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CORRELATION OF RADIOLOGICALLY ABNORMAL SELLAS WITH SMALL PITUITARY ADENOMAS IN AN UNSELECTED AUTOPSY SERIES

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Great stress is paid to the demonstration of the asymmetrical sella in the diagnosis of microadenomas of the pituitary gland by complex motion tomography. Though 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 75 ng/ml an adenoma was present in only one.

Tomograms were reported by two observers together, then independently, and were considered positive and compatible with a microadenoma in 27 cases. Correlation with pathological findings showed there to be 20 false positives and 26 false negatives. In the 7 cases in which the films and pathology were both positive, the x-ray findings corresponded to the site of the adenoma in 5. In 2 of these the adenoma was too small to believe the correlation valid.

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

BRAIN STEM GLIOMA: ABRUPT ONSET OF HEMIPLEGIA WITH RAPIDLY PROGRESSIVE COURSE

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We describe two children, ages 7 and 16, with malignant brain stem gliomas who presented with the abrupt onset of a right hemiplegia affecting the arm before the leg and sparing the face. They were hospitalized within one to five days and progressed to severe bulbar palsy requiring endotrachial intubation within 15 and 23 days from onset. The initial studies including computed tomography, radio-istope scanning, three-vessel angiography and lumbar puncture, failed to reveal the location or the character of the lesion. The abrupt onset and rapid course initially suggested vascular or inflammatory conditions. The subsequent neurologic deterioration localized the lesion to the brainstem 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 highly malignant tumor with rapid deterioration to complete bulbar paralysis.

CEREBRAL CYSTICERCOSIS IN CANADA: A REPORT ON THREE CASES FROM MONTREAL

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Montreal, P.Q.

Parenchymatous and meningeal cerebral cysticercosis were diagnosed in two Haitian and one Italian adult male immigrants residing in Montreal. The presenting problem was either a seizure disorder (partial or generalized) or headache or both. Examinations in two of three cases revealed increased intracranial pressure and on x-ray typical calcified subcutaneous nodules.

CT scans showed numerous hypodense cystic lucencies, 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 caused acute right hemiparesis and aphasia. Despite posterior 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 were of long duration and imported.

In immigrants cerebral cysticercosis should be included among the differential diagnosis of seizure and headache with 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 cysticerci.

CHEMOTHERAPY OF PEDIATRIC BRAINSTEM TUMORS

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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 adjuvant chemotherapy immediately following radiation therapy. Twenty-two patients received radiation therapy alone following diagnosis and later, at the time of tumor progression, were given chemotherapy. The median 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 adjuvant chemotherapy and 13 months for those given chemotherapy at the time of progression following radiation therapy. For patients receiving at least 2 courses of chemotherapy, median time to progression was 12.5 weeks for those receiving CCNU 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 chemotherapy, 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 12.5 weeks for those receiving a nitrosourea plus procarbazine. The results indicate no therapeutic benefit for adjuvant chemotherapy following radiation therapy in our patient group. While we were able to document response of some brainstem tumors to chemotherapy at the time of tumor progression, we found no clear advantage of one drug or drug combination over the others. New chemotherapeutic 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 this tumour 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 tumour 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 5. Three cases presented with multiple cranial nerve lesions and one with optic atrophy. A history of treatment of Otitis Media was common.

Sixty percent of patients with tumour arising from the nasopharynx presented with variable cranial nerve lesions and optic atrophy in one.

Two patients with tumour arising from the retroperitoneal area presented with multiple root disease. One patient with RMS arising from the neck presented with Horner's syndrome.

There were 10 patients with orbital tumours and all presented with proptosis rather than any specific neurological manifestation.

The importance of early clinical diagnosis of this highly malignant tumour will be stressed and the rapid neurological progression illustrated by the case material.

INTRACRANTIAL PRESSURE MONITORING

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I.C.P. monitoring has been in clinical use for over 20 years and is now advocated for management of serious head trauma and diffuse encephalopathies such as Reye's Syndrome.

This display outlines the pathophysiology of raised I.C.P., the principal points of management and the methods used to measure 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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In 24 adult patients with severe head injury, the intracranial pressure (ICP) was monitored for one to 6 days with a ventricular cannula. 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 Mannitol and, in 9 instances, barbiturate coma. The EEG is a practical method of monitoring barbiturate dosage. In selected cases concentrations of barbiturate 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., no correlation between outcome and ICP were detected in this series. Eleven patients were initially decerebrate and one flaccid; five of them died and 3 remained in a persistent vegetative state. Two of the remaining patients (Glasgrow coma sum 5, 6, 7) died. Elevated ICP is 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 cannula 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

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The flow chart is a well-recognized tool that is useful in the preparation of computer programs. Its utility for this purpose resides in the fact that the computer proceeds from operation to 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 described in general terms by the presence or absence of very few attributes, since there is only a small number of diagnostic and therapeutic procedures that may be applied and since the choice among these procedures 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 can account for virtually every possible hospital course of a patient with craniocerebral trauma. The flow chart has been used in the retrospective analysis of the hospital course of 30 patients admitted to Sunnybrook Medical Centre with a diagnosis of multiple trauma. The method has proved useful in identifying delays in the application of treatment and deviations from the expected course. Since the chart embodies treatment priorities it serves to teach correct decision-making. Finally, since even a complicated hospital course may be succinctly 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 damage following cardiac arrest. Three of the patients survived, one died a cardiac death on day three, and three appeared to proceed on to "brain death". Thiopental was administered from 4-72 hours and maximum blood levels attained ranged from 30 to 135 mg per litre 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 finding 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 iso-electric 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 intravenous barbiturate and maintained blood levels) was uniformly associated with a bad outcome. The EEG patterns' correlation with the barbiturate 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 ½ 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 hypertension (BP > 160/90), 14 had excessive anticoagulation (PT or PTT > 2 ½ control), and 8 had previous cerebral infarction. Seventeen had two or more risk factors. The prognosis was poor: intracerebral hemorrhage - 12/17 died, 3 17 left with major neurological deficit; subdural hematoma - 2 8 died, ⅛ major deficit; subarachnoid hemorrhage - ½ died.

Anticoagulants are still an important cause of intracranial hemorrhage. The risk of hemorrhage is greater if the patient is hypertensive, has had a cerebral infarct, or is receiving excessive anticoagulants. The incidence of intracerebral hemorrhage in our series is greater than in previous reports, probably due to improved detection by computerized tomography brain scans.

INTRAVENOUS BARBITURATES IN COMATOSE HEAD INJURIES: LATE FAILURE TO CONTROL ELEVATED INTRACRANIAL PRESSURE

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Toronto, Ontario

Intravenous barbiturates have been shown to effectively decrease raised intracranial pressure when conventional therapy has failed in comatose patients with head injuries.

Continuous intracranial pressure monitoring was performed on twenty-six patients with major closed head injuries at Sunnybrook Medical Centre over the past year, utilizing either an intraventricular catheter or a subarachnoid screw. All patients received conventional management which included controlled hyperventilation to a $PaCO_2$ of 25-30 torr, and intravenous dexamethasone and mannitol.

In fifty percent, the intracranial pressure remained elevated at greater than 25 torr. These patients received intravenous pentobarbital to maintain serum barbiturate levels of 2.5-4.5 mg %. Ninety-two percent showed an initial stable reduction of intracranial pressure, but effective control of elevated intracranial pressure was not subsequently maintained in fifty-four percent of the whole group.

In the majority of late failures, the reasons for loss of control were not obvious. In two patients, increase in $PaCO_2$ coincided with loss of control and in one instance, premature reduction of barbiturate dosage may have been causative. Despite correction of the apparent precipitating abnormality, subsequent control intracranial pressure elevation was achieved in only one patient.

CEREBRAL EDEMA IN ACETAMINOPHEN OVERDOSE

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

A 15 year old girl was admitted after taking 30 grams of Acetaminophen (Tylenol). She received N-acetyl cysteine 10 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 and transtentorial herniation.

In our patient, severe cerebral edema and increased intracranial pressure were present as proven at craniotomy, and appeared to be the immediate cause of death. Intracranial pressure monitoring would appear indicated in patients with coma from severe acetaminophen overdose. Further studies are needed to determine if barbiturate therapy and possibly surgical decompression can increase survival in acetaminophen overdose when intracranial pressure cannot be controlled by conventional methods, as the liver may be able to regenerate if the patient does not die from CNS causes.

THE RELATIONSHIP OF A FOCAL MOTOR NEUROLOGICAL DEFICIT TO INTELLECTUAL FUNCTION IN EXTERNAL HEAD TRAUMA

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Toronto, Ontario

The relationship between external head trauma and cerebral function is of considerable concern to the clinician. In addition, the presence or absence of a neurological deficit and its relationship to be severity of intellectual impairment following head injury has practical significance for patient management. The present study examined the relationship between hemiparesis following head trauma and intellectual function on a standardized test, the Wechsler Adult Intelligence Scale. Three groups of patients, similar in age and education were compared on measures of overall abilities (WAIS Full Scale IQ), verbal (Verbal IQ), visual spatial abilities (Performance IQ), and subtests scores. Group One consisted of patients with external cerebral trauma and hemiparesis as sequelae; Group Two consisted of patients with external cerebral trauma and no focal motor deficits; Group Three consisted of psychiatric patients, serving as a control.

The results of the study showed that as a group, patients with head injury obtained significantly lower scores than the control subjects on the Full Scale IQ ($p<.001$) Verbal IQ ($p<.02$) and Performance IQ ($p<.001$). Subjects with hemiparesis (Group One) obtained significantly lower scores than control subjects on both language and visual spatial tasks and significantly lower scores than Group Two on six subtests. Patients without focal motor deficits (Group Two) obtained significantly lower scores than the control group on selected visual spatial subtest scales.

The implications of this preliminary study are discussed with respect to patient adjustment and therapeutic management. The presence of motor sequelae in patients with head injury should alert the clinician to the increased likelihood of intellectual impairment; however the absence of focal motor signs does not preclude the presence of intellectual deficits.

SPECTRAL ANALYSIS OF THE ELECTROENCEPHALOGRAPHIC RESPONSE TO EXPERIMENTAL CONCUSSION IN THE NON-ANESTHETIZED RAT

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This investigation studied the pathophysiology of concussion by recording the electroencephalographic response to a concussive blow administered to 27 Sprague-Dawley rats, previously implanted with chronic epidural EEG electrodes. The blow was administered to the head by a blunt dart, shot from a spring-loaded pistol. The calculated momentum (mv) and kinetic energy (½ mv ^{2}) were constant upon repeated testing. Spectral analysis of the continuously recorded EEGs was performed off-line on a PDP8 computer. Statistical analyses were done using one-way analyses of variance and Duncan's multiple range test. Fifteen rats were concussed to Stage 1-2, that is, transcient loss of consciousness without associated autonomic disturbances. Their EEGs were characterized by a decrease in the power spectra of the alpha, beta, and theta frequencies by 25%, 37%, and 10% respectively. The delta spectrum alone was increased, by 15%. These EEG changes were similar in pattern, but less profound than those observed in Stage 3-4 concussion. The EEG power spectra returned to control values within 2 hours of the concussion. These observations support the theory that concussion represents a reversible alteration in cerebral cortical activity, rather than a transient dysfunction of the reticular activating system alone. In fact, previous authors have demonstrated EEG hypersynchrony in response to a lesion in the reticular activating system — a finding directly opposite to the above results. (Supported by the M.R.C.)

NEUROSURGICAL APPLICATIONS OF DOPPLER

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Sunnybrook Medical Centre, Toronto

Two hundred and eight neurosurgical patients were referred for continuous wave cerebrovascular Doppler assessment. The clinical diagnoses were transient ischemic attack (TIA), recovering ischemic neurological deficit (RIND) or cerebral infarction in 134 patients (64.4%), amaurosis fugax in 33 (15.9%), asymptomatic bruit in 25 (12%) and other in 16 (7.7%). One hundred and six (51%) of the cases were in connection with endarterectomy, 26 (12.5%) with extracranial-intracranial (EC-IC) anastomosis.

