and fields occurred in three of fourteen such eyes. All eyes without visible NFL atrophy demonstrated total or near total resolution of visual defects. Severe atrophy of the NFL correlates with persistent visual loss. Apparent normality of the NFL portends visual recovery after decompression of the optic chiasm.

CEREBRAL NON-VISUAL CONTROL OF THE VESTIBULO-OCULAR REFLEX

James A. Sharpe and Alex W. Lo

Toronto, Ontario

Orientation of gaze, the sum of head position in space and eye position in the orbit, can be controlled by modulation of the vestibulo-ocular reflex (VOR) during passive head movement. This requires summation of head position and retinal target information. In both animals and humans, visual information is derived from on-center off-surround cells in the visual cortex and is transmitted through the lateral geniculate nucleus to the abducens nuclei in the medulla. The abducens nuclei send axons to the oculomotor nuclear complex which in turn send axons to the medial rectus muscles. The muscle fibers in the medial rectus muscles are innervated by the oculomotor nerves. The oculomotor nerves are composed of two types of nerve fibers: type I fibers, which are slow-conducting fibers, and type II fibers, which are fast-conducting fibers. Type I fibers are responsible for the maintenance of eye position in the orbit, whereas type II fibers are responsible for the generation of eye movement.

Clinical symptoms of vestibular-ocular reflex dysfunction can be seen in patients with cerebellar or brainstem lesions. The most common clinical manifestation is called the nystagmus syndrome. Nystagmus is characterized by a sudden, rapid, involuntary movement of the eyes, usually back and forth or from side to side. The disorder is often accompanied by vertigo, headache, and dizziness.

The vestibulo-ocular reflex (VOR) is a reflexive response to changes in head position. The reflexive responses are mediated by the vestibular system, which is located in the inner ear, and the oculomotor nuclei, which are located in the brainstem. The VOR is important for maintaining visual stability during head movement and for helping to prevent falls.

THE INFLUENCE OF THE HISTOCHEMICAL PROFILE OF MUSCLE ON THE IN VITRO CAFFEINE CONTRACTION TEST

A.K.W. Brownwell, M. Szabo

Calgary, Alta

In vitro contracture tests on skeletal muscle are recognized as the most sensitive method for detection of individual at risk for malignant hyperthermia (MH). Several factors are known to influence the outcome of these tests. These include the temperature at which the muscle preparation is carried out and the duration of drug injection by the subject prior to the biopsy. This report describes the effect of the histochomical profile of the biopsy specimen on the results of in vitro caffeine contraction studies in patients at risk for MH.

THE DESIGN AND EVALUATION OF A PROBLEM BASED LEARNING APPROACH IN UNDERGRADUATE NEUROLOGY

H. Barrows

Hamilton, Ontario

Problem based learning requires the student to learn while attempting to evaluate and understand neurological patient problems. The student is taught to apply the hypothesis-generating, deductive logic of the clinician to determine the anatomical, physiological, biochemical, psychological, pathological processes responsible. Guided by faculty in this process the student discovers the concepts in basic neuroscience and clinical neurology that must be learned by self study to understand neurological problems and the nervous system in general. Following self-study from a wide variety of resources, the knowledge and skills acquired are applied back to each problem undertaken. Advantages of this approach include: 1) acquisition of problem solving (clinical reasoning) skills, 2) integration of knowledge from the many disciplines in neuroscience, 3) recall of learned information in working with clinical problems, and 4) reinforcement of learning through active participation in the problem-solving process. 7) Learning is individualized to the educational needs of each student, 8) acquisition of self-educational skills in neurology for lifelong learning.

CLINICAL VALUE OF THE CORNEOMANDIBULAR REFLEX

A. Guberman

Ottawa General Hospital

Although the corneomandibular reflex has been recognized since 1902, its significance remains unknown. The reflex is known to be elicited by compressing the corneomandibular nerve behind the ear or by squeezing the masseter muscles. The reflex is thought to be mediated by the trigeminal nerve and is thought to be a protective reflex that prevents damage to the eye when the head is struck. However, the clinical significance of the reflex is unknown.

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THE RATIONALE AND PROGRESS OF THE COLLABORATIVE ECIC BYPASS STUDY

H.J.M. Barnett

University Hospital, London, Ontario

RATIONALE

Many surgeons are capable of performing superficial temporal to middle cerebral artery microvascular anastomosis. Morbidity can be kept to a minimum when the anastomoses are performed without occlusion of the carotid artery. As a result of our experience, we have developed a protocol for performing carotid endarterectomy in the presence of severe stenosis of the internal carotid artery. We have found that the use of a carotid clamp can be avoided in most cases, and that the use of an internal shunt is rarely, if ever, necessary during carotid endarterectomy.

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A NEW INHERITED CANINE HYPMYELINATING DISORDER ASSOCIATED WITH CONGENITAL TREMOR

J.D. DUNCAN, I.R. GRIFFITHS, G.M. BRAY
Montreal, P.Q.

An inherited canine disorder, characterised by the development of a white body tremor, has been found in 6 male pups from three litters produced by a Springer spaniel bitch. The tremor develops around 12 days post-partum and involves the limbs, body, head and eyes. Three affected pups were perfused at 2.6 and 11.6 weeks and the central and peripheral nervous system samples stained. Grossly, there was an obvious deficiency of myelin throughout the entire CNS; the nerve roots appeared normal. Light microscopy revealed striking hypomyelination of all tracts of the spinal cord, brain and optic nerves, with an apparent paucity of glial cells. Longitudinal sections demonstrated that intra­nodal myelin lamellae and without the normal membrane specializa­tions or maturation will be discussed.

CEREBRAL PHOSPHOLIPIDOSIS EXPERIMENTALLY-INDUCED WITH CHLORPHAMERONE

J.R. WHERRETT
S. HUTERER, M. KHAN AND N.B. NEWCASTLE
University of Toronto

An increasing number of neurotoxic drugs have been found to cause a systemic lipidosis in animals morphologically similar to the Niemann-Pick syndromes. These experimental phospho­lipodonts could provide conversion models in which to analyze pathological changes in cerebral storage disorders. Here we describe morphological and lipid changes in brain and other tissues of rats and baboons after intranasal administration of all trans retinaldehyde. The histopathological changes were also investigated. Plasma levels of 14 amino acids were given either Fluosol-DA 35% (15 ml/Kg) or mannitol 20% (2 g/Kg). Animals in the experimental groups were divided into three groups of 12 animals each. All animals were sacrificed at the end of the second week post-operation. The brains were perfused with formalin. Light microscopic analysis of ischemic cortical neuronal change after middle cerebral artery occlusion. The brains were perfused with formalin. Light microscopic analysis of ischemic cortical neuronal change after middle cerebral artery occlusion. The brains were perfused with formalin. Light microscopic analysis of ischemic cortical neuronal change after middle cerebral artery occlusion. The brains were perfused with formalin. Light microscopic analysis of ischemic cortical neuronal change after middle cerebral artery occlusion. The brains were perfused with formalin. Light microscopic analysis of ischemic cortical neuronal change after middle cerebral artery occlusion. The brains were perfused with formalin. Light microscopic analysis of ischemic cortical neuronal change after middle cerebral artery occlusion. The brains were perfused with formalin. Light microscopic analysis of ischemic cortical neuronal change after middle cerebral artery occlusion.

A PERSONAL PERSPECTIVE

D. FAIRHOLM
Taipei, Taiwan

The challenge and responsibility of Medical Education is one of which we are all a product and in which many of us are active participants. Medical Education in the developing world is even a much greater challenge. Many simple or complicated solutions to the problems of medical education in this developing world have been tried. But have resulted in an incredible "Brain Drain" towards the developed world with adverse effects on the sending country. North American manpower saturation and government awareness of this has resulted in immigration policies which will make it difficult or prohibitive for perspective candidates in medical education. Consequently, we need to take a fresh look at involvement in International Education and development.