Cerebral angiography was also performed on 89 patients proving the Doppler examination 95% accurate in the assessment of the cervical carotid arteries. In another 11 (5.3%) the surgical (or autopsy) specimens were compared with Doppler and angiographic results showing that the former provided a more accurate assessment of the degree of luminar narrowing in high grade stenoses. Plaques were detected in 3 of the 18 post-endarterectomy cases, while 10 of 46 plaques were found to be progressive by Doppler in non-endarterectomized arteries. The ultrasound method was accurate in post-operative assessment of all 12 EC-IC anastomoses whereas angiography failed to detect existing shunt function until up to 11 months post-operatively in 2 patients. Eleven of 14 patients (78.6%) referred because of incidentally discovered asymptomatic bruits had atherosclerotic plaques demonstrated by Doppler. On the other hand, of 11 patients with postoperative bruits, only 2 (18.2%) demonstrated recurrent stenosis.

Doppler proved useful in the pre- and post-operative evaluation of carotid endarterectomy patients, in assessing patency of EC-IC anastomoses and for following asymptomatic bruits.

INTRACRANIAL VENOUS THROMBOSIS OCCURING IN EARLY PREGNANCY

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Ottawa General Hospital

Intracranial venous thrombosis in pregnancy classically occurs in the post-partum or late gestational 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 temporoparietal hemorrhagic lesion surrounded by edema and later extensive edema with focal hemorrhagic areas bilaterally, especially parasagittally. Angiography showed the disappearance of previously seen venous structures. Death occurred after 17 days and extensive venous thrombosis with widespread cerebral infarction and edema were found at autopsy.

The second case was a 29-year-old who presented with status epilepticus in the fourth month of gestation. She remained in coma up to and following delivery. The diagnosis was made angiographically.

The diagnosis of intracranial venous thrombosis is facilitated by computerized cranial tomography. The outcome of cases in early pregnancy is less favourable than those presenting post-partum and treatment remains symptomatic.

"ASSESSMENT OF EXPERIMENTAL MICROVASCULAR SURGERY BY SCANNING ELECTRON MICROSCOPY"

G-E. OUAKNINE, G. MOHR AND J. HARDY
Notre-Dame Hospital and University of Montreal

This poster presents the surgical evaluation by scanning electron microscopy (S.E.M.) of various microvascular operations performed in 60 rabbits and 40 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 after few hours of application. The HEIFETZ 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 ACLAND and KEES microclips demonstrated the least traumatic modifications. The best suture material to minimize the endothelium lacerations appeared to be 11.0 and 12.0 threads mounted on 50 needles (⅜ circle). The continuous suturing technique appeared more traumatic and thrombogenic than the interrupted stitches technique.

Formation of fibrin and platelet agregation on the intima and suture line was often observed. Loops of threads found in the lumen, mainly due to continuous sutures appeared less thrombogenic than endothelial ruptures due to surgical manipulations. Intimal modifications and reconstruction were accurately demonstrated by the S.E.M. like the proliferation of endothelial cells covering the edges of the suture line, filling out intimal gaps and lacerations or covering threads.

In conclusion, the S.E.M. appears to be the method of choice for studying the differents endothelial modifications due to the surgical trauma.

MOYA-MOYA DISEASE: A CLINICAL PATHOLOGICAL CORRELATION

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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 arterial trunks with a collateral system of fine vessels in the region of the basal ganglia and transdurally) 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 ½ 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 proliferation of intimal connective tissue, splitting of the internal elastic lamina and chronic inflammatory cells in the adventitia, particularly in the occluded areas). Immunofluorescent stains IgG, IgA, IgM, C_3 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.

CAROTID DOPPLER IMAGING IN A COMMUNITY HOSPITAL

D.L. CHEW AND R. CURRY
Niagara Falls

When facilities for arteriography and invasive vascular surgical procedures are some distance away, the decision to send a patient many miles from home for potentially dangerous investigation and therapy is fraught with uncertainty. The doppler imaging system, "Echoflow", is a non-invasive ultrasound technique allowing the practising physician to distinguish between those patients with obstructive disease of the carotid arteries amenable to surgical intervention and those patients for whom surgery is not indicated.

Our experience over the past 12 months includes over 400 evaluations. We have verified various reports regarding clinical signs in vascular disease as well as collecting information on a number of different diagnostic conditions which previously would not have been identified. This report illustrates how the results of a non-invasive imaging system can identify vascular disease and the subsequent effect on the management of such patients.

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

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Computed mapping of electroencephalogram (CME) is a newly developed microcomputer system to display equipotential maps of square root of average powerspectra over each frequency band on color television. This new device was employed in examination of 22 aphasics due to cerebral infarction in comparison with CT and regional cerebral blood flow (rCBF) study. High voltage foci in slow wave bands and asymmetry of alpha distribution were regarded as functional lesions on CME. In 12 patients, high voltage foci 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 left hemispheric asymmetry of alpha distribution in spite of no abnormal finding on CT until 8 months after the episodes. Comparing with conventional EEG interpretation, CME is more useful in diagnosis of functional lesions topographically and objectively though the source of the data is same as conventional EEG.

CEREBRAL BLOOD FLOW AND ARTERIAL VASOSPASM IN PATIENTS WITH SUBARACHNOID HEMORRHAGE

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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 Botterell classification were $37 \div 6$ ml 100g min, $35 \div 7$ ml 100 min and $32 \div 6$ ml 100 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 49 of 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 $39 \div 6$ ml 100g min, while flow was $31 \div 6$ ml 100g in in those patients with spasm ($p<0.01$).

The results demonstrate that CBF is significantly reduced in patients of poor clinical grade, and also that arterial 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

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We have previously documented an increase incidence of cardiac arrhythmias, elevated serum cardiac enzymes and ischemic ECG 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 focal myocardial lesions in these patients who die acutely.

We have systematically studied cardiac sections in 70 consecutive autopsies, using histochemical methods sensitive to mitochondrial 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, brain tumors and spinal subarachnoid hemorrhage. In 36 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.

MONITORING REQUIREMENTS DURING CAROTID ENDARTERECTOMY

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Concern for the possibility of shunts causing emboli or ischemia led to a progression of intra-operative monitoring proce-

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dures during arterial clamping without shunting. Initially, only carotid artery stump pressure less than 50 torr guides shunt intervention. Subsequently, a two lead EEG oscilloscope display further added diagnosis of possible ischemia. With the addition of multi-channel EEG recording the prime indication for shunting became the appearance of EEG ischemic changes. Lastly, manipulation of the systemic arterial pressure (SAP) with drugs, per cent inspiratory oxygen (FiO_2) and arterial CO_2 pressure (PaCOd22) by ventilation has been used to control ischemia prior to shunt intervention. Throughout all cases the occlusion time was kept to a minimum with the mean being fifteen minutes.

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

Our results suggest that in most situations continuous multichannel EEG monitoring provides enough data to allow sufficient time for intervention with either SAP FiO_2 $PaCO_2$ manipulation and or shunting.

PROXIMAL NERVE CONDUCTION IN IDIOPATHIC CHRONIC RELAPSING POLYNEUROPATHY

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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 ataxia, 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 patients, mild abnormalities in distal motor nerve conduction were 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 conductions in spite of a severe sensory ataxia.

These results confirm the diffuse distribution of the lesions in idiopathic chronic relapsing polyneuropathy, often with a greater proximal involvement. F wave determinations show definite abnormalities in proximal nerve function in patients with idiopathic chronic relapsing polyneuropathy who are clinically normal and have normal, or borderline distal nerve function.

MIDDLE MOLECULE CLEARANCE IN UREMIA: EFFECTS ON PERIPHERAL NERVE CONDUCTION AND PLATELET FUNCTION

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W.F. CLARK, A.L. LINTON

Several investigators have suggested that toxins of "middle" as opposed to smaller, molecular size may be responsible for polyneuropathy and other complications of uremia. Using a crossover design, we tested this hypothesis on 12 patients, 9 having polyneuropathy. A baseline period of observation established that each group of 6 patients was similar. At two-monthly intervals for six months, each group was alternated between either conventional hemodialysis by a Homoclear dialyzer, or hemodialysis by a Sorbiclear dialyzer. The latter contains a two layered membrane of cellulose and activated charcoal designed to clear middle molecules. At the beginning and end of each two-monthly period the patients were tested for clearance of middle molecule plasma fractions, platelet function (bleeding time, platelet count and platelet aggregation), and nerve conduction (median motor and sensory conduction velocities, compound action potential amplitudes, and F response latencies). A paired sample T test applied to this crossover design was used to analyse the data.

Despite improved clearance of middle molecules and improved platelet function during Sorbiclear hemodialysis, no significant change in median nerve conduction occurred. There are several possible explanations for this result. However, we speculate that there is more than one "toxin" in uremia, the one affecting platelet being different than the one affecting peripheral nerve.

DOES THE USE OF CHAIN-SAWS PRODUCE NEUROPATHY IN CANADIAN FORESTRY WORKERS

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As part of a pilot study on the vibration white finger (VWF) syndrome in Quebec foresters, neurological examination and nerve conduction studies were performed. Eighteen of 29 chain-saw users reported having white fingers when working in the cold, compared with 2 of 12 forestry workers not using saws. Mild clinically detectable sensory abnormalities were more common in the hands and feet of the sawyers (with and without VWF symptoms) compared with the non-sawyers.

The motor and sensory nerve conduction studies, performed on the forestry workers as well as on a group of sedentary persons, showed the following:

1. Distal sensory action potentials were of considerably lower amplitude in forestry workers.
2. Motor conduction velocities and distal motor latencies were similar in all groups.
3. Increased numbers of mild ulnar nerve conduction blocks across the elbow were found in the sawyers with VWF, otherwise no entrapment neuropathies were detected in any group.

It is tentatively concluded that the abnormalities demonstrated probably relate to manual work, rather than to the specific use of chain-saws.

PERIPHERAL NERVE SEGMENTS PROMOTE NEURITE OUTGROWTH FROM EMBRYONIC SENSORY AND SPINAL CORD NEURONS IN VITRO

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Single cell cultures of neurons from 7 and 8 day chick embryo dorsal root ganglia and spinal cord have been established and used to detect the presence of factors from peripheral nerve segments of adult mice that promote neurite outgrowth.

In the absence of NGF, 8 day sensory neurons show little or no neurite outgrowth; a single 0.5 mm segment of peripheral nerve (in a 200 microlitre culture well) induces a neurite outgrowth response from sensory neurons that is greater than the response seen with concentrations of NGF that promote maximal neurite outgrowth. This response can be inhibited by approximately 50% upon coincubation with antibody to purified mouse BNGF at concentrations that produce 100% inhibition of NGF-induced neurite outgrowth. Ligation of the nerve in vivo results in a rapid decrease of neurite outgrowth promoting activity proximal to the ligature.

Spinal neurons from 7 day embryos have been established in culture in a defined serum-free medium. In this setting, adult mouse peripheral nerve segments cause a quantitative enhancement of neurite outgrowth.

These observations suggest the presence within peripheral nerve, of factors, including NGF, that are transported to sensory and spinal cord neuronal cell bodies from the periphery and act to maintain the functional integrity of these neurons.

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EVALUATION OF RADICULOPATHIES AND PLEXOPATHIES USING SEGMENTAL SENSORY STIMULATION

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Available electrophysiological methods have been diagnostically disappointing for evaluating radiculopathies and plexopathies. Poor specificity of segmental stimulation and difficulty in recording from proximal structures being the main problems. Recording somatosensory responses (SEPs) should be potentially helpful in these areas, but as conventionally evoked, multisegmental stimulation is utilized. In this study, we evoked SEPs by stimulation of different sensory nerves each subserving one or at most two segmental levels. The following sites were stimulated: thumb (median, C_6); between the second and third digits (median, C_7); fifth digit (ulnar, C_8); ventro-lateral forearm (musculocutaneous, C_5) and the following sensory nerves at the ankle - saphenous ($L_{2,3}$); superficial peroneal ($L_{4,5}$); sural ($S_{1,2}$). Sensory nerve action potentials (SNAPs) were simultaneously recorded at the elbow and knee following stimulation of upper and lower extremity nerves respectively. This allowed assessment of more distal parts of the peripheral nervous system. "N_{20}", the initial cortical event of SEPs evoked by upper limb stimulation measured (msec): 22.2 ± 1.2 (N = 33); 22.5 ± 1.1 (N = 22.0 ± 1.4 (N = 32) and 17.4 ± 1.2 (N = 24) for each stimulation site respectively. "P_{40}", the equivalent peak evoked by lower limb stimulation measured (msec) 43.4 ± 2.2 (N = 25); 39.9 ± 1.8 (N = 26) and 42.1 ± 1.4 (N = 24) respectively for each nerve stimulated. Twelve patients were studied with myelographically proven radiculopathies due to disc disease, six of whom had operative confirmation of their lesions. In each case, the SEPs evoked by stimulation of an appropriate segmental level(s) showed evidence of slowed conduction and or block in conduction. In 5 other patients with brachial plexopathies due to various causes, it was possible to delineate which trunk(s) or cord(s) were primarily involved. The technique holds considerable promise for the electrophysiological investigation of proximal segments of the peripheral nervous system.

TRANS-SPHENOIDAL ADENOGRAPHY

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Trans-sphenoidal resection of pituitary tumours has usually been preceded by pneumoencephalography. Although this technique is helpful, it has some limitations. It can demonstrate only the superior surface of the tumour and even this may not be clearly outlined. We wish to present another and, in our experience, superior method of tumour delineation.

After fenestration of the sellar floor, but before incision of the dura mater, a lumbar puncture needle is introduced into the tumour. A small amount of contrast medium is gently injected and its passage is monitored by fluoroscopy. Radiographs are obtained for more precise study. In the first 10 patients an oily contrast medium was used, but in 10 subsequent patients we have found a water soluble medium more satisfactory. It disperses uniformly throughout the tumour, outlining its intrasellar and perisellar dimensions. In 12 patients, defects in the tumour capsule were indicated by spread of the contrast medium into the subarachnoid space, the third ventricle, the cavernous sinus and/ or the carotid sheath. Contrast medium remaining within the lesion as operation proceeds indicates that tumour excision is yet incomplete.

The advantages of this technique will be illustrated by specific radiographs.

OCCURRENCE OF CREUTZFELDT-JAKOB-LIKE VACUOLAR CHANGE IN ALZHEIMER'S DISEASE AND OTHER CONDITIONS

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The intracerebral inoculation of brain from a patient with Alzheimer's disease produced in the primate a neurological disorder with all the pathological features of Creutzfeld-Jakob disease. (Rewcastle, Gibbs and Gadjusek, J. Neuropathology 37: 679, 1978). Study of the original biopsy demonstrated the sparse presence of large vacuoles containing multivesicular structures indistinguishable from those encountered in Creutzfeld-Jakob disease and considered typical for this latter disorder.

Review of 7 other biopsies from patients with Alzheimer's disease has demonstrated by electron microscopic study the presence of identical vacuoles in each instance. In order to test the specificity of this observation, we have further studied 7 biopsies from a variety of pathological conditions and an equal

number of "normal" biopsies where no structural changes were present at either the light or electron microscopic levels to indicate a pathological process.

We conclude that this vacuolar change is not specific for Creutzfeld-Jakob disease. Identical changes are also present in Alzheimer's disease, though much less frequently, and in inflammatory conditions resulting from known or suspected viral agents. Rather than being taken to indicate the nature of the infective agent, they more likely represent a tissue reaction of unknown nature that would seem to be restricted in our material to damage resulting from suspected viral agents, thus suggesting indirectly that Alzheimer's disease might also have an infective aetiology.

NEUROBEHAVIORAL DYSFUNCTION IN MULTIPLE SCLEROSIS: A MAJOR CAUSE OF ADAPTATIONAL BREAKDOWN?

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Although M.S. is the most common CNS disease in young adults, relatively little has been learned about the nature and extent of neurobehavioral deficits associated with cerebral demyelination. The neuropathological literature on cerebral disease and the clinical epidemiological and neuropsychological literature on behavioral deficits is reviewed. It is proposed that neuro-psychological dysfunction is associated with overall functional disability but not with disease duration or extent of disease in discrete neurological systems. Fifty-five sequential referrals to a Neuropsychology Service from a Regional M.S. Clinic with definite 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 small number of recall tests confirmed the tendency for general cerebral deterioration in the more disabled patients. Duration of disease and extent of neurological disorder in separate systems showed no systematic relation to psychological performance. Evidence of structural cerebral disease in neuro-psychologically disabled patients is offered by examination of a subset of patients submitted to CTT scans. Suggestions are made as to why neurobehavioral symptoms are given little research emphasis and why patients, families and professionals receive relatively little information about the unique adaptational demands these deficits pose. A brief formulation for coping with neurobehavioral abnormalities is proposed.

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

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In the course of a systematic investigation of different forms of spinocerebellar 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 father, all siblings, were retarded and could not walk, and may have been similarly affected. Some members of this family were previously studied by Rothman and Olanow.

The five affected siblings, ranging in age from 19 to 27 years, had delayed developmental milestones. Recurrent vomiting and seizures precipitated by fever were present in all five in the first year, and remitted spontaneously. By the mid-teens, a combination of pyramidal, cerebellar, posterior column and peripheral nervous system involvement developed, the older siblings being more severely affected. There were also severe progressive pes cavus and scoliosis. The patients had extraocular movement abnormalities, suggesting dysfunction in cerebello-vestibular pathways. Dysmorphic features included: large heads, high foreheads, long narrow facies, deepset eyes with synophrys, high-arched palates, irregular teeth and prognathism.

Urinary and plasma amino acid chromatography and chromosome studies were normal. EEG studies were also normal, although some had shown epileptiform abnormalities during infancy. EMG studies suggested involvement of dorsal root ganglia or sensory neurones as well as central sensory pathways, and possibly some anterior horn cell involvement. Sural nerve biopsy showed evidence of axonal degeneration with loss of large myelinated fibers. Gastrocnemius biopsy showed mild denervation with probable reinnervation.

This syndrome does not correspond to any recognized spinocerebellar or pyramidal system degeneration, to our knowledge.

FAILURE OF REGENERATING DORSAL ROOT AXONS TO REGROW INTO THE SPINAL CORD

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The relative role of neuronal and sheath cell responses in mammalian regeneration has not been clearly delineated. Previous studies using CNS or PNS transplants inserted into peripheral nerves or spinal cords suggest that injured CNS glia blocks axonal regrowth while Schwann cells may facilitate the elongation of spinal axons (Aguayo et al. Soc. Neurosci. Symp. 4:361-383,1979). In the present experiments the fate of peripheral axons regenerating along a continuous path of sheath cells that includes both schwann chells and CNS glia (Berthold & Carlstedt, Acta Physiol. Scand. 446: 23-42, 1977) was studied in 32 adult Sprague Dawley rats up to 11 months after the lower lumbar-upper sacral spinal roots were crushed at a site approximately equidistant from the dorsal root ganglion and the cord, thus avoiding direct damage to CNS glia in the root entry zone. Axons were found to regenerate along the Schwann cell ensheathed segments but stop at the boundary between PNS and CNS. Hyperplastic astrocytic processes appear to play an important role in blocking the regrowth of these axons.

SPINAL SUBDURAL EMPYEMA WITH ANAEROBIC ORGANISMS

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We describe a spinal subdural abscess in a 15 year old boy. The organisms isolated were anaerobes usually associated with infections of the paranasal sinuses and mouth, including Fusi-bacteria, Actinomycosis and Bacteroides. There is no previous report in the English literature of these organisms being implicated in spinal subdural abscess.

Onset of the illness was subacute with low back pain, intermittent fever, parapareses and urinary incontinence developing over one week. Laminectomy was performed. The dura, which was bulging, was incised; copious viscid pus was released and the subdural space irrigated. Diagnosis was made by microscopy of the pus and confirmed by culture. Antibiotic treatment was with Penicillin, for eight weeks, Chloramphenicol and Metronidazole. Recovery was complete and he remained well six months later. Differential diagnosis is mainly from meningitis and epidural abscess. The aetiology is obscure but may be post traumatic.

THE CORTICAL SOMATOSENSORY EVOKED POTENTIAL IN ACUTE SPINAL CORD INJURIES

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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 injury in incomplete, alterations 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 trauma and progressive normalization of the wave for are senistive early indications 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. We have also shown that a 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

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Phagocytic cells are commonly present in central nervous system (CNS) lesions in many diseases; thus implicating free radical production in the pathology. 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 — H_2O_2 must be removed by catalase. Low levels of catalase could lead to increased tissue damage or increased levels might follow 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 flotation technique developed by Gagnon was utilized with bovine liver catalase (Pharmacia) standards. Fresh 3% H_2O_2 (Pharmacia) was stabilized with EDTA. All test specimens were examined concomitantly with standard dilutions. Multiple Sclerosis CSF gave a mean of 18 ± 4 U dl, while normals gave a mean of 11 ± 2 U/dl. Chronic degenerative CNS disease gave a mean of 9 ± 2. Patients in exacerbation gave values significantly greater than those with chronic progressive or stable disease. Multiple sclerosis does not appear to be due to abnormally small amounts of catalase.

LISSENCEPHALY: THREE CASES DIAGNOSED BY CT SCAN

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Lissencephaly 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 agyria, with associated cerebral anomalies. Microscopically, heterotopic nests of grey matter were found in the subependyma which gave rise to the theory that the basic defcct was an abnormal migration of neuroblasts in fetal life. Several studies show convincing evidence for a genetic origin of this defect, while others present isolated examples.

We recently observed three patients with the lissencephaly 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 the syndrome of pachygyria-agyria providing fuel for speculation as to the etiology of this aberration of development.

HEREDITARY DEMYELINATING INFANTILE NEUROPATHY WITH UNUSUAL CLINICAL AND PATHOLOGICAL FINDINGS

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

S.B., age 12, showed nystagmus, slow motor development and progressive gait and limb ataxia since early infancy. He developed pes cavus and mild progressive distal lower limb weakness. Reflexes became absent, vibration sense was decreased distally and pseudoathetoid movements were ob-

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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 action potentials were recordable with surface electrodes at age 3. Needle electrode study showed no active denervation, recorded action potentials were small and dispersed, estimated MNCV 3.5 m/sec. CSF protein 37 mg %.

Father, age 42, had slowed motor development, pes cavus and slowly progressive distal atrophy and weakness. MNCV was 15 m.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 partially demyelinated along 80% of their lengths. Globules of folded 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.

VISUAL EVOKED AND AUDITORY BRAINSTEM RESPONSES IN PATIENTS WITH SPINAL CORD DEMYELINATION

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Pattern visual evoked responses (VERs) and auditory brainstem responses (ABRs) were studied in 26 patients aged 18 to 63 with a clinical diagnosis of spinal cord demyelination. All patients had symptoms and signs implicating the spinal cord. None had ever had symptoms or signs suggestive of lesions above the foramen magnum. Almost all had had myelography. Duration of symptoms ranged from 1 month to 13 years. 5 patients had had symptoms for less than 1 year.

For VERs, positive peak latency and inter-eye latency difference were measured. For ABRs, wave I-III, III-V, and I-V intervals were measured, as well as interear differences for these intervals. All values were compared to our normal control group (N=24) of similar age.

Using 3 standard deviations (SD) above the normal mean as the upper limit of normal, VERs were abnormal in 17 patients, and ABRs in 13. ABRs were abnormal in 3 patients with normal Vers. Only 6 patients were normal on both tests.

Using 2 SD above the normal mean, VERs were abnormal in 20, and ABRs in 18. Only two patients were normal on both tests.

These results are compared with a second group of 20 patients with predominantly spinal cord demyelination but with some clinical features suggestive of disease above the foramen magnum.

VER and ABR testing is a useful investigation in patients with demyelinating disease affecting predominantly the spinal cord.

FAVOURABLE RESULTS WITH THE SYRINGO-SUBARACHNOID SHUNT FOR THE TREATMENT OF SYRINGOMYELIA

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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 tonsillar ectopia. There were 14 patients with idiopathic syringomyelia, three with post-traumatic syringomyelia, one with spinal arachnoiditis and one with a spinal arachnoid cyst. The operations were performed with an operating microscope and attention was directed to preserving the arachnoid membrane to ensure proper placement of the distal end of the shunt in an intact subarachnoid space. A Pudenz ventricular catheter 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 with cessation of progression 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.

SPINAL CORD SWELLING IN MULTIPLE SCLEROSIS

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In subacutely 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 tumour. Widening of the cord, similar to that seen with intramedullary tumour, was seen in the upper thoracic cord in two patients and in the mid-cervical cord in one. However, CSF protein electrophoresis showed oligoclonal IgG banding in each case. This, in conjunction with the subsequent clinical course, confirmed a diagnosis of multiple sclerosis (MS) in all three patients. Repeat myelography after 21 days in one case showed a reduction in cord swelling.