THE CANADIAN JOURNAL OF NEUROLOGICAL SCIENCES

The theory of Gardner relates to the development of syringo­myelia cavities by abnormal CSF pulsation pressure transmis­sion between the fourth ventricle and the spinal canal of the spi­nal cord. This explanation is not adequate for all categories of syringomyelia, especially the non-communicating varieties (e.g. spina bifida and post-traumatic). Alternative theories include abnormal CSF passage through the cord substance via Venous-Robins spaces or a result of a mechanical compromise of CSF by glial cells lining the cavities. While most neuro­radiological assessment has been anatomic, i.e. evaluation of cord elongation and confirmation of the presence of cysts, the advent of water soluble contrast media and CT allows a more dynamic study of CSF pathways in syringomyelia. Five cases of syringomyelia, four of which were post-trauma­tic, are presented showing metrizamide taken up in the cord syrinx, five or six hours following metrizamide myelography. Three of these, on myelography, showed small cords, that would otherwise be called atrophic. This appears to confirm the theory of subarachnoid fluid passage through the spinal cord between a syringe and the surrounding subarachnoid space. This also may be the most definitive way of confirming the presence of the syrinx.

WE BELIEVE THAT TENSION PNEUMOCEREBRALIS SHOULD BE CONSIDERED IN THE DIAGNOSIS OF SYRINGOMYELIA.
22 control probands the mean plasma levels of ASP, THR, SER, ILE, LEU and TYR were significantly decreased along with a significant decrease of TAU/SER, GLN/GLU, THR/GLY and SER/GLY. When the relatives of epileptic probands were compared with the control probands, they showed a significant decrease in TAU, ASP, GLN, GLU, TYR, TAU/GLU and GLN/GLU, and a significant increase in VL. This group of epileptic patients was also found to be significantly different from those non-epileptic probands. The examiner observing a significant decrease of THR, SER, GLY, AME, ILE, LEU, TYR, THR/GLY and SER/GLY. Discrepancy Analysis it was possible to distinguish epileptics from controls with 100% accuracy, and from 3/sec spike-wave epilepsy with 90% accuracy.

The results of this indicate: 1) the epileptic probands whose electroencephalograms show focal, abnormal or diffuse type of patterns, show plasma amino acid patterns which are significantly different from those of probands with 3/sec spike-wave epilepsy. 2) that the plasma amino acid patterns appear to be genetically controlled at least in part and 3) that the plasma amino acid patterns of these epileptic probands are also significantly different from those of probands with 3/sec spike-wave epilepsy.

PLASMA AND ERYTHROCYTE FLOW IN ACUTE FOCAL CEREBRAL ISCHEMIA
J. R. LITTLE, A. COOK, S. A. COOK AND W. J. MACINTYRE
Cleveland Clinic, Cleveland, Ohio

The object of the investigation was to study plasma and erythrocyte flow and morphological changes in an area of acute focal cerebral ischemia. The right middle cerebral (MCA) of 18 ketamine-anesthetized cat was exposed microsurgically. Plasma and erythrocyte flow in the right Sylvian artery was determined by measuring the transit of $^{11}{}$ albumin and $^{99m}{}$ Tc-labeled erythrocytes injected into the ipsilateral carotid artery.

In the past twenty years the anterior approach to the cervical spinal canal pioneered by Cloward has gained wide acceptance in the surgical treatment of spondylosis resulting in medullary or radicular compression. Surgical experience gained in using the Cloward technique has led to the development of a series of modifications suited to the individual problem. Unilateral or contralateral cervical spine following trauma. Thirty cases of post-traumatic cervical spine instability are reviewed in which stabilization was accomplished using an anterior approach. Modifications of the classic Cloward interbody fusion utilizes include vertical pegs, trapezoid autogenous bone strut and vertebral body replacements using acrylic implants. Follow-up clinical and radiological assessments demonstrate that, when the anterior vertebral elements are the pivot of instability, patients possessing with complete quadriplegia are best treated with acrylic implants. For those with incomplete neurological insults and patients without neurological deficit, stabilization using autogenous bone struts or pegs in one or several stages produces the most acceptable outcome.

EXTRALUMINAL DISSECTIONS WITH CAROTID ENDOARTECTOMIES
K.S. POLYZOIDIS AND J.D. McQUEEN
University Hospital, Saskatoon

The media was dissected from the plaque in 40 cases prior to vessel clamping with the aid of the operation microscope and microinstrumentation. A small (Bmrm.) longitudinal incision was made over the lower external carotid arteries. The media and the elastic lamina were separated from the calcified plaque with a small, curved instrument. Circumferential dissection was only complete. When the plaque was then removed. Three vessels were clamped; the incision was passed into the lumen and extended. The plaque was then removed in one or two pieces in the routine way.

The major advantages are: (1) the shortening of dissection times (and particularly in the setting where pressor agents are used in cardiac patients) (2) facilitation of removal of intact plaques. Intraoperative monitoring and post-operative clinical evaluations gave no evidence of embolization.

BLOOD FLOW CHARACTERISTICS OF THE EXTRA-CEREBRAL CIRCULATION
M.I. VILAGHY, V.C. HACHINSKI, J.W. NORRIS, R.D. RUDDELL AND P.W. COOPER
Sunnybrook Medical Centre, Toronto

The extra-cerebral circulation is an important factor in cerebral blood flow measurements by non-invasive techniques and in certain diseases like migraine. It has been generally assumed that it is a small, non-quantifiable and inert compartment. In order to re-examine this question, we have studied three groups of patients using the xenon 133 intra-arterial injection technique and the initial slope index method.

In the first group (11 patients) regional cerebral blood flow (rCBF) values were compared by ipsilateral internal carotid artery and common carotid artery injections. In a second group (14 patients) selective extra-cerebral artery (ECA) injections were performed and the rCBF values were compared in rest and during carotid compression. In the third group (13 patients) CFB pattern changes were studied following extracranial intracranial (EC-EC) anastomosis surgery by injecting the tracer via the shunt.

Our results suggest that the extra-cerebral circulation 1) is not homogeneous 2) responds paradoxically (as compared with brain to arterial PCO$_2$ changes) and 3) EC-IC anastomosis surgery results in marked and unpredictable changes in both the regional pattern and paradoxical PCO$_2$ rection of the extra-cerebral circulation.

INTERNAI FIXATION OF CERVICAL SPINE DISLOCATIONS WITH AN INTERLAMINAR CLAMP
R.O. HOLMES, W.S. HUESSTIS, R.A. LANGILLE AND W.J. HOWES
Victoria General Hospital, Halifax

For many years we have utilized a locally made clamp device, originally designed by Dr. H.H. Tucker, for fixation of cervical fracture dislocations or unilateral subluxations. This method has evolved to be our procedure of choice in injuries where posterior cervical instability is the main defect. Thirty-two patients were treated between 1972 and 1978 and all were initially placed in cervical traction and most reduced prior to operative treatment. Fifteen had fractures through the posterior elements of the cervical spine, 12 neuro root deficit, 4 mild spinal contours and 3 severe cervical cord injuries.

The clamp is applied to the adjoining laminae of the involved level and on the opposite side. The fusion is then performed. In 2 patients bilateral clamps were applied because of severe instability. Two were re-explored in the immediate postoperative period for replacement of stripped clamps. Postoperatively a cervical collar and in some cases, a Minerva cast was used. The great majority left hospital within 3 weeks.
PERCUTANEOUS LUMBAR RHIZOTOMY FOR SPASMS IN PARALYSIS

I.M. TURNBULL
Vancouver, B.C.

Many operations have been recommended to help paraplegics with uncontrollable spasms of the legs. Routes L1 to L5 can be reached percutaneously without entering the subarachnoid space. Thermal lesions readily abolish spasms in the distribution of these roots without affecting the sacral visceral reflexes. Over the past two years, sixteen patients have been so treated.