Two of our patients had myelography with water-soluble contrast medium which may facilitate demonstration of minor cord swelling associated with MS which might otherwise be missed with oil myelography. These cases demonstrate that spinal cord enlargement may be seen in MS. This observation is of special importance in evaluating myelographic findings in subacute myelopathy. It underlines the importance of alternate diagnostic techniques, particularly the CSF protein electrophoresis, for a correct diagnosis and avoidance of surgical intervention in demyelinating myelopathies.

RELATIONSHIP BETWEEN CORD INJURY, SPINAL COLUMN INJURY AND RECOVERY IN 358 SPINAL CORD INJURED PATIENTS

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The management of acute spinal cord injured patients is made difficult because of the multitude of possible combinations of types of spinal cord injury and types of spinal column injury. There have been very few studies which have attempted to determine if there are any consistent patterns of relationship between the cord and the vertebral injuries. In addition there have been very few attempts to relate these two factors to recovery. It was felt that it would assist the management of acute cord injured patients to attempt to identify relationships between the neurological and boney injuries and recovery. Diagnosis of the injuries in the acute stage might be improved by a knowledge of the frequencies with which certain types of neurological injuries are associated with certain types of vertebral injuries. The establishment of prognosis or the treatment likely to be most successful for certain associations of neurological and vertebral injuries might also be facilitated. This study is based on a computerized analysis of the records of 358 patients managed between 1948 and 1973.

It was found that fracture-dislocations produced more complete cord injuries and a higher incidence of death than other types of injuries, while those with compression fractures had a lower incidence of death and fewer complete cord injuries. There was an effect of level of injury on completeness with thoratic injuries tending to be more complete. Level of injury had no effect on recovery. The direction and severity of dislocation also affected the degree of neurological deficit on admission. With the exception of falls at home, the type and level of boney injury were not related to the cause of the injury. Falls at home produced more compression fractures and more thoraco-lumbar injuries than traffic, sports-recreational or work-related causes of cord injury.

"AN EVALUATION OF SILASTIC SHEATHING IN NEUROLYSIS OF THE ULNAR NERVE"

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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 effectiveness of sheathing the ulnar nerve with silastic, following neurolysis, as a means of minimizing the effects of recurrent scar formation.

A total of 43 procedures was 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 electrophysiological 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 neuropathy or had a metabolic predisposition towards entrapment.

It is concluded that simple neurolysis, 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

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To investigate the observation that testing of 256 Hz vibration sense is valuable in assessment of peripheral nerve dysfunction, we analysed 120 consecutive patients referred to an electrodiagnostic 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 mseconds, compared to 4.6 ± 0.9 mseconds for the others (P 0.005).

It is concluded that, contrary to current 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 CERVEAU ISOLE CATS

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Gloor and others have suggested that generalized spike discharges 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 Cerveau Isole preparation in cats. A significant association between spikes and spindles occurred in 23 out of 27 foci. Immediately after establishment 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 latent period of 1 to 40 minutes, spikes occurred only during a spindle (i.e. 100% association). No change in the spindles occurred after establishment of the epileptic focus.

The morphologic relationship of the focal spike discharge to the spindle wave was assessed using a PDP1160 computer and will be presented. These findings will be discussed with respect to the thalamocortical interaction in the production of focal Penicillin spikes.

PERIPHERAL NEUROPATHIES IN CHILDHOOD

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In children, electrodiagnostic studies are required more often to aid in the diagnosis of polyneuropathies than in the investigation of isolated nerve lesions. The author has tested 126 children with polyneuropathies as compared to only 35 with mononeuropathies due to trauma or entrapment (excluding facial palsies). The former may be grouped as follows -

Genetic	-	hypertrophic type of Charcot-Marie-Tooth disease	20
59		Friedreich's ataxia	15
		Krabbe's leucodystrophy	6
		metachromatic leucodystrophy	4
		ataxia-telangiectasia	4
		hereditary sensory neuropathies	3
		others, e.g mucopolysaccharidoses	7
Metabolic	-	hypothyroidism	6
13		subacute necrotizing encephalomyelopathy	3
		diabetes mellitus	2
		chronic renal failure	2
Inflammatory	-	Guillain-Barré syndrome	16
20		others, e.g. diphtheria, post-measles	4
Nutritional	-	coeliac disease	1
1			
Toxic	-	vincristine	11
18		diphenylhydantoin	7
Cryptogenic	-	chronic sensori-motor neuropathy	9
15		others, e.g. dysmyelination	6

These polyneuropathies may now be divided into axonal (e.g. in Friedreich's ataxia) and demyelinating (e.g. in leucodystrophies, diphtheria). Electrical studies may assist in the diagnosis and management of the underlying disease. Even in some seemingly more localized lesions like neuralgic amyotrophy (paralytic brachial neuritis) conduction studies and electromyography may demonstrate more widespread affection of peripheral nerves. Some practical aspects of conduction studies will be discussed.

AN UNUSUAL CASE OF ACUTE INFLAMMATORY POLYNEUROPATHY — THE ROLE OF PREGNANCY, RHESUS IMMUNE GLOBULIN, AND PLASMAPHERESIS

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A 27-year-old G4P3 female presented at 38 weeks of gestation with a one month history of a rapidly evolving sensory-motor disturbance of the upper and lower limbs consistent with a diagnosis of acute inflammatory polyneuropathy. This was supported by evidence of an *increased CSF protein (97%), nerve conduction studies and a* sural nerve biopsy.

Four weeks prior to the development of the neuropathy, the patient had received 300 mg of rhesus immune globulin (Rho(D) Immune Globulin — Connaught). A small group of patients is known to develop this disorder two to four weeks following administration of a vaccine. The association following the administration of anti-D globulin has not been previously reported.

The association with pregnancy is believed to be coincidental and in a few previously reported cases, delivery has appeared to have a beneficial effect on the course of the disease. However, in our patient, within 48 hours of delivery (7 weeks following onset) a rapid deterioration in neurologic function occurred with the development of profound weakness of extremity function, facial diplegia, and bulbar palsy. The CSF protein became 226 mg%.

The patient was treated with steroids (prednisone 80 mg per day). Ten days following the acute deterioration, she was treated with four 3 L exchange plasmapheresis. This latter treatment was associated with subjective and objective improvement. Following the fourth exchange she had recovered close to her pre-delivery status. Her subsequent course has been that of continuing recovery.

DERMATOGLYPHIC ALTERATIONS ARE A SUBTLE INDICATOR OF ANTICONVULSANT EFFECT ON THE FETUS

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Gestational exposure to anticonvulsant medication has been associated with an increased risk of major congenital malformations, dysmorphic craniofacial features, and digital defects. A clinical investigation of children of epileptic women was designed to assess the risk of minor congenital abnormalities in relation to the presence of major congenital malformations and maternal use of anticonvulsant medication during pregnancy.

To date, 72 children ascertained retrospectively and prospectively have been examined, of whom 50 were exposed to anticonvulsant medication in utero. Preliminary results showed a high frequency of hypoplasia of the distal phalanges with or without nail hypoplasia only in children of treated epileptic women (22.0%). These digital abnormalities, diagnosed on clinical and/or radiographic grounds, were variable in severity. Dermatoglyphic analyses indicated a significantly higher frequency of digital arches in children of the medication group (18.7 vs 2.7%). Although the higher frequency of digital arches was associated both with normal and abnormal digits, it was particularly evident on hypoplastic phalanges (34.5%).

These results suggest that dermal ridge patterning, which is laid down between the 13th and 16th week of gestation, may serve as a sensitive indicator of drug induced dysmorphogenesis early in development.

LIFE THREATENING COMPLICATIONS FROM TRADITIONAL ANTICONVULSANT DRUGS

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Among 1000 consecutive children with newly diagnosed seizure disorders seen at the I.W. Killiam Hospital for Children during the past 4 years, four developed life-threatening complications from anticonvulsant drugs. Phenobarbital was the first drug used in all and a rash was the initial adverse reaction. The drug was continued for 3-10 days after the rash appeared. Three subsequently received diphenylhydantoin (DPH) or carbamazepine (CBZ) before the rash subsided. Hepatitis developed in all four and other complications included Stevens-Johnson syndrome and a lupoid syndrome. All four survived although one suffered a cardiac arrest. One patient had reactivation of lupus, when later reexposed to CBZ, and another developed bone marrow suppression when subsequently exposed to CBZ for the first time. These patients illustrate cross reactivity of phenobarbital with DPH or CBZ, although a "priming" effect of phenobarbital or an interaction between the drug and an infectious agent are other possible mechanisms. Subsequently Valproic Acid has been used uneventfully in two of these patients and Clonazepam in one. We suggest that these newer drugs may be less likely to produce cross sensitivity.

Life-threatening reactions to traditional anticonvulsant drugs may be more common than is appreciated. Hypersensitivity to one anticonvulsant drug should warn of the risk of adverse reaction to the second.

SURGICAL AND ELECTRONIC INSTRUMENTATION FOR THE FUNCTIONAL EXPLORATION OF EPILEPSY

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Surgical treatment of intractable seizures is continuously improving. Recording of spontaneous seizures either from the scalp or from the depth with chronic depth electrodes has allowed for a better definition of the epileptogenic zone. The success of surgery depends directly on the precise localisation of this zone.

In order to achieve this goal, a new stereotactic frame was developed to introduce precisely and safely many small flexible electrodes having from 5 to 15 contacts. An attachment for this frame was also built to introduce electrodes in direction not necessarily in the AP or lateral projections.

It is desirable to pick up all the available data (up to 130 separate signals) and to shorten the duration of monitoring.

A 32 chanel prototype was designed and built recording the electrical signals on the videotape which also record the behavior of the patient.

The availability of the SEEG signals on a videotape will allow repeated analysis using different strategies.

We will be demonstrating surgical instruments and technic and operate the electronic equipment.

CLINICAL SEIZURE FORMS IN TEMPORAL LOBE EPILEPSY

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The clinical seizure forms in thirty-three (33) children who had a temporal lobectomy for intractable seizures were analysed. Neocortical seizures occurred in ten (10), allocortical in twenty (20), focal motor in seventeen (17), tonic adversive in thirteen (13), automatisms in thirty-one (31) and generalized tonic clonic in twenty-five (25).

The results of surgery and the pathology of the temporal lobes varied with the different seizure types. The relationship between the different types of neocortical seizures and the verbal and performance intelligence quotients will be discussed.

THE ANATOMY OF JARGON

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Neologistic jargonaphasia is a distinct entity characterized by fluent speech under pressure, poor comprehension and copious neologistic paraphasias. It is often associated with anosognosia for the speech defect, apraxia, alexia and graphorrhea. From a larger population of 52 Wernicke's aphasics, we chose 10 with jargonaphasia and localizing information on the isotope scan, CT scan or autopsy. The lesions were traced without the knowledge of clinical features. The lesions were overlapped and the anatomy of this unique language disturbance is analysed. Some lesions involve the supramarginal gyrus predominantly, others are more temporal but it is concluded from the overlap technique and the autopsied case that both superior posterior temporal lobe and supramarginal gyrus has to be involved to produce this unique clinical disturbance. Whole brain sections of an autopsied case gave information beyond the in vivo localization techniques. Even though much of the lesion was in the supramarginal gyrus, inferior parietal lobule region, the temporal operculum and the planum temporal were also involved.

A model of neural processing of language is constructed on the basis of the anatomical evidence to elucidate the neurolinguistic and behavioural aspects of jargonaphasia.

BRAINSTEM AUDITORY EVOKED POTENTIAL (BAEP) — THE IMPORTANCE OF NOISE

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In the clinical use of BAEP, one is commonly faced with abnormal waveforms which change character on repeating the test. Since many artifacts ("noise") can distort BAEP morphology, it is important to guard against making a false positive diagnosis on this basis. This requires knowledge of the noise content.

A simple method is described which allows on-line statistical estimation of the signal noise ratio (SNR), and objective determination of the presence or absence of significant peak compo-

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nents. It is shown that high SNR is associated with "clean" and highly reproducible BAEPs, and low SNR with poorly reproducible ones. The SNR thus provides an instant objective ability to judge the acceptability of single BAEP, or whether further averaging is required. The effect of noise is shown to be quite marked, and can cause gross distortion of peak amplitude, latency and shape.