Patients treated fall into three categories. The first are young paraplegics who find their independence impaired by spasms. The second group are paraplegics of longer standing with decubitus ulcers which will not heal because spasms cause insurmountable nursing problems. The third group have multiple sclerosis and characteristically, have more sensory function in the legs than do the traumatic paraplegics.

Initial response to operation has been good. Early problems with alteration ofbalance and diminished sensation have not proved difficult. Two of the first patients treated developed recurrent spasms which did not respond completely to repeated operations. Stimulation ofroots prior to lesion making at the second operations evoked twitching in muscles not innervated by those roots, a finding never seen at the initial procedure. Indications for the operation will be discussed in the light of our experience.

EARLY vs. DELAYED REPAIR OF RUPTURED INTRACRANIAL ANEURYSM

H. HUGENHOLTZ, F.J. PAYNE and R.G. ELGIE
Scarborough General Hospital, Toronto

100 consecutive stable (Gr. I-III, Botterell) patients with ruptured berry aneurysms were analysed to compare the results of early and delayed operative repair. Patients following admission occurred in 28 patients, resulting in 8 deaths, deterioration preceding further surgical consideration (4), and persistent clinical deterioration prior to eventual surgery (17). Therefore, only 88 patients came to surgery, with only 64 having maintained their admission grade or better. In this group, 31 were operated on within 7 days of their most recent hemorrhage and 33 thereafter.

The outcome following operation is shown -

<table>
<thead>
<tr>
<th>Last hemorrhage &amp; Good</th>
<th>Fair &amp; Poor</th>
<th>Deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 - 2 days</td>
<td>11</td>
<td>2</td>
</tr>
<tr>
<td>3 - 7 days</td>
<td>13</td>
<td>3</td>
</tr>
<tr>
<td>&gt; 7 days</td>
<td>25</td>
<td>5</td>
</tr>
</tbody>
</table>

The incidence of significant post-operative vasospasm following surgery within 48 hrs. of a bleed, 2/15 (13%), resembled that when surgery was delayed at least 7 days; 7/47 (15%), but was lower than its occurrence in 2/26 (31%) of cases repaired 2-7 days following a bleed.

Early repair of ruptured intracranial aneurysms within 48 hrs. of a bleed in clinically stable Gr. I-III patients, appears to be a reasonable alternative to a policy of delayed intervention, and merits further experience.

HERPES SIMPLEX VIRUS ENCEPHALITIS ISOLATES ANALYZED BY RESTRICTION ENDONUCLEASES.

K.G. WARREN, M.L. LEWIS and S.M. BROWN
University of Alberta

J. SUBAK-SHARPE
Glasgow, Scotland

B. YOUNG AND A. ZBITNEAU
Saskatoon

D.W. PATY and G.D. KETTLYS
Boston, Massachusetts

The DNA of Herpes Simplex Virus may be purified and digested into fragments with restriction endonucleases. The DNA fragments, which radioactively labelled with P32, can then be subjected to agarose gel electrophoresis and digested into fragments with restriction endonucleases. The DNA restriction enzyme profiles obtained readily differentiate Herpes Simplex Virus type I from type II.

We previously reported that 31 spontaneous isolates of herpes simplex virus type I from infantile encephalitis, superior cerebral, and vagus ganglia from 17 individuals could be classified as 15 different virus strains by analysis of DNA restriction profiles. Virus isolates from different individuals could be differentiated from one another. Furthermore multiple virus isolates from the same individual are indistinguishable.

We report here the restriction endonuclease profiles of eight strains of herpes simplex virus which cause encephalitis of humans. The isolates were obtained from London, Ontario, Saskatoon, Saskatchewan, Edmonton, Alberta, and Vancouver, British Columbia. DNA virus was digested with the restriction endonuclease Bam H1 and KpnI. All eight isolates exhibited restriction enzyme profiles characteristic of Herpes Simplex Virus type I, and they could all be differentiated from one another. It is concluded that endonucleases may be caused by many strains of Herpes Simplex Virus type I, and as yet no subgroup of strains is associated with that disease.

PROSPECTIVE STUDY OF EFFICACY AND TOXICITY OF CARBAMAZEPINE AND PHENYTOIN AS PRIMARY ANTI-EPITHELIC DRUGS

J. BRUNI, B.J. WILDER and A. BAUMAN
Wellesley Hospital, Toronto and University of Florida

A prospective double blind study evaluating carbamazepine and phenytoin was performed using newly diagnosed epileptics with generalized tonic-clonic and partial seizures. Twenty-five patients randomly received carbamazepine or phenytoin and have now completed 6 months. The patients receiving phenytoin did not differ significantly from the carbamazepine-treated patients with regards to seizure type and frequency, age, sex, and duration of the seizure disorder. Doses were adjusted according to response, toxicity, and plasma levels. Of 11 carbamazepine-treated patients, 2 were discontinued because of non-compliance and two patients were discontinued because of an allergic skin rash. Seven of 9 patients who completed the study all had excellent seizure control (5 were seizure-free). Plasma carbamazepine concentrations ranged from 3 to 10μg/ml. Of 12 phenytoin-treated patients, 5 patients had to be terminated because of lack of efficacy, 2 patients were discontinued because of an allergic skin rash and only 5 achieved excellent seizure control in the absence of toxicity. Plasma phenytoin concentrations ranged from 5 to 17μg/ml. Side effects were similar in severity and frequency in both groups of patients, however, only patients in the phenytoin-treated group showed a lack of response (5 patients). Carbamazepine was a more useful drug in the treatment of these patients.

EEG ABNORMALITIES AND CONVULSIONS IN JUVENILE DIABETES MELLITUS - A FOLLOW-UP STUDY

D.L. KEENE, K. METRAKOS, M. BELMONT, G.V. WATERS AND S. SINGER
Montreal Children’s Hospital

Every diabetic child is exposed to the risk of hypoglycemic convulsions. Now with a tighter diabetic control being advocated for coma, intracranial infection (n = 94) and convulsions

A. ROY
Clarke Institute of Psychiatry, Toronto

22 in-patients with a discharge diagnosis of hypoglycemic convulsions were matched for age and sex with 22 in-patients with a discharge diagnosis of epilepsy. The hypothesis tested was that psychiatric variables would differentiate the two groups. The di-agnoses were made independently by Psychiatrists with a special interest and knowledge of epilepsy after usually a lengthy admission which invariably involved observation of the attacks with post-mortem physical examination and EEG’s. A sleep and sleep EEG’s and in some sphenoidal EEG’s, psychometric tests and specialized radiology. Some had an historical attack during an EEG recording.

Both groups completed the General Health Questionnaire, the Waterfield self-assessment depression inventory, the Mood Anxiety Inventory, the Eysenck personality and the Foulds Hostility questionnaires. The Hamilton Rating Scale for depression was completed during a psychiatric interview.

There were no statistically significant differences on psychologi- cal variables between the two groups. The practical implication is that a female patient presenting with seizures, who is currently depressed, has a past and family history of psychiatric disorder, has attempted suicide and is sexually maladjusted should be admitted for further investigation before a diagnosis is made.

SEIZURES PRECIPITATED BY MENTAL ARITHMETIC

B. ZIFKIN, F. ANDERMANN, E. MCGOVERN
Montreal Neurological Hospital

AND A. WILKINS
Cambridge, England

A patient with generalized convulsions noted that seizures were reliably precipitated by mental arithmetic. Immerical EEG revealed only a mild diffuse nonspecific disturbance but performance of mental arithmetical activity evoked burst of generalized epileptiform activity with no obvious clinical accompaniment. Extensive psychological testing was performed to confirm this finding statistically and to further specify the nature of the cognitive activity responsible for epileptiform discharge. Tasks involving multiplication, division, and manipulation of spatial information were significantly associated with paroxysmal epileptiform EEG. Staging subtraction, and the simple retention of spatial information were not associated with such discharge. The likelihood of paroxysmal discharge was not related to the number of cognitive steps involved in solving a problem but problems of division yielding a quotient and remainder were more likely to evoke discharges than one yielding a quotient alone. Simple attention to a task was not sufficient to yield epileptiform activity. The majority of epileptogenic tasks in this patient, including mental arithmetic, involved processing of spatial information and are similar to those whose performance is impaired by parietal lobe lesions. Generalized epilepsy induced by thinking may relate to parietal lobe dysfunction in a manner analogous to the involvement of the occipital lobe in pattern-sensitive epilepsy.