The technique is useful in establishing sensory threshold in evoked response audiometry, and can be applied directly to the analysis of other kinds of evoked potentials.

THE TREATMENT OF PROGRESSIVE MULTIFOCAL LEUCOENCEPHALOPATHY WITH ANTI-DNA VIRUS AGENTS: A CASE REPORT

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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 papova virus infection of the central nervous system, is available. Another DNA virus infection, namely herpes simplex encephalitis, will respond to adenosine arabinoside (Ara-A) therapy and acyclic quanosine shows promise. Cytosine arabinoside (Ara-C) and 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 5 ½ year history of Hodgkin's disease (Stage III B) and for which he had received previous x-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 oro-pharyngeal 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 brain biopsy tissue.

On each of nine days he received intravenously 30 mgm/Kgm of Ara-A. There were no significant side effects. Because of continuing deterioration of his neurological status and beginning 16 days after the completion of his ARA-A therapy he received intravenously on each of six days 105 mgm of Ara-C plus 15 mgm of AraC intrathecally via lumbar puncture on the 2nd, 3rd and 4th days. Again there were no significant side effects but his neurologic status continues to deteriorate to the present (3 weeks after completion of Ara-C therapy).

ABERRANT WAVE FORMS TO PATTERNS REVERSAL STIMULATION: CLINICAL SIGNIFICANCE AND ELECTROGRAPHIC "SOLUTIONS"

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Although a delay in appearance of the major electropositive potential (P2) evoked by pattern stimulation correlates reliably with anterior visual system lesions such as optic neuritis, the test becomes clinically valueless when more than one potential can be plausibly identified as (P2). This phenomenon, which we have termed an aberrant wave form (AWF) occurred in 20% of our patient population.

AWFs did not appear in 34 normal controls. 40% of patients with AWFs for at least one eye had definite MS, an additional 28% had questionable MS, while the remainder had chronic myelopathy or other central nervous system disease. These findings did not depend on whether the possible (P2) peaks were delayed or not. Similar percentages occurred among patients in whom the (P2) peak for at least one eye was clearly defined but delayed. In contrast, among patients with a normal PVER response in each eye, MS occurred in 10% and questionable MS in an additional 33%.

Using field potential characteristics of clearly defined (P2)s as a guide, multiple channel recording of full-field pattern evoked potentials identified the "true" (P2) peak in 55% of AWFs and that of hemi-field stimuli in 70%.

We conclude that an aberrant wave form as above defined carries a similar clinical significance as a normally formed but delayed (P2) peak. Hemi-field stimuli is the best method of discerning which of several peaks evoked by full-field stimuli is the "true" (P2).

SOME EMG OBSERVATIONS ON CROSSED REFLEX ACTIVITY IN HUMANS

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In 1898 Sherrington demonstrated in the experimental animal that deep reflexes spread across the midline through a pathway extrinsic to the spinal cord. This mechanism is different from the intraspinal pathway which has been postulated for superficial reflexes. Since these reflexes are also encountered in humans, this study was undertaken to determine whether similar mechanisms might exist in man. Patients were selected who had brisk crossed adductor reflexes and others who had bilateral contractions of abdominal muscles when the deep and superficial abdominal reflexes were elicited. These responses were recorded electromyographically. The amplitude of the action potentials provided information as to the mechanism of crossing for adductor and superficial abdominal reflexes. Latency determinations were of value in defining the pathways utilized for the deep and superficial abdominal reflexes. From these EMG observations in man, the crossed adductor reflex is mediated by a pathway extrinsic to spinal cord. A similar manner of crossing is postulated for the deep abdominal reflex while the superficial abdominal reflex utilizes an intraspinal pathway. These patterns of crossed reflex activity in humans are similar to those which have been demonstrated in the experimental animal.

ELECTROGRAPHIC AND CLINICAL CORRELATES OF SECONDARY BILATERAL SYNCHRONY (SBS)

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

Defined as generalised epileptiform discharges (GEPs) consistently preceded by focal spikes or sharp waves, SBS occurred in 57 patients.

56 cases (98%) had more than one spike focus: 35 (61%) showed three or more non-contiguous foci in the inter-paroxysmal period. The most active spike focus triggered the GEPs in all cases. 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 ipsilateral to the focus of onset.

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%).

Grand mal seizures occurred in 46 patients (80%) and focal seizures occurred in 29 (50%). The focal seizures clinically implicated the frontal lobe in 13 patients and the temporal lobe in 10.

32 patients were mentally subnormal. Focal neurological signs which occurred in 19 patients (33%) correlated with the site of SBS origin in each instance.

MULTIMODALITY EVOKED POTENTIAL TESTING IN MULTIPLE SCLEROSIS

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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 33 were classified as clinically definite, 21 as probable, 20 as suspected and 12 as progressive Multiple Sclerosis (MacDonald-Halliday 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). It was found that 91% in the definite, 81% in the probable, 55% in the suspected and 42% in the progressive diagnostic groups had significant evoked responses abnormalities in at least one of the three systems. 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 BAEP in 35% and one or more responses abnormal in 78% of the total of these patients. In the 33 patients outside the diagnostic classifications there was an 18% abnormal rate and these abnormalities and the clinical situation in which they occurred will be considered.

The addition of the BAEPs and the SEPs to the PVEP does increase the yield of diagnostically useful information from sensory evoked potentials in M.S., but the PVEP is still the single most sensitive indicator of subclinical involvement, with the SEP slightly less effective and the BAEP the least sensitive indicator of disseminated demyelination

SEROTONIN METABOLISM IN EPILEPTIC PATIENTS

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Modifications of biogenic amine levels in CNS alter the seizure threshold in animal models and thus may play a role in the pathophysiology of human epilepsy. We measured the concentration of the serotonin metabolite 5-hydroxy-indoleacetic acid (5HIAA) in CSF of patients un-dergoing diagnostic pneumoencephalography (PEG). The first and last samples drawn originate from lumbar and cisternal compartments respectively. A "U" shaped age dependent curve was found, women had higher levels, and CSF tryptophan concentrations correlated with 5HIAA levels in both compartments. No difference was found between non epileptic and untreated epileptic patients, but there was a significant reduction of 5HIAA in the lumbar sample of treated epileptics. Tryptophan oral loading 8 to 12 hours prior to PEG increased 5HIAA in both compartments. A load of 3g increased 5HIAA as much as 6g, but with the larger load the rise of 5HIAA lasted longer. No seizures resulted from such loads. In conclusion, central serotonin turnover as reflected by CSF 5HIAA concentration is affected by age, sex and precursor availability, and does not appear to be reduced in untreated epileptics. 3g of oral tryptophan three times a day appear to be a safe way to provide a serotonergic stimulation in adult epileptics and could be tested as an anticonvulsive treatment.

SPINAL SUBDURAL HEMATOMA

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Intracranial subdural hematoma is well recognized and fully documented, but the occurrence of a similar lesion within the spinal canal is rare. Our recent experience with 2 cases prompted a review of this condition.

46 cases are reviewed. There were 26 female and 20 male patients, ranging in age from 6 months to 80 years (average 47 years). Percipitating factors included blood dyscrasia and anti-coagulants (with and without lumbar puncture), lumbar puncture alone, and both minor and major trauma. 4 cases were considered to be spontaneous.

The lesions were situated primarily in the mid to lower thoracic region of the spinal canal. The duration of symptoms ranged from a few hours to several years. Myelography showed either a complete or partial block. 9 patients required myelogram via cysternal puncture because the clot mass prevented an adequate study via the lumbar route. 34 cases were treated surgically. The operative findings and results are discussed.

The clinical presentation of this lesion is relatively uniform, particularly in the presence of bleeding disorder or anti-coagulant therapy, but the diagnosis is often delayed to the point of permanent spinal cord injury.

MONITORING DURING CAROTID ENDARTECTOMY: FURTHER EVIDENCE THAT AN INTERNAL SHUNT IS NOT NECESSARY

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We believe that most, if not all, neurological deficits arising as a consequence of carotid endartectomy are the result of emboli during the surgery rather than hemodynamic intolerance to carotid cross-clamping. This conviction is based on observations

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in 105 consecutive cases. An internal shunt was used only three times. Three of the 105 patients had a new deficit post-operatively. In every instance the deficit was transient.

In 64 cases continuous 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 was there an EEG change. In 18 patients measurements of cerebral blood flow (CBF) and internal carotid 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 pressure paralleled the changes in CBF.

Our results demonstrate that major EEG changes, and profound reductions in CBF (hemispheric flow as low as 14 ml/100 gm/min in one case) may occur during carotid endartectomy without any new post-operative deficit occurring. These observations support our view that an internal shunt is rarely, if ever, necessary during carotid endartectomy.

RETINAL NERVE FIBRE ATROPHY IN COMPRESSION OF THE CHIASM A PROGNOSTIC SIGN

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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 funduscopic signs of diffuse or hemioptic retinal NFL atrophy before and after treatment of compressive lesions of the optic chiasm. Redfree fundus copy and retinal photography were performed in ten patients who presented with visual field defects documented by kinetic Goldman perimetry and tangent screen examination. Serial visual fields and assessment of descending atrophy of the NFL were correlated over follow-up periods of one to five years (mean 2.2 years) after surgery.

Degeneration of the nasal hemioptic NFL was present in seven of twenty eyes. Seven eyes showed diffuse attrition of optic axons on the retina. Six eyes showed no NFL atrophy. Field loss persisted in eyes with hemioptic 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 attrition 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

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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 to yield a smooth eye movement velocity command. In the absence of retinal input, eye position information is derived from outflow, the corollary discharge of eye position commands, and perhaps from orbital proprioceptive inflow. We investigated voluntary control of the VOR in ten normal subjects and three patients after cerebral hemidecortication. During sinusoidal whole body rotation in darkness, subjects were required to enhance the VOR by fixating an imaginary stationary target and to suppress it by fixating an imaginary target moving with the head.

Normal subjects generated smooth eye movements toward imaginary stationary or moving percepts by enhancing or suppressing the VOR, at will. Patients could not enhance the VOR ipsilateral to the decorticate hemisphere or suppress the VOR contralateral to the decorticate hemisphere. We infer that the cerebrum uses a corollary discharge of eye position and a vestibular head position signal to compute gaze position and direct smooth eye movement to a desired orbital position. A cybernetic model is presented. One cerebral hemisphere regulates the ipsilateral smooth eye movements that mediate voluntary control of the VOR.

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

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In vitro contracture tests on skeletal muscle are recognized as the most sensitive methods for the detection of individuals at risk for malignant hyperthermia (MH). Several factors are known to influence the outcome of these tests. These include the temperature at which the experiment is carried out and recent drug ingestion by the subject prior to the biopsy. This report describes the effect of the histochemical profile of muscle on the in vitro caffeine contracture test (with and without halothane).

Male white rats (University of Calgary strain) were sacrificed by decapitation. Soleus (mainly type I fibers) and extensor digitorum longus (mainly type II fibers) muscles were removed and studied in the standard experimental set-up.

Dose response curves for soleus were consistently to the left of those for extensor digitorum longus. Threshold concentrations for soleus were also less than those for extensor digitorum longus.

These findings indicate that, in the rat, the interpretation of the in vitro caffeine contracture test results is influenced by the histochemical profile of the specimen being tested.

A prospective study is being undertaken, in our laboratory, to ascertain the influence of the histochemical profile of the biopsy specimen on the results of in vitro caffeine contracture studies in patients at risk for MH.

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

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Problem based learning requires the student to learn while attempting to evaluate and understand neurological patient problems. The student is taught to apply the hypothetico-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 through 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 work with clinical problems, 4) reinforcement of learning through reuse in subsequent problems, 5) perceived relevance of learning, 6) high motivation for learning through active participation in the problem-solving process, 7) learning is individualized to the educational needs of each student and, 8) acquisition of self-educational skills in neurology for life long learning.

This paper will describe; 1) criteria for problem selection, 2) design of problem formats, 3) structure of problem based learning and, 4) faculty skills required.

An evaluation of; 1) student acceptance, 2) teacher acceptance and, 3) student learning relevant to the advantages described in a recurrent six week block in neuroscience, a subunit of Phase III McMaster's curriculum will be presented.

CLINICAL VALUE OF THE CORNEOMANDIBULAR REFLEX

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Although the corneomandibular reflex has been recognized since 1902, its significance remains unknown. The reflex, consisting of a contralateral jaw deviation (pterygoid contraction) following a brisk corneal stimulation, was found in 40 cases out of several hundred representative neurological patients examined over a five-year period.