COMA IN CHILDHOOD WITH PARTICULAR REFERENCE TO IT’S AETIOLOGY, CLINICAL FINDINGS, MORTALITY, MORBIDITY AND PROGNOSIS

A.O. OGUANNEKAN
Lagos, Nigeria

We reviewed the records of patients seen from 1976 to 1978 for coma, intracranial infection (n = 94) and convulsions (n = 94) in children under 15 years of age. Of the 225 children (age ranged 6 weeks to 10 years) seen in the Children’s Emergency Room of the Lagos University Teaching Hospital, excluding neonatal and traumatic causes of coma,
seizures occurred in 152 children (68%) and were more frequent in those aged between 1 to 3 years than in the other age groups. Sixty children died. Ocular and motor findings correlated significantly with mortality. Metabolic acids and a variety of other systems disturbances, usually multiple, occurred in nearly half the children. At discharge, 102 of the remaining 165 children were normal and the remainder had some degree of handicap (mild to severe). The importance of detecting and correcting accompanying systemic disturbances is discussed.

THE RESULTS OF SURGICAL TREATMENT OF TEMPORAL LOBE EPILEPSY. A PERSONAL EXPERIENCE WITH THE FIRST 100 CONSECUTIVE CASES

A. OLIVIER
Montreal Neurological Institute

From 1971 to 1979 inclusive, the author has carried out 124 temporal lobeectomies including the amygdala and the pes of the hippocampus in 124 patients, of whom 100 had a minimum follow-up of one year. For the purpose of analyzing the results, the patients have been classified in three main groups:

GROUP A (seizures-free or maximum alleviation of seizures)
1) totally seizure-free since discharge
2) patients who have become seizure-free (minimum one year)
3) patients still having a maximum of 3 seizures per year

GROUP B (patients improved or only slightly improved)

GROUP C (patients having less than 50% of the number of seizures)

The results do not seem to differ when operations are performed on the left (57%) or on the right (43%). Ten patients (10%) in this series have had pre-operative stereotaxic depth electrode studies. There were no death and no permanent hemiparesis or dysphasia in this series.

The results will be discussed on the background of the total number of temporal lobe cases treated surgically at the Montreal Neurological Institute with emphasis on the preoperative investigation.

NEUROLOGICAL INVOLVEMENT IN CHILDREN WITH MORPHEA (LOCALIZED SCLERODERMA)

P. HWANG, G. WATTERS, K. METRAKOS, B. MOROZ, F. ANDERMANN AND B. ROSENBLATT
Montreal Neurological Hospital and Montreal Children's Hospital

In a retrospective study of 25 children with morphea (mean age at diagnosis was 4 years), neurological abnormalities were found in 65% including psychomotor retardation, partial or generalized seizures, hemiparesis, and dysphasia. Thirty percent had seizures. Electroencephalographic (EEG) abnormalities were found in 75% of the patients, including paroxysmal or focal abnormalities while a few had epileptic discharges spontaneously or with photic stimulation. The type and localization of the EEG abnormality did not seem to correlate well with the site of the lesions.

Associated abnormalities included hemichromic atrophy, ecoli- nephalia, increased sedimentation rate and hyperglobulina- mia. Radiological studies done and in progress including CT-Scans showed cranial and cerebral asymmetry appropriate for the side of the face involved only in some cases. The incidence of neurologic abnormalities in these patients is not only directly related to the cerebral lesions but is also caused by the systemic disease.

EXTRA PYRAMIDAL COMPLICATIONS OF METOCLOPRAMIDE THERAPY

J.D. GRIMES, M.N. HASSAN AND D.N. PRESTON
Ottawa Civic Hospital

Metoclopramide is now a widely prescribed drug. Despite being a central dopaminergic receptor (DA 2) antagonist the drug was considered safe to use in Parkinson's Disease. Apart from acute dystonic reactions (1%), extrapyramidal complications have been rare. Over a 15 month period 11 patients with metoclopra- mide-induced extrapyramidal disorders were assessed in a Parkinson's Disease Clinic. Eight patients (average age 54) had developed acute tran- sient dystonic reactions. Eight patients (average age 71) deve­loped parkinsonism (average 1.7 years metoclopramide treat­ment). Six of these were referred as classical Parkinson's Disease and 4 were already receiving L-Dopa (average 1 yr). Metoclopramide withdrawal resulted in complete clearing of par­kinsonism within 2 weeks in 5 patients. The other 3 patients rem­ained with some residual symptoms and may have had pre-existing undiagnosed Parkinson's Disease.

Six patients (5 of the parkinsonism group and 1 other) developed tardive dyskinesia on continuing metoclopramide (average 2.6 yrs treatment). This was transient in 3 but occasionally multiplemovements persisted at 8 months in the others.

Chronic metoclopramide therapy can induce transient or per­manent extrapyramidal disorders in older patients. Its chro­nic use seems contra-indicated in Parkinson's Disease and the drug should be placed in the same extra-pyramidal risk category as the neuroleptics.

CEREBRAL TUMOURS IN CHILDHOOD PRESENTING AS CHRONIC FOCAL EPILEPSY

J.P. GIRVIN, W.T. BLUME
London, Ontario

That a longstanding uncontrolled focal seizure disorder may represent a supratentorial tumour neoplasm in childhood is uncommonly appreciated. We found tumours in 14 of 29 patients less than 20 years old operated on for intractable epilepsy from 1974 to 1979. This contrasted markedly with the 5% tumour incidence of the MNI series covering the same age group (Mathiesen, 1975).

In our series, the majority of seizure disorder prior to surgery was 7.3 years. 11 of these 14 patients (79%) had normal neurological examinations; 4 of 15 patients (27%) without tumours had normal EEGs.

Sixty children died. Ocular and motor findings correlated significantly with mortality. Metabolic acidosis and a variety of other abnormalities were more frequent in those aged between 1 to 3 years than in the other age groups, including insulinomas and in 6% of these children have had both pre-operative stereotaxic depth electrode studies. There were no death and no permanent hemiparesis or dysphasia in this series.

The results do not seem to differ when operations are performed on the left (57%) or on the right (43%). Ten patients (10%) in this series have had pre-operative stereotaxic depth electrode studies. There were no death and no permanent hemiparesis or dysphasia in this series.