The reflex was unilateral in 14/40, bilateral but asymmetrical in 11/40 and bilaterally symmetrical in 15/40. Twenty-eight of 40 patients were stuporous or comatose.

The reflex was seen in 3 patients without evidence of neurological disease; in 1 patient with motor neuron disease in 16 patients with unilateral hemispheric lesions where it was usually contralateral to the lesion and often associated with elevated intracranial pressure (13/16); in 2 patients with acute bilateral hemispheric lesions; in 7 patients with intrinsic upper brainstem lesions and in 11 cases with diffuse (usually anoxic) or metabolic brain disease.

In the comatose or stuporous patient the corneomandibular reflex tended to suggest an intrinsic brainstem lesion or a hemispheric lesion with elevated intracranial pressure and secondary brainstem distortion. It was also useful for following the evolution of the depth of coma. In the alert patient the reflex correlated best with recent or remote interruption of supranuclear pathways to the trigeminal nerve nuclei.

POST-OPERATIVE SYMPTOMS IN UNRECOGNIZED MALIGNANT HYPERTHERMIA (M.H.) REACTIONS

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Patients who survive documented intra-operative M H reactions often complain of muscle pains and weakness. We are reporting 16 patients who developed muscular symptoms, low-grade fever or fatigue after general (13) and or local (6) anesthesia. In no instance was the diagnosis suspected, at the time of surgery, or when the symptoms first appeared. When operative records were reviewed no abnormality was seen. One woman presented with a 6 months history of low-grade fever and malaise starting 36 hours after repair of a scaphoid fracture. Another's leg cramps began 17 years earlier, as an episode of "phlebitis" 8 days after epidural anesthesia. Thirteen of 14 patients with muscular pains, cramps or weakness reported either initiation or exacerbation of their symptoms with surgery. Nine of these had elevated serum CPK. In all patients the diagnosis was confirmed at muscle biopsy by the caffeine contracture test of Kalow et al (1970). In 6 families other biopsy positive individuals were discovered. In 3 other families anesthetic reactions were clearly present by history.

Malignant hyperthermia should be suspected in all patients with unexplained muscular symptoms, elevation of serum CPK or persistent fevers. Careful anesthetic histories of probands and family should be obtained and consideration given to diagnostic muscle biopsy.

THE RATIONALE AND PROGRESS OF THE COLLABORATIVE EC/IC BYPASS STUDY

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RATIONALE

Many surgeons are capable of performing superficial termporal to middle cerebral artery microvascular anastomosis. Morbidity can be kept to a minium and mortality is low. Patency rates exceed 90% in experienced hands. Anecdotal evidence encourages the possibility of stroke prevention by this technique. Suggested indications are internal carotid artery thrombosis, surgically inaccessible (high cervical and intracranial) internal carotid stenosis, middle cerebral artery stenosis and occlusion. The natural history in patients with these conditions and evidencing TIA or partial stroke is not known in more than a fragmentary way. From the slender evidence available, two to ten percent of carotid occlusions will have stroke each year after the occlusion, mostly embolic from alternate carotid channels, some from carotid stumps. Data is totally lacking about prognosis for inaccessible carotid artery stenosis and middle cerebral occlusion. Middle cerebral stenosis by best available figures has a 4% per year risk of stroke. These imperfect figures cannot be used to control any evaluation of the bypass.

PROGRESS OF STUDY

From 58 centres patients with TIA and partial stroke and having the radiological lesions mentioned have been submitted by random selection to best known medical therapy or to the same plus the anastomosis. Sample size calculations indicate a requirement of 1,000 cases followed for an average of five years if a 50% stroke rate reduction is to be achieved. 620 cases have been randomized to date. From North American and European centres the ratio of internal carotid to middle cerebral cases is 3:1, from Japan 3:7. Patency rate in post-operative angiograms is 92%. Risk factor balance to date indicates an effective randomization process.

Data respecting the longterm patency of the bypass is accumulating from pathological and angiographic data. Examples of narrowing of the lumen of the supervicial temporal artery on late follow-up have been collected and will be shown.

Dementia, neurological disability and the use of the procedure as adjunct to aneurysm and basal tumor surgery are not being tested in this trial.

A NEW INHERITED CANINE HYPOMYELINATING DISORDER ASSOCIATED WITH CONGENITAL TREMOR

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An inherited canine disorder, characterised by the development of a whole 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 weeks and the central and peripheral nervous systems sampled. 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 internodes were short, thin and often found on axons intercalated between naked segments. On EM, large axons were either unensheated and apposed, or thinly myelinated; there was no evidence of myelin breakdown. There were many astrocytic processes, some of which ensheated axons or ran between the axon and its myelin sheath. Freeze fracture EM showed that some paranodal specializations had formed in the absence of myelin lamellae and without the normal membrane specializations. In conclusion, these abnormalities of myelination provide a morphological basis for this inherited tremor syndrome. The possibility that these morphological abnormalities are due to aberrant axon-glial interactions or to deficient glial cell multiplication or maturation will be discussed.

CEREBRAL PHOSPHOLIPIDOSIS EXPERIMENTALLY-INDUCED WITH CHLORPHENTERMINE

J.R. WHERRETT

S. HUTERER, M. KHAN AND N.B. REWCASTLE
University of Toronto

An increasing number of neurotropic drugs have been found to cause a systemic lipidosis in animals morphologically similar to the Niemann-Pick syndromes. These experimental phospholipidoses could provide convenient models in which to analyze pathophysiological disturbances in cerebral storage disorders. Here we describe morphological and lipid changes in brain and other tissues of rats who had received the anorectic drug chlorphentermine, (50 mg per kg intraperitoneally, 5 times weekly for 3 weeks). Electron microscopy confirmed the formation of intracellular lipid cytosomes in lung and liver and in cerebral neurones after exposure to the drug. Chemical analysis of lung, liver, spleen and brain revealed variable alterations in organ weight, lipid and phospholipid content. In all tissues there was an increase in the lysosome-specific phospholipid, bis-(monoacylglycero)-phosphate ranging from 4-fold in brain to 14-fold in lung. This phospholipid is thought to play an important role in the lysosomal metabolism of fatty acids and is known to accumulate in human Niemann-Pick syndromes. In this study we have shown that neuronal lipidosis may be induced readily in vivo by a neurotropic drug and is accompanied by a disturbance in glycerophosphatide metabolism.

INTERNATIONAL NEUROSURGICAL EDUCATION A PERSONAL PERSPECTIVE

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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 the 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 man power saturation and government awareness of this has resulted in immigration policies which will make it difficult or prohibititive for perspective candidates in medical education. Consquently, we need to take a fresh look at involvement in International Education and development.

Chang Gung Memorial Hospital is a new well equipped institution with a growing clinical volume. The average monthly load at present is 40 - 55, major procedures including trauma (20 - 30), tumors, (12 - 18), cerebral hemorrhage (4 - 10), and aneurysm (2 - 3). The pathology is varied and usually advanced. The attending staff includes three neurologists and three neurosurgeons. Neurology and Neurosurgery share about 100 ward beds. There are good, well equipped, computerized ICU's and 9 bed nursing observation unit.

Operation room facilities are as advanced as most in Canada. The diagnostic facilities include polytomography, biplane stereo angiography and CT Scanning.

The primary areas of teaching involvement are with staff surgeons who have recieved one or two years training locally in Neurosurgery and residents. Nursing care requires considerable up grading.

In the presentation, I would attempt to outline the needs and problems of International Medical Education, suggest a new way to meet the needs and relate our personal experience in the developing world.

DOWN'S SYNDROME IN FAMILIES WITH ALZHEIMER'S DISEASE

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Neuropathologic changes identical to those seen in Alzheimer's Disease develop in almost all patients with Down's Syndrome after the third decade of age. Clinical manifestations of dementia however, occur less often. Down's Syndrome occurs more frequently than expected in families of patients with Alzheimer's Disease.

As part of the Collaborative Case-Control Study of Alzheimer's Disease we have found three such families which are the subject of this report. In family one, the index case was a 57 year old woman with Alzheimer's Disease, the seventh of eight children. That sibship included two sisters with Down's Syndrome. One died at the age of 4 ½ years but the surviving affected sister developed dementia at the age of 51 years. These siblings have on first cousin, two second cousins and one grandniece with Down's Syndrome. Thus, within three generations, five persons have Down's Syndrome alone, another has Alzheimer's Disease alone, and one has both conditions. This family had a 15/21 translocation but the other two families had trisomy 21.

Our observations suggest that Down's Syndrome and Alzheimer's Disease may have a common genetic defect, and underline the importance of obtaining a detailed family history and karyotyping in patients with pre-senile dementia.

POST-OPERATIVE TENSION PNEUMOCEPHALUS

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Recent reports suggest the association of surgery performed in the sitting position using nitrous oxide anaesthesia with the accumulation of subdural air under tension causing a deterioration in level of consciousness.

We report two such patients who worsened in the early post-operative period.

The presence of air was demonstrated using computerized tomography and skull films. Aspiration of the air resulted in marked sustained improvement in their conditions.

We believe that tension pneumocephalus should be considered in the differential diagnosis of any patient whose conscious level fails to improve post-operatively.

DELAYED METRIZAMIDE CT IN SYRINGOMYELIA

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H.J.M. BARNETT AND G. DEBRUN
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The theory of Gardner relates to the development of syringomyelic cavities by abnormal CSF pulsation pressure transmission between the fourth ventricle and the central canal of the spinal cord. This explanation is not adequate for all categories of syringomyelia, especially the non-communicating varities (e.g. spinal arachnoiditis and post-traumatic). Alternative theories include abnormal CSF passage through the cord substance via Virchow-Robins spaces or as a result of production and absorption of CSF by glial cells lining the cavities. While most neuroradiological assessment has been anatomic, i.e. evaluation of cord enlargement 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 syrinx and the surrounding subarachnoid space. This also may be the most definitive way of confirming the presence and extent of the syrinx.

PROTECTIVE EFFECT OF FLUOSOL IN ACUTE CEREBRAL ISCHEMIA

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Perfluorochemical Blood Substitutes (Fluosol-DA) are small particle fluorocarbons (0.2 micron) suspended in an emulsion and with a high propensity for oxygen and carbon dioxide. We have studied Fluosol-DA as compared to Mannitol in the modification of acute focal cerebral ischemia. Adult mongrel cats were divided into three groups of 12 animals each. All animals underwent a transorbital tourniquet ligation of the middle cerebral artery. Control animals were treated with intravenous 0.9% saline (15 ml/Kg). Animals in the experimental groups were given either Fluosol-DA 35% (15 ml/Kg) or Mannitol 20% (1.2 grams/Kg). All animals were treated in an oxygen chamber and four cats from each group were sacrificed at 1, 3 and 6 hours after middle cerebral artery occlusion. The brains were perfused with a mixture of trypan blue and buffered formaldehyde and light microscopic analysis of ischemic cortical neuronal change was assessed and graded for each animal. Control animals demonstrated an average percentile infarction of 57% whereas Mannitol treated animals had a 29% area of infarction and the Fluosol animals 27%. Moreover, those animals treated with Fluosol showed a marked decrease in the severity of neuronal change. It is suggested that Fluosol, because of its low viscosity, may support the microcirculation and maintain cortical collateral flow as well as supplying oxygen to the ischemic area. The implications in the treatment of ischemic stroke will be discussed.

PLASMA AMINO ACIDS IN EPILEPSY

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McGill University

N.M. VAN GELDER
University of Montreal

K. METRAKOS
The Montreal Children's Hospital

We have previously reported plasma amino acids in patients with 3/sec spike-wave epilepsy. In the present study plasma amino acid investigations were made in a heterogeneous group 11 epileptics patients whose electroenphalograms exhibited cerebral dysrhythmia other than the classical 3/sec spike-wave abnormality. 11 first degree relatives of these epileptic patients were also investigated. Plasma levels of 14 amino acids were determined and ratios of certain metabolically related amino acids calculated. In the epileptic probands when compared with

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22 control probands the mean plasma levels of ASP, THR, SER, ALA, ILE, LEU and TYR were significantly decreased along with a significant decrease in the ratios of TAU/GLU, GLN/GLU, THR/SER, 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, ALA, 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 that with 3/sec spike-wave elilepsy, the former showing a significant decrease of THR, SER, GLY, ALA, MET, ILE, LEU, TYR, THR/GLY and SER/GLY Discriminant Analysis it was possible to distinguish epileptics from controls with 100% accuracy and from 3/sec spike-wave epileptics with 90.5% accuracy.