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MANAGEMENT OF VASCULAR AND TUMOURAL LESIONS OF THE PINEAL REGION IN CHILDREN

E.G. VENTUREYRA AND V.F. DASILVA
Children's Hospital of Eastern Ontario, Ottawa

Since August 1977, 10 patients have been operated upon at the Children’s Hospital for lesions in or around the pineal region. (Ages 6 months to 17 years, 6 males, 4 females.) Depending upon the location and type of lesion, a different surgical approach was used. Six patients (1 each of pineal germinoma, pineal mixed germ cell tumour, pineoblastoma, teratoma and aneu­rysm of the Vein of Galen) were approached using the occipital transtentorial route. A venous malformation of the Vein of Galen was exposed through the infratentorial supracerebellar approach, a thalamo-epidural cut through the transtentorial approach and a transventricular approach was used for an exten­sive gmrinoma of the pineal region. A giant aneurysm of the parietal approach. Intraoperative Doppler recordings were used during the clippings. Five lesions were totally excised, 4 were subtotally and 1 thrombosed. Four patients harboring malignant tumours received postoperative radiation and 2 in addition received chemotherapy. Magnification and illumination of the deep surgical field was obtained with a fiberoptic head light, and magnifying loupes and the operating microscope. The postoperative morbidity was negligible and no permanent sequelae were observed but one patient developed seizures following clamping and excision of the giant AVM. The present series document the feasibility of the direct surgical attack on vascular and tumoural lesions of the pineal region with little mortality or morbidity. Pineal surgery can be carried out safely and successfully providing the patient would benefit from CSF shunting. An abnormal test consists of a rise in CSF pressure of more than 1.5, 2.0 and 2.5 mm Hg/min during the infusion of artificial CSF into the lumbar subarachnoid space at rates of 1.5, 2.0 and 2.5 cc/min for 10 minutes respectively. Another constant feature of an abnormal test is the persistence of days to weeks. No patients had clinical or historical evi­dence of pore-tosystemic encephalopathy, although 3 patients had mild cardiac liver abnormalities. We propose that acornol­ism unmasked or augmented pre-existing Parkinisonism. Three cases had prior episodes of transient Parkinisonism relieved by abstinence. Animal studies have shown significant impairment of basal ganglia dopamine metabolism during ethanol intoxica­tion and ethanol withdrawal. A movie of two patients will be shown.

PARKINSONISM PROVOKED BY ALCOHOLISM

M.A. LEE, P.L. CARLEN, M. JACOB AND O. LIVSHITS
Toronto Western Hospital

Six cases of chronic alcoholics are described wherein Parkinonsm was provoked by alcohol withdrawal (4 cases) or by chronic intoxication (3 cases). One patient demonstrated a lingual-oral dyskinesia as well. All patients showed partial or complete recovery of their Parkininosms with maintained abstinence of at least 6 months. No patients had had a typical or historical ev­i­dence of pore-tosystemic encephalopathy, although 3 patients had mild cardiac liver abnormalities. We propose that acornol­ism unmasked or augmented pre-existing Parkinisonism. Three cases had prior episodes of transient Parkinisonism relieved by abstinence. Animal studies have shown significant impairment of basal ganglia dopamine metabolism during ethanol intoxica­tion and ethanol withdrawal. A movie of two patients will be shown.

CSF DYNAMICS IN ADULTS WITH HYDROCEPHALUS

H. SCHUTZ AND F. TAYLOR
Toronto

This paper presents the results of 85 lumbar CSF infusion studies (Katzman Test) in 79 patients. The purpose of this test is to aid in the identification of those patients with hydrocephalus who would benefit from CSF shunting. An abnormal test consists of a rise in CSF pressure of more than 1.5, 2.0 and 2.5 mm Hg/min during the infusion of artificial CSF into the lumbar subarachnoid space at rates of 1.5, 2.0 and 2.5 cc/min for 10 minutes respectively. Another constant feature of an abnormal test is the lack of return to baseline pressure within 15 minutes between infusions. A third constant factor is the ability of the test to be reproducible. In the past, the results of CSF shunting in hydrocephalic pa­tients, selected solely on the basis of the clinical presentation, air study and CT scanning, have produced disappointing results. 37 of the 79 patients were selected for shunting on the basis of an abnormal infusion test. 31 of the shunted patients returned to an independent life style. The fa­vorable result was maintained for a mean follow-up period of 2.5 years. Follow-up was complete for 77 of 79 patients.

The correlation between an abnormal infusion test and a di­agnostic CT scan was only 59%. This suggests that an assess­ment of the dynamics of CSF circulation should be included in order to refine the indications for CSF shunting.

GUIDELINES FOR MANAGEMENT OF INTRA-UTERINE HYDROCEPHALUS

D. COCHRANE, T. MYLES
Calgary, Alberta

The aggressive treatment of intratventricular hydrocephalus has been of benefit to many children. Somatic and 2-dimensional ultrasound techniques (grey scale, color Doppler and spectral analysis) of hydroce­phalus more reliable than previous methods.

LEUKOCYTE GLUTAMATE DEHYDROGENASE IN VARIOUS FORMS OF ATAXIA

A. BARBEAU, M. CHARBONNEAU AND T. CLOUTIER
Montreal Neurological Institute

Many studies from our group have indicated that pyruvate metabolism may be impaired in certain sporadic cerebellar depen­dencies (CJNS: 389-397, 1976). In other studies we have demonstrated a marked decrease in cerebellar glutamic acid and occasional decreases in GABA and/or aspartic acid in Friedreich’s Ataxia (FA) (CJNS 6: 311-319, 1979). To further in­vestigate the origin of this glutamic acid deficit, we measured the activity of leukocyte glutamate dehydrogenase (GHD) in a vari­ety of cerebrovascular degenerations. In this series, we reported elsewhere (A.N.A., April 1980 meeting) indicated that leukocyte GHD was significantly decreased in 12 FA pa­tients as compared to controls (p < 0.001). We now report extended re­sults of leukocyte GHD in 44 control subjects and 44 ataxic sub­jects, all determined as sex- and age-matched pairs. The ataxic subjects included 23 classical Friedreich’s ataxia (FA), 8 dominant olivo-ponto-cere­bellar degenerations (OPCA Type I), and 5 recessive ataxias of Charlevoix-Saguenay type (CS), 8 dominant olivo-ponto-cere­bellar degenerations (OPCA Type II), and 5 recessive ataxias of spinocerebellar degeneration (OPCA Type III). The group with a significant leukocyte GHD decrease was the FA group; CS, OPCA and RA groups, which were not signifi­cantly different from controls, included a few patients with very low values, generally far advanced or long-standing cases. This important finding deserves further studies as to its cause and relation­ship to the decreased serum (lipoamide dehydrogenase) values found in the same patients. Similarly, we are also investiga­ing methods for replacement therapy of glutamic acid. This is supported by us at the Association Canadienne de l’Ataxie de Friedreich.

SUPPRESSOR CELL ACTIVITY AND MITOGEN RESPONSES IN VARIOUS PHASES AND STAGES OF MULTIPLE SCLEROSIS (MS)

D.W. PATY, H.K. COUSIN AND B. BASS
University Hospital, London, Ontario

Several investigators have reported changes in non-specific suppressor activity (NSC) during various phases of MS. We have looked at mitogen responses and NSC activity in 40 MS patients that meet Schumacher criteria. Mitogen responses (PHA, ConA, PWM) were measured by a micromethod in triplicate. There was a consistent increase in responses during the acute relapse (+43.5%, P < 0.01; N = 7) using patients as their own controls. The following Correlation of NSC activity was found in 40 MS patients and 17 normals.

Phase of MS | Mean P | SD | P Value
--- | --- | --- | ---
Stable MS (N = 22) | 65.8 | 13.72 | 0.05
Acute Relapse MS (N = 9) | 52.4 | 16.79 | 0.02
Chronic Prog. MS (N = 6) | 70.9 | 10.86 | 1.00
Benign MS (N = 3) | 32.7 | 19.70 | 0.02
Normal (N = 17) | 62.1 | 14.76 | 0.02

Stable MS patients were not significantly different from normal; NSC activity was reduced during relapses, elevated in progressive and very low in benign MS. These data show recipro­cal changes in mitogen responses and NSC activity in the relapsing remitting NSC and progressive forms of MS. This study increases our understanding of the role of the suppressor cell in the MS disease process. These data are consistent with the concept of a disorder of immune regulation in MS. A relevant antigen specific model is needed.

AN UNUSUAL FORM OF CENTRONUCLEAR MYOPIA WITH COPIOUS LIPOFUSCIN ACCUMULATION IN THE CENTER OF MUSCLE FIBERS

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A 60 year old man had a progressive painless loss of muscle bulk and strength in the limb girdles and proximal limb regions for about 10 years. The lower extremities were more affected. The disability was relatively mild. Serum CK was elevated less than twofold. Electromyography revealed some single fiber ac­tivity and some abnormal small and large motor units. In a quadriceps biopsy, the majority of muscle fibers had single or multiple clustered central nuclei; some of these were bizarre in shape. The long axis of many peripheral nuclei pointed towards the center of the fibers. The central nuclei often surrounded a myofibrillar core (up to 50 microns in diameter) which was often filled with copious amounts of myogen and/or typical lipofuscin. Both fiber types were equally affected by the central nuclei; this suggested a genetic etiology (probably autosomal dom­inant). Two of the patient’s 4 sons had a very mild shoulder girdle atrophy and weakness, myopathic electromyographic pat­terns, and type I fiber atrophy on biopsy, but only rare ncu­clei; this suggested a genetic etiology (probably autosomal dom­inant) for the disease. The pathological picture in the father’s

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biopsy differs from that of other forms of centronuclear myopa­
thy, because it shows large clusters of central nuclei, and be­
cAUSE OF THE COPIOUS CENTRAL ACCUMULATION OF GLYCOPEN AND LIPIDOC.
the pathophysiology of these changes is presently unknown.

FACIO SCAPULO HUMERAL DYSTROPHY: THE INFANTILE VARIETY (FSH)
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FSH muscular dystrophy is generally considered a restricted disorder with onset in the second decade; running a relatively benign course. An infantile FSH disease has been described by Duchenne, a century ago, forgotten and rediscovered in rare contemporary reports. It is not a widely recognised entity. We describe a 13 year old male patient showing facial diplegia in the neonatal period with a progressive muscle weakness leading to loss of ambulation at about 7 years. Outward signs are the peculiar shoulder slouch and hamstring tightness. He is intellectually normal. Se­
rum enzymes are markedly elevated and EMG studies show myopathic motor units. The muscle biopsy taken at 5 and 13 years of age revealed prominent replacement of muscle fibre by delicate fibrous tissue infiltration. The patient's mother showed mild facial, neck and shoulder weakness in line with a low penetrance of the FSH phenotype. Biopsy of masseter muscle was consistent type I fibre atrophy. This is more typical of myotonic dystrophy (MD) with which this family has little in common clinically. However like in congenital myo­
tonic dystrophy most reported cases of FSH had an early disabling myopathy most likely inherited through an autosomal dominant gene from a very mildly affected parent. A major differ­
ence seems to us the absence of the intermediate disba­
tility between the two ends of the FSH spectrum unlike (MD).
A movie will show the patient at the near end of his ambulatory pe­
riod and literature will be reviewed on the subject.

END-PLATE ACTIVITY IN MAN
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Two types of electrical activity are evident at the End-Plate zone with concentric needle electrodes. These include (1) low voltage negative potentials which correspond to miniature End-Plate potentials and (2) other larger voltage negative potentials which frequently have pre-photons evident in the rising phase of the initial negative voltage deflection. The origin of the latter has been uncertain but most authors have claimed that the latter discharges represent nerve fibre activity. Theoretically, this explanation is not particularly attractive.

For this reason the whole diaphragm preparation was investigated by means of intra-cellular electrode techniques, and parallel needle electrode investigations were carried out with the conventional concentric needle electrodes used in human electromyography. The spontaneous larger voltage negative potentials were rarely detected with the intra-cellular electrode, even though the miniature End-Plate Potentials were readily detected by both electrode techniques. The negative potentials were clearly triggered by contact of the panconic needle electrode and its fibres. They were abolished by curare. The latter evidence led to the conclusion that these positive negative discharges at the End-Plate zone represent, not nerve fibre activity, but muscles fibre action potentials, probably pre-synaptically activated by mechanical irritation of the motor axon terminal and pre-terminal branches. The latter has been uncertain but most authors have claimed that the latter discharges represent nerve fibre activity.

PREDICTION OF RESPONSE OF HYPERACTIVE CHILDREN TO METHYLPHENIDATE
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Follow up studies of hyperactive children have demonstrated persistent academic, social and/or behavioral difficulties in spite of longterm treatment with stimulant medication. Hyperactivity in children seems to be the final common pathway for a number of different conditions and it is possible that a favourable academic and social outcome of certain subgroups of hyperactive children treated with methylphenidate is masked if the subgroups are not differentiated.

This study reports a longitudinal investigation of 96 boys aged 8-11, the aim of which was to develop a rationale for treatment of hyperactivity. Each child received detailed neuropsychologic, neurological, psychiatric and electrophysiological studies with the aim of delineating specific subgroups. Different treatments including behavioural, diet manipulation and stimulant drugs were then systematically tried with each subgroup and the most effective treatment for each subgroup was determined.

A neuropsychological test battery was predictive of response to stimulant drug treatment. In contrast the presence of learning disability, personality adjustment problems and/or family pathology were not predictive of a favourable drug response. The neuropsychological profile of the typical "favorable responder" will be presented.

PREDICTION OF OUTCOME IN NONTRAUMATIC COMA IN CHILDHOOD
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The data in 102 comatose children (seen Feb. 76-Dec. 78) were analysed. The median age was 30 months (range 1-17 yrs) and the median duration of coma, 30 hrs. (range 3 hrs-35 days). 33 children died, 51 were normal and 18 had mild, moderate or severe handicap (follow-up 6 mths-3 yrs). The rela­
tionships between clinical variables (examination, 21/2 hrs. of the nearest adult) and outcome were:
1. Aetiology: 35% with meningitis and 62% with encephalitis were normal; 72% with anoxic-ischaemic insults died. 2. Seizures: 63% with seizures at onset of coma and 6% with seizures after the onset were normal; 70% with generalized sei­ures and 16% with multifocal seizures were normal. 3. Coma Severity (Grade 1 to 4); 70% in grades 1 or 2 coma were normal. All those in grade 4 coma died. 4. Motor: 58% with normal motor patterns were normal. All those faccicled and ataxic were normal. 5. Ocular findings: 67% with normal EOM, 61% with reactive pupils and 70% with corneal reflexes were normal. All with ab­sent EOM or non-reactive pupils and 64% with absent corneal reflexes died. 6. Respiratory: 62% with normal respiration and 31% who re­quired artificial respiration died. 7. Blood pressure: 58% able to maintain BP were normal, 93% unable to maintain BP died.

8. Body temperature: 58% able to maintain body temp. were normal; all those unable to maintain temp. tested by the patients to their primary symptoms of excessive daytime drowsi­ness, sleep attacks, cataplexy, vivid hypnaglog hallucinations, and also to other frequent symptoms such as visual problems (blurring, diplopia) and memory impairment.

SPECTRAL ANALYSIS OF THE BASIC ACTIVITY FOR CLINICAL APPLICATION OF THE EEG APPLIED IN PATIENTS WITH SPACE OCCUPYING LESIONS
O. MAGNUS, D.C.J. POORTVLIET
C.J.M. VAN DER WULP AND A.C. VAN HUFFELEN
The Hague, Netherlands

This method has been applied to 49 EEGs recorded in 37 pa­
tients with intra-cranial space occupying lesions (24 tumours, 9 intracerebral hematomas, 3 extracerebral hematomas and 1 large aneurysm). It will not only improve prognosis (i.e. of death but (ii) will also provide baseline data to evaluate therapeutic measures.

SOCIO-ECONOMIC AND RELATED LIFE-EFFETS IN 180 PATIENTS WITH NARCOLEPSY-CATAPLEXY
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Y. HISHIKAWA AND Y. SUGITA, Osaka
S. NEVSIMALOVA AND B. ROTH, Prague

A questionnaire survey of some 180 life effects items has been made in 180 narcoleptics, 60 from each of North America, Asian and European populations with 180 age and sex matched controls selected at random. Most of the patients were tested by the patients to their primary symptoms of excessive daytime drowsi­ness, sleep attacks, cataplexy, vivid hypnagog hallucinations, and also to other frequent symptoms such as visual problems (blurring, diplopia) and memory impairment.