The results of this indicate; 1) that epileptic probands whose electro-encephalograms show focal, abnormal or diffuse type of patterns, show plasma amino acid patterns which are significantly different from non-epileptic control probands, 2) that these altered 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

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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 artery (MCA) of 18 ketamine-anesthetized cat was exposed microsurgically. Plasma and erythrocyte flow in the right Sylvian cortex was determined by measuring the transit of ^{131}I albumin and ^{99}Tc-labeled erythrocytes injected into the ipsilateral carotic artery. The right MCA was occluded and the flow studies were repeated. Groups of 6 cats each had flow studies after 1 hour, 3 hours, or 6 hours of occlusion. The cats were then killed by the intra-aortic perfusion of a colloidal carbon-buffered fixative solution. Sodium fluorescein (50 mg) and Evans blue (50 mg) were given intravenously 20 minutes before death. Mean transit times before occlusion were 6.0 ± 0.5 sec for ^{131}I albumin and 11.5 ± 0.5 sec for ^{99}Tc-labeled erythrocytes. They increased slightly after occlusion to 9.5 ± 1.0 sec for ^{131}I albumin and 15.5 ± 1.0 *sec for* ^{99}Tc *erythrocytes. Progressive delay in* mean transit time was seen with longer periods of ischemia. At 6 hours, mean transit time was 22.5 ± 3.5 sec ^{131}I albumin and 24.0 ± 4.0 sec for ^{99}Tc erythrocytes. Prolongation of transit times correlated with the development of cortical edema and microvascular narrowing. Three cats with evidence of caudate nucleus ischemia only had little change in their flow patterns. The findings of this study suggest that plasma and erythrocyte flow changes are similar and that a state of plasmapheresis does not develop.

"THE VALUE OF SCANNING ELECTRON MICROSCOPY IN EXPERIMENTAL MICROVASCULAR SURGERY"

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Experimental microvascular surgery has proven to play an important role in clinical research for neurosurgery: firstly, for teaching purposes in performing micro-anastomoses on vessels measuring 1mm in diameter or less (extra-intracranial bypass operations for treatment of cerebral vascular insufficiency) and secondly for the basic knowledge of the long-term evolution of operated microvessels. Scanning electron microscopy (S.E.M.) is a highly valuable method for studying reparative processes of the vascular endothelium following surgical manipulations.

More than 150 microvascular interventions have been performed in rabbits and rats: end-to-end anastomoses, end-to-side anastomoses, longitudinal sutures and venous patches. The sequential modifications of the endothelial surfaces have been scrutinized using S.E.M. following various surgical techniques and suture materials.

The patency rate of these anastomoses has been slightly above 90%. The surgical trauma upon the intima has been particularly studied and the lacerations of needles and sutures have been determined with accuracy on "empty specimens". Intimal lacerations have also been observed following the technique of continuous suturing as compared to the conventional technique with interrupted stitches. The intimal trauma following applications of microvascular clips for temporary occlusion has also been investigated: the low-pressure ACLAND clip has proven to be less traumatic for the endothelium than any others.

The process of endothelial reconstruction can be followed most accurately with the S.E.M.: there is early elongation of endothelial cells which tend to fill out progressively the surgical gaps and to cover the suture material in consecutive layers.

The observation of the endothelial regeneration will contribute to develop less traumatic surgical techniques and suture materials in order to lessen the myo-intimal thickening observed after any microvascular operation and hence improve the long-term permeability.

STABILIZATION OF THE CERVICAL SPINE USING THE ANTERIOR APPROACH

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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 treatment of the unstable 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 classical Cloward interbody fusion utilized include vertical pegs, trapezoid autogenous bone struts 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 presenting 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 ENDARTERECTOMIES

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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 (8mm.) longitudinal incision was made over the lower internal and upper common carotid arteries. The media and the elastic lamina were separated from the calcified plaque with a small, curved instrument. Circumferential dissection was carried out to avoid embolization. It was also possible to carry out a similar separation of the diseased intima from the outer layers of the lower external carotic artery. The microscope 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 EEG monitoring and post-operative clinical evaluations gave no evidence of embolization.

NEUROPSYCHOLOGICAL ASSESSEMENT OF THE EFFECTS OF CAROTID ENDARTERECTOMY

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Carotid endarterectomy has demonstrated prophylactic value, generally resulting in improved neurological status and relief of symptoms. It has been reported in the literature that some patients "feel smarter" after surgery, however these effects have not been adequately researched.

Extensive neuropsychological results were obtained on 55 patients who were examined prior to and 5 months following carotid endarterectomy. Clamping time, size of lesion and history variables were also analyzed. Patients were classified according to side of surgery and whether TIA or completed stroke. A carefully selected surgical group and vascular group were followed in a similar testretest fashion.

Operated and control groups showed improvement on tests due to practice but a significant improvement was noted in patients receiving right carotid endarterectomy with a history of completed stroke. Patients with TIAs seemed to have solely prophylactic effects. The right operative stroke patients improved significantly more than left. Surgery was delayed a significantly longer time for the patients with left-sided stroke and these patients were significantly poorer on cognitive, motor and sensory tests both prior to and following operation. These results raise the possibility that carotid endarterectomies performed quickly on patients with completed strokes will result in a significant restoration of function, whereas only prophylactic benefits accrue if surgery is delayed.

BLOOD FLOW CHARACTERISTICS OF THE EXTRA-CEREBRAL CIRCULATION

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RUDELLI 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. Still, it has been generally assumed that it is a small, homogeneous 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 carotod artery and common carotid artery injections. In a second group (14 patients) selective extra-carotid artery (ECA) injections were performed and the rCBF values were compared in rest and during hyperventilation. In a third group (13 patients) rCBF pattern changes were studied following extracranial intracranial (EC-IC) 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 reaction of the extra-cerebral tissues.

INTERNAL FIXATION OF CERVICAL SPINE DISLOCATIONS WITH AN INTERLAMINER CLAMP.

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AND W.J. HOWES
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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 nerve root deficit, 4 mild spinal contusions and 3 severe cervical cord injuries.

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

Follow-up was 3 to 7 years in 16 cases and 1 ½ to 3 years in 16. Flexion-extension views of the cervical spine 3 months postoperatively showed no evidence of recurrent subluxation. Late follow-up x-rays, when available, showed evidence of fusion of the involved disc space and even of the laminae in some cases. No patient developed infection, delayed neurological deficits, or pain which could be attributed to the presence of the clamp. The advantages of this method will be discussed.

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PERCUTANEOUS LUMBAR RHIZOTOMY FOR SPASMS IN PARAPLEGIA

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Many operations have been recommended to help paraplegics with uncontrollable spasms of the legs. Roots L1 to L5 can be reached percutaneously without entering the subarachnoid space. Thermal lesions readily abolish spasms in the distribution of these roots without risk to the roots serving 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 of balance and diminished sensation have not proved difficult. Two of the first patients treated developed recurrent spasms which did not resond completely to repeated operations. Stimulation of roots 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

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100 consecutive stable (Gr. I-III, Botterell) patients with ruptured berry aneurysms were analysed to compare the results of early and delayed operative repair.

Rebleeds following admission occurred in 28 patients, resulting in 8 deaths, deterioration precluding further surgical consideration(4), and persistent clinical deterioration prior to eventual surgery (7).

Therefore, only 88 patients came to surgery, with only 64 having maintained their admission grade or better. In this group, 31 were repaired within 7 days of their most recent hemorrhage and 33 thereafter.

The outcome following operation is shown:-

Interval from Last hemorrhage	*Excellent & Good*	*Fair & Poor*	*Deaths*
0 - 2 days	11	2	-
3 - 7 days	13	,3	2
> - 7 days	25	5	3

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 3/26 (31%) of cases, repaired 3-7 days following a bleed.

Early repair of ruptured intracranial aneurysms within 48 hrs. of a bleed in clinically stable G. 1-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.

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University of Alberta

J. SUBAK-SHARPE
Glasgow, Scotland

B. YOUNG AND A. ZBITNEAU
Saskatoon

D.W. PATY AND G.D. KETTYLS
University of British Columbia

The DNA of Herpes Simplex Virus can be purified and digested into fragments with restriction endonucleases. The DNA fragments, which radioactively labelled with P^{32}, can then be subjected to agarose gel electrophoresis and autoradiography. The DNA restriction endonuclease profiles obtained readily differentiates Herpes Simplex Virus type I from type II.

We previously reported that 31 spontaneous isolates of herpes simplex virus type I from explants of human trigeminal, superior cervical, 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, whereas multiple virus isolates from the same individual are indistinguishable.

We report here the restriction endonuclease profiles of eight strains of herpes simplex virus which caused encephalitis of humans. The isolates were obtained from London, Ontario, Saskatoon, Saskatchewan, Edmonton, Alberta, and Vancouver, British Columbia, Virus DNA was digested with the restriction endonucleases Bam H1 and KpN1. 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 encephalitis 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 ANTIEPILEPTIC DRUGS

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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 and 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 10 l3g/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 5.0 to 17.9 μg/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

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G.V. WATTERS AND S. SINGER
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Every diabetic child is exposed to the risk of hypoglycemic convulsions. Now with a tighter diabetic control being advocated the occurrence of hypoglycemic episodes and convulsions is going to increase.

The Montreal Children's Hospital diabetic clinic has routinely ordered a baseline EEG on all new diabetics at the time of initial diagnosis. There were one thousand patients in the series, but it was decided to review the last three hundred patients now actively being followed at the Montreal Children's Hospital diabetic unit. This was in an attempt to correlate the initial EEG with future convulsions associated with hypoglycemia.

When the initial EEG of diabetic patients who later develop a seizure disorder was compared to a control group there was a significant difference in the number of epileptiform EEG's seen in the patients with diabetes and seizures (26%) compared with 10.4% of a control group ($P = 0.001$). The differences in the initial epileptiform EEG were not as significant ($P = 0.01$) in the patients with diabetes and seizures (26%) when compared to diabetic patients without seizures (16%). However, the initial epileptiform EEG did not help differentiate those diabetics with recurrent seizures from the diabetics who had only a single clinical episode.

It was concluded that diabetics with an epileptiform abnormality in the initial EEG recording should be followed more closely for the possibility of convulsive episodes with hypoglycemic events.

DIFFERENTIAL DIAGNOSIS OF HYSTERICAL CONVULSIONS FROM EPILEPSY

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22 in-patients with a discharge diagnosis of hysterical 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 diagnoses were made independently by Consultants with a special interest and knowledge of epilepsy after usually a lengthy admission which invariably involved observation of the attacks with post ictal physical examination and EEG's; serial and sleep EEG's and in some sphenoidal EEG's, psychometric tests and specialized radiography. Some had an hysterical attack during an EEG recording.

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

There were nine statistically significant differences on psychiatric 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

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AND A. WILKINS
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A patient with generalized convulsions noted that seizures were reliably precipitated by mental arithmetic. Interictal EEG revealed only a mild diffuse nonspecific disturbance but performance of mental arithmetic 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 discharges. Addition, 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

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We reviewed the records of patients seen from 1976 to 1978 for coma, intracranial infection ($n = 94$) and convulsions ($n = 64$) were the most frequent causes of the coma. In 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 acidosis 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

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From 1971 to 1979 inclusive, the author has carried out 124 temporal lobectomies including the amygdala and the pes of the hippocampus in 124 patients, of which 100 have 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 of 3 seizure per year)	55%
1) totally seizure-free since discharge	34%
2) patients who have become seizure-free (minimum one year)	13%
3) patients still having a maximum of 3 seizures per year	8%
GROUP B (patients having less than 50% of the original number of attacks)	27%
SUCCESS GROUP (Groups A and B)	82%
GROUP C (patients not improved or only slightly improved)	11%
Follow-up incomplete	7%

The results do not seem to differ when operations are performed on the left (57%) or on the right (43%). 10 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 epilepsy cases treated surgically at the Montreal Neurological Institute with emphasis on the preoperative investigation.