Occupational problems were extremely prevalent (over 75%) and included statistically significant deleterious effects upon: performance; promotion; earning capacity; fear of, or actual, job loss; and more disability insurance. Driving was greatly affected and patients more frequently fell asleep at the wheel (69%), had near or actual accidents from drowsiness or falling asleep at the wheel (67%), and could experience cataplexy (29%) or sleep paralysis (12%) at the wheel. Accidents attributed to sleepiness or falling asleep at the wheel (49%) or related to smoking (49%) were much more frequent in patients. There were also deleterious effects on education, recreation and per­
sonality related to the symptoms. Narcolepsy can therefore pro­
duce a variety of serious side-effects probably more serious and pervasive than, for instance, those of epilepsy. This emphasizes the importance of adequate and easy diagnosis and treatment.

CAPABLE NOSMARKS: CLINICAL AND PATHOLOGICAL ASPECTS
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In the past 12 months, five cases of primary brain lymphomas with clinical manifestations resembling multiple sclerosis. Par­
kinson's disease and encephalitis respectively (three cases), and narcolepsy (two, including one with seizures) were seen and followed.
Lymphoma was diagnosed by brain biopsy in four cases and autopsy in one. Extracerebral primaries were excluded by clinical investigations or autopsy. CT scan was positive in four but negative in one patient with splenial involvement; multifocal tumor was present in one case. CSF showed pleocytosis, increased total protein and gamma globulins, and positive cytology in two cases. Pathological examination in all cases included light microscopy and immunocytochemistry for gamma globulins and lysozyme. The tumors were diagnosed as poorly differentiated or anaplastic lymphohematopoietic neoplastic (one) and lymphoplasmocytic (two). Light microscopy and immunocytochemistry were diagnostic in three cases but both PD and tumor markers were negative in the fourth case for unequivocal diagnosis. None of the tumors was clearly monoclonal.

Both cases received cranial irradiation and chemotherapy and were followed for up to 12 months. Because of the potential that therapeutic response of brain lymphoma, importance of early biopsy with the aid of electromyography and immunochemistry is stressed in cases with obscure diffuse or multifocal CNS disease which may represent lymphomatoform disorder.

CLINICAL APPLICATIONS OF SUBCORTICAL SOMATOSENSORY EVOKED RESPONSES

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Five patients are presented with myasthenia confirmed by clinical, electrical and serological criteria. All were taken to early thymectomy and to other anti-myasthenic therapy being en­
ployed. These patients experienced persistent ocular myasthe­nic features (diplopia and ptosis) following thymectomy, despite resolution of proximal muscle and bulbar weakness. Wrist ocu­lar symptoms failed to resolve during the ensuing 12 months, plasmapheresis was performed. These patients were consecutively studied Tension positive and on no other medic­ations. All five patients failed to show any improvement in the ocular features of myasthenia gravis, immediately after plasma­pheresis or during the ensuing two months. Acetylcholine receptor antibody titers were elevated in each patient initially and came down with plasmapheresis without corresponding clinical benefit. All patients were then treated with Prednisone and showed dramatic improvement.

Clinical improvement in myasthenia gravis with plasmaphere­sis has been postulated to be the result of the removal of a circulating humoral factor. The acetylcholine receptor antibody has been implicated. The failure of these post-thymectomy symptoms to respond to plasmapheresis despite normalisation of the antibody titre suggests that another mechanism may be operational. The striking response to steroids in preference to another anti-myasthenic therapy being en­ployed is explained by the continuing production of new antibody. One of the advantages of plasmapheresis is that it can be used in serial fashion and is not limited to a single use. This study demonstrates the value of repeated plasmapheresis in patients with refractory myasthenia gravis.
hours nor received any sedative, hypnotic, or analgesic for 12 hours prior to surgery. After informed consent, they randomly received (n = 11) or did not receive (n = 12) electrode stimulation immediately prior to surgery. MAC for surgical incision was produced by periaqueductal gray stimulation. The duration of analgesia was less by the Waud test than 0.50%, the MAC for patients in the unstimulated group was 966 at 15 minutes after stimulation, 1011 at 30 minutes, and less by the Waud test than 0.50%, the MAC for patients in the unstimulated group was 966 at 15 minutes after stimulation, 1011 at 30 minutes, and less by the Waud test than 0.50%, the MAC for patients in the unstimulated group was 966 at 15 minutes after stimulation, 1011 at 30 minutes, and less by the Waud test than 0.50%, the MAC for patients in the unstimulated group was 966 at 15 minutes after stimulation, 1011 at 30 minutes, and less by the Waud test than 0.50%, the MAC for patients in the unstimulated group.

Both nerve injection and crush injury resulted in a significant breakdown in the BNB with marked endoneurial edema which was felt to contribute to the nerve fiber damage seen with these injuries.

Following nerve section and suture, there was an immediate breakdown of the BNB at the site of anastomosis which, over the following 3 weeks, spread throughout the proximal segment undergoing Wallerian degeneration. Regeneration was followed closely by restoration of the BNB along the course of the regenerating nerve.

Our results also indicate that a peripheral nerve at the site of a traumatic end bulb neuroma lacks a normal BNB. The disturbed endoneurial environment may contribute to the painful symptoms often seen with these lesions.

MANAGEMENT OF SEVERE HEAD INJURIES WITH BARBITURATE PROTOCOL

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The University of Vermont protocol for the management of head injuries is presented. In this protocol is the mandatory administration of high dose intravenous barbiturates in all severe head injuries (Grade 3, 4, 5) beginning immediately after neurological assessment in the Emergency room. These agents are used to attempt to maintain the intracranial pressure within normal limits and hopefully influence quality of neurologic recovery. Our observations, based on a detailed analysis of 25 cases treated to date, do not support the contention that barbiturates favorably influence the intracranial hypertension or the quality of the patient's outcome.

ROYAL COLLEGE LECTURE: "MALIGNANT" BRAIN EDEMA IN CHILDREN

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Sixty-one of 215 children who had early CT scans following head trauma exhibited a pattern which we have described as diffuse cerebral swelling. This is marked narrowing of the ventricular system, compression of the perimesencephalic cistern and a general loss of CSF spaces. Three quarters of these children had Glasgow Coma scores of 8 or less, and 24 had Glasgow Coma scores of 5 or less.

The patients were divided into 2 groups: Those with a lucid period and the onset of secondary deterioration and those who were immediately rendered unconscious. In the former group, the acute deterioration is suspected to be due to acute brain swelling produced not from the central component of the brain but by acute congestion of the brain, vasodilatation and hyperaemia. Studies of cerebral blood flow, cerebral blood volume and a new indicator of cerebral blood flow have been performed in the second group of children with immediate onset of unconsciousness, 60% demonstrated evidence of extracranial collections, progressive ventricular dilatation, and a pattern which looked like atrophy. Their time to recovery was much longer than the group with a lucid interval and their residual neurologic deficits much worse. It is concluded that acute diffuse brain swelling due to increased blood volume and hyperaemia is a common accompaniment of acceleration-deceleration head injury in children. In those with a lucid interval, if the hyperaemia and acute brain swelling are controlled, rapid recovery will occur with a return to normal. In those patients with an acute onset of unconsciousness and evidence of diffuse brain injury, recovery will be slow, delayed intracranial hypertension can be expected and the majority of these children will take many months to recover. We believe that the term "malignant" cerebral edema is a misnomer.

THE ROLE OF THE BLOOD NERVE BARRIER (BNB) IN NORMAL AND INJURED PERIPHERAL NERVE

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Nerve fibres in a peripheral nerve function in a unique environment created and maintained by special barrier mechanisms (blood nerve barrier) analogous to the blood brain barrier in the central nervous system.

The present experimental study was designed to examine the role of the blood nerve barrier in both the normal rat sciatic nerve and in nerves under various pathological conditions. The BNB was assayed by a fluorescent tracing of intrafascicularly injected bovine serum albumin labeled with Evans' blue and by electronmicroscopic tracing of 1.4 J. injected horseradish peroxidase. The right sciatic nerve of 2 animals of each group of 25 animals were subjected to either 1) nerve injection with a variety of anesthetic, steroid and local anaesthetic agents, 2) nerve transaction followed by epineural suture, 3) nerve transaction with neuroma formation, or 4) crush injury.

The left sciatic nerve was used as the control throughout. The nerves were examined at varying periods from 1 to 12 weeks following injury. Results revealed that in the normal nerve the intraneuronal fluorescence is destroyed in the lumen of the endoneurial microvessels and efferent with no passage into the endoneurium. The anatomic sites of the BNB appeared to be the tight junctions of the endoneurial capillary endothelium and perineurial cells.

There is controversy concerning the role of internal neurolysis in nerve surgery. Many workers consider the procedure valuable in treatment of a variety of nerve lesions associated with fibrosis. It is also a fundamental component of newer techniques of nerve repair which employ fascicular suture and interfascicular nerve grafting. Others view internal neurolysis as a potentially hazardous procedure which adds another potential site of injury to the nerve and to damage to the microcirculation, occurring in intrafascicular and perineural swelling. Pain frequency and work laboratory on the subject proposed the present study to examine morphological and physiological alterations in nerve fibres following experimental neurolysis.

Internal neurolysis was performed on the right sciatic nerve of 108 Wistar rats using the operating microscope and microroughmic techniques. Groups of 8 animals were studied from 1 to 12 weeks postoperatively. Nerve conduction studies were carried out on both the operated upon and control nerves in all animals. Alterations in the blood nerve barrier (BNB) were assessed by both fluorescent and horseradish peroxidase (BNB) blue albumin and by electronmicroscopic tracing of horseradish peroxidase.

Careful performed internal neurolysis does not result in any significant nerve fibre damage as assessed by both light and electronmicroscopic. Consecutive tissue stains revealed only minimal degree of epineural and perineural fibrosis and no evidence of interfascicular swelling. Blood nerve barrier studies showed transient increased permeability of penicillinum and endoneurial microvessels to tracer proteins at 1, 24 and 48 hours, no longer evident at 1 week. Electrophysiological studies showed transient early 20-40% dip in nerve conductance velocity which appeared to correlate with the disturbance in the BNB. A more persistent breakdown in the BNB, seen in 7 animals, was in each instance associated with inadvertent operative trauma to perineurum.

It is concluded that experimental internal neurolysis per se does not result in significant lasting morphological and physiological alterations in normal nerve. Nevertheless, care must be taken to avoid damage to the perineurum which can result in more persistent damage to the BNB and decreased electrical function of the nerve. The significance of these results in the light of our clinical experience will also be discussed.

RADIOTHERAPY AND CHEMOTHERAPY IN THE TREATMENT OF GLIOMAS: AN EXPERIMENTAL STUDY

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The avian sarcoma virus-induced glioma model in rats was used to evaluate radiation dose response (survival curves), combining fractions treatments to the whole head. Groups of 30 rats received total doses of 1000-4000 rads in 40 fractions over 2, 4, or 5 weeks, respectively. Median group survival times were compared to controls and to each other. All doses of radiation significantly prolonged survival with reference to control. The 4000 and 4575 rads doses were significantly more effective than 2300 rads. In other series of experiments, BCNU chemotherapy (10 mg/kg) was combined with 2400 and 4600 rads radiation therapy. Synergies of therapies was demonstrated. Mytphilic monoclonal antibody to gliomas (10 mg/kg) was given on survival and its combination with 4600 rads radiation therapy.

CEREBROVASCULAR PERMEABILITY IN MECHANICALLY INDUCED HYPERTENSION

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Our previous studies of cerebrovascular permeability in angiotensin-induced acute hypertension have demonstrated that the acute intravenous administration of angiotensin is enhanced protein synthesis. In order to exclude the possibility that the enhanced protein synthesis was due to angiotensin, cerebrovascular permeability alterations were studied in rats with nonpharmacologically induced hypertension.
Rats received horseradish peroxidase (HRP) intravenously, following which hypertensive was induced by clamping the abdominal aorta. Animals were sacrificed 2½ minutes after the onset of the hypertensive episode. The results show the same pattern of permeability alterations as observed in angiotensin-induced acute hypertension. In hypertensive animals local segments of penetrating arterioles in the temporal and parietal cortex showed increased permeability to HRP. Permeable vessels showed increased numbers of pycnocylic vessels as compared with controls. The interendothelial junctions revealed no alterations. A few vessels demonstrated HRP diffusely in endothelial cytoplasm but this was not associated with extravasation of tracer into the endothelial basement membranes. Enhanced pinocytosis appears to be the principal mechanism resulting in increased cerebrovascular permeability in this model as well. (Supported by Grant MRC MA-7191.

"QUANTITATIVE HISTOTOPOGRAPHY IN DOWN'S SYNDROME: COMPARISONS WITH NORMAL AGING AND ALZHEIMER'S DEMENTIA"

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An XY pen recorder linked potentiometrically to a sampling stage microscope permitted the plotting of topographic "scattergrams" to determine the precise cytoarchitectonic localization of neurofibrillary tangles and granulovacuolar degeneration in hippocampal regions of adult mongol's brains. The area (and hence volume) of six cortical "zones" surveyed was measured with a digitizer and programmable calculator. Larger more striking regional predispositions suggest that a common denominator in these cases is the enlargement of the 3rd ventricle associated with hydrocephalus or intraventricular cystic tumor. It is postulated that this interfrees with the function of periventricular structures through direct pressure, oedema or reduction in blood flow to cause this syndrome.

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Macrocrania as a feature of neurofibromatosis has not been much appreciated in the past. Some children with neurofibromatosis, half of whom were familial cases were assessed regarding head size. All had measurements of the occipital frontal cirumferences (O.F.C.) and standard skull x-rays. Six children had pneumo-encephalograms and 25 had CT-Scans to investigate macrornia or confirm the presence in nine patients of optic nerve or chiasm gliomas. Head size by O.F.C. was skewed towards upper percentiles for age. (15 of 61 at or greater than the 98th percentile) white stature (height) was skewed towards the lower percentiles for age. There was a low order correlation between macrornia and other features including intellectual defect, motor deficits, and EEG abnormalities.

From plain skull radiographs, the cranial capacity was estimated and found to be above the 95th in 60% of the patients. The volume of the sella turcica was likewise measured and found to be above the maximum normal volume in only 20%. From CT examination ventricular enlargement was found in 20% and ventricular asymmetry in 48%.

DISCREET LOCALIZATION OF HUMAN ENTRAPMENT NEUROPATHIES

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In human entrapment neuropathies, it is sometimes important to learn as precisely as possible the actual level of the nerve injury. Errors in localization may lead to occasional failure in the surgical decompression; for example, in the median nerve entrapment at the level of the carpal tunnel. It has been established by direct intra-operative stimulation of the involved nerves at short intervals that the functional abnormalities may be restricted to quite short segments of the nerve. This latter technique led us 4 years ago to attempts to better localize nerve injuries by combinations of stimulation or recording at short intervals (20 mm. or less) both proximal and distal to the level of the nerve injury by means of surface electrodes. The method was utilized primarily to investigate ulnar or peroneal nerve entrapments, but has been extended to the median nerve in the last 6 months following the publication of Kimura's observations with this technique in the median nerve (Brain, 102, 619-625). The surface electrode methods have had two important advantages:

1. It has been possible to identify abnormalities in nerve function, particularly in the median nerve at the level of the carpal tunnel segment not clearly identifiable by conventional EMG techniques.
2. The methods have made it possible to more precisely localize the level of the nerve injury, and hence establish the cause of failures in prior surgical decompression attempts.

The methods have therefore helped not only to extend our knowledge about the patho-physiology of human nerve entrapments, but represent a significant improvement with which nerve entrapments can be identified and precisely characterized in clinical laboratories.