NEUROLOGICAL INVOLVEMENT IN CHILDREN WITH MORPHEA (LOCALIZED SCLERODERMA)

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In a retrospective study of 25 children with morphea (mean age at diagnosis was 7 years), neurological abnormalities were found in 65% including psychomotor retardation, partial or generalized seizures, hemiparesis, as well as dyslexia, proximal and distal muscular atrophy, anisocoria and optic atrophy.

Electroencephalographic (EEG) abnormalities were found in 75% of the patients, including paroxysmal or focal abnormalities while a few had epileptiform 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 skin lesions.

Associated abnormalities included hemifacial atrophy, eosinophilia, increased sedimentation rate and hypergammaglobulinemia. 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 high indicating primary or secondary involvement of the nervous system plays a role in the expression of this disorder.

EXTRAPYRAMIDAL COMPLICATIONS OF METOCLOPRAMIDE THERAPY

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Metoclopramide is now a widely prescribed drug. Despite being a striatal 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 metoclopramide-induced extrapyramidal disorders were assessed in a Parkinson's Disease Clinic.

Two patients (average age 54) had developed acute transient 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 remain with some residual signs and may have had pre-existing *undiagnosed Parkinson's Disease*.

Six patients (5 of the parkinsonism group and 1 other) developed tardive dyskinesia on discontinuing metoclopramide (average 2.6 yrs. treatment). This was transient in 3 but oro-buccallingual movements persist at 8 months in the others.

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

HYDROCEPHALUS FOLLOWING BIRTHWEIGHT LESS THAN 1500 g.

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Although very small preterm infants have been reported to frequently develop hydrocephalus following intraventricular hemorrhage (IVH), we have found a very low incidence of hydrocephalus in the Province of Nova Scotia in infants weighing less than 1500 g at birth. Of 341 infants weighing <1500 g born during 1976-78 in Nova Scotia, 314 (92%) were cared for in 3 neonatal intensive care units. Four infants developed hydrocephalus prior to discharge, 2 of whom died with diagnoses of Arnold-Chiari and Dandy Walker cyst, a 3rd died without having cause for hydrocephalus determined, the 4th survived with CT scan-documented postIVH hydrocephalus. 182 (58%) survived to be discharged home from hospital. Of the 132 deaths, 113 (86%) had a complete autopsy; IVH was found in 48 113 (42%). Head circumference at age 11-36 months in 75% of survivors followed was >97%ile in only 2 other infants, both of whom had hydrocephalus excluded by CT scan. Retrospective review of shunt procedures at the only hospital with pediatric neurosurgical services revealed no additional *infants with hydrocephalus developing after discharge*.

Our survival rate comparable to the experience of others and the low incidence of hydrocephalus in our survivors suggest that the incidence of IVH in this population was lower than in other populations reported to date. Although this difference may be due to the unselected nature of this population base, it may be related to the perinatal care which these infants and their mothers received.

RESPONSE OF RECURRENT MEDULLOBLASTOMA TO HIGH DOSE CYCLOPHOSPHAMIDE

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Eight patients with malignant neuroectodermal brain tumors (7—medulloblastoma, 1—pineoblastoma) recurrent after radiation therapy were treated with high intravenous doses of cyclophosphamide. Six of the seven medulloblastoma patients had recurrent posterior fossa disease, one had gross nodular seeding on myelography, five had malignant cells in the CSF and three had systemic spread. The patient with pineoblastoma had a large intradural, extramedullary recurrence at the cervico medullary junction.

The treatment consisted of escalating doses of intravenous cyclophosphamide beginning with a dose of 100mg·kg given as 50mg·kg QD times two days. The dose was raised at monthly intervals by 20mg·kg to a maximum dose of 160mg kg for a maximum of five courses. Alkylating activity was detected in spinal fluid within one hour after intravenous administration when a single intravenous dose exceeded 50mg·kg. Toxicity of this regimen was considerable including profound hematologic depression, sepsis and occasional hemorrhagic cystitis.

Five patients had an unequivocal response (improvement in clinical status and a diagnostic test longer than three months) and three of the seven had a partial response (improvement in either clinical status or a diagnostic test longer than three months).

This aggressive chemotherapy regimen was relatively well tolerated in individuals who had received prior neuraxis radiation. The high response rate observed (100%) suggests that this chemotherapy regimen could be incorporated in a phase III regimen for newly diagnosed disease prior to the completion of neuraxis radiation therapy.

CEREBRAL TUMOURS IN CHILDHOOD PRESENTING AS CHRONIC FOCAL EPILEPSY

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That a longstanding uncontrolled focal seizure disorder may represent a supratentorial cerebral 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 9% tumour incidence of the MNI series covering the same age group (Mathieson, 1975).

In our series, the mean duration 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 examinations. Intelligence was normal in 85% of the tumour patients and 43% of the non-tumour patients.

EEGs showed multiple independent spikes in both hemispheres in 12 of 14 tumour patients. (86%) and in 11 of 15 patients (73%) without tumours. However, persistent delta activity on most sequential EEGs appeared in 71% of tumour patients and only 27% of non-tumour patients.

Initial CTT scans failed to diagnose the tumour in 9 of 14 patients.

CONCLUSIONS: (1) a longstanding therapy resistant focal seizure disorder may be the only clinical manifestation of a childhood tumour, (2) possibility of tumour increases when intelligence and CNS examination are normal, (3) persistent focal EEG delta activity has greater differentiating value than spikes, and (4) initial contrast radiography may be non-diagnostic for tumour.

CAROTID-OPHTHALMIC ANEURYSMS: 19 CASES PRESENTING WITH COMPRESSION OF THE VISUAL APPARATUS ALONE

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A recent review of our experience with 72 cases of carotid-ophthalmic aneurysms reveals that visual involvement is a more important and frequent finding at the time of presentation of these aneurysms than previously thought. Altogether, 25 patients were noted to have visual signs. In six of 30 patients presenting with subarachnoid hemorrhage, visual involvement was a secondary finding. In the other 19 patients, however, the aneurysms were intact and visual deterioration simulating the more common causes of optic nerve and chiasmatic compression was the presenting feature in each case.

These 19 cases were distinctive. All were women. Eighteen of the 19 aneurysms were of "giant" size. Nine patients had *more than one aneurysm. In five, there were bilateral* carotid-opthalmic aneurysms. Visual acuity was impaired in every case, and with the exception of one case, was the symptom bringing the patient to medical attention. In 12 cases, there was bilateral loss, always greatest on the side of a single aneurysm or on the side of the largest of bilateral aneurysms. The visual loss was usually slowly progressive over many months or a few years. Abnormalities of the visual fields were found in all 19 patients. These were remarkably diverse. The most common were bilateral temporal sector defects or hemianopsias; but nasal defects, altitudinal hemianopsias, and central scotomas were also seen.

A wide variety of surgical techniques were required to treat these aneurysms. The overall neurological and visual results of direct treatment (13 cases) was better than the results of indirect treatment. The visual results with direct treatment are somewhat unpredictable. Factors associated with a favourable outcome include a short history, sparing of the ophthalmic artery, and good decompression of the optic apparatus at the time of the initial operation.

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

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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 tactical 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 thalamic epidermoid cyst through the transcallosal approach and a transventricular approach was used for an extensive germinoma of the pineal region. A giant aneurysm of the posterior cerebral artery was successfully clipped through a right subtemporal craniotomy and later excised through a left parietal approach. Intraoperative Doppler recordings were used during the clipping. 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, magnifying loupes and the operating microscope. The postoperative morbidity was negligible and no permanent sequelae were observed but one postoperative death occurred after clipping 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 low mortality and no morbidity. Pineal surgery can be carried out safely and successfully provided that a number of steps are taken in the planning and during the course of the surgical intervention.

CSF DYNAMICS IN ADULTS WITH HYDROCEPHALUS

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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. There were no complications attributable to the test.

In the past, the results of CSF shunting in hydrocephalic patients, selected solely on the basis of the clinical presentation, air study, RISA cisternography 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

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The aggressive treatment of infantile hydrocephalus has been of benefit to many children.

Sophisticated 2-dimensional ultrasound techniques (grey scale, B mode) have made the prenatal diagnosis of hydrocephalus more reliable than previous methods.

Medical decisions regarding the management of hydrocephalus in utero, were formely made by obstetricians, and were directed at reducing maternal morbidity and mortality. Now, however, neurosurgical input is being requested, as more concern for the fate of the fetus is expressed.

Based on our experience with 5 cases of intra-uterine hydrocephalus in the past year, we are proposing guidelines for management of this problem.

If antenatal ultrasonography and roentgenographic studies show only hydrocephalus, we recommend that the fetus be born by elective cesarean section at the time of pulmonary maturity, and early ventricular shunting be carried out. This plan should result in the smallest amount of nervous system damage from the hydrocephalus, according to present knowledge of the pathological effects of hydrocephalus.

However, if antenatal diagnostic studies show cerebral, or other major system anomalies combined with hydrocephalus, then standard obstetrical practice (vaginal delivery of the fetus with cerebrospinal fluid drainage) should prevail.

We have found antenatal ultrasonography to be reliable in assessing fetal lateral ventricular size and shape, and to correlate well with the results of post-natal C.T. scanning.

PARKINSONISM PROVOKED BY ALCOHOLISM

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Six cases of older chronic alcoholics are described wherein Parkinsonism 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 Parkinsonism with maintained abstinence of days to weeks. No patients had clinical or historical evidence of porto-systemic encephalopathy, although 3 patients had mild chemical liver abnormalities. We propose that alcoholism unmasked or augmented pre-existing Parkinsonism. Three cases had prior episodes of transient Parkinsonism 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.

OPTIC NEURITIS (O.N.) IN FAMILIAL MS

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Although the proportion of patients with optic neuritis who do not go on to develop MS is highly variable in reported series, there does appear to be a subgroup of patients with O.N. who on prolonged follow-up do not develop symptoms or signs of disseminated disease (idiopathic O.N.). The relationship of such cases to MS is unclear.

In a study of the families of 80 patients with familial MS, 4 family members with a well documented history of optic neuritis were identified. This had occurred 3, 10, 12, and 25 years prior to their evaluation. Three of these patients underwent visual and auditory evoked responses, electrically and flash evoked blink reflexes, and CSF electrophoresis (isoelectric focusing in polyacrylamide gel). Excepting for the visual evoked response, all studies were normal, tending to confirm the monosymptomatic involvement.

The cumulative appearance of the second symptom in a group of 60 women and 24 men with MS, having O.N. as their first symptoms was plotted. The 4 patients with the period of follow-up noted above were evaluated using this curve. Accordingly, an estimate was made that 50, 75, 80, and 96% of the risk of getting a second lesion in these patients had passed if they were destined to develop disseminated disease. These cases of isolated O.N. in relatives of patients with clinically definite MS suggests a close relationship between MS and idiopathic O.N.

LEUKOCYTE GLUTAMATE DEHYDROGENASE IN VARIOUS FORMS OF ATAXIA

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Many studies from our group have indicated that pyruvate metabolism may be impaired in certain spinocerebellar degenerations (CJNS 3: 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 dehydro-genase (GDH) in a variety of spino cerebellar degenerations. Preliminary results reported elsewhere (A.A.N.-April 1980 meeting) indicated that leukocyte GDH was significantly decreased in 12 FA patient-control pairs ($p < 0.001$). We now report extended results of leukocyte GDH in 44 control subjects and 44 ataxic subjects; all determined as sex and age-matched pairs. The ataxic subjects included 22 classical FA, 9 recessive spastic ataxia of Charlevoix-Saguenay type (CS), 8 dominant olivo-ponto-cerebellar degenerations (OPCA Type I), and 5 recessive ataxias of slow progression without pes cavus and kyphoscoliosis (RA). The only group with a significant leukocyte GDH 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 investigating methods for replacement therapy of glutamic acid.

Studies supported by l'Association Canadienne de l'Ataxie de Friedreich.

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

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Several investigators have reported changes in non-specific suppressor cells (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 response during the acute relapse ($+43.5\%$, $P = 0.01$, $N = 7$) using patients as their own controls. The following ConA generated NSC activity was found in 40 MS patients and 17 normals:

Phase of MS	*Mean % Suppression*	*± SD*	*P Value*
1) Stable MS (N = 22)	65.8	13.72	<0.05
2) Acute Relapse MS (N = 9)	52.4	16.79	<0.02
3) Chronic Prog. MS (N = 6)	79.0	10.88	<0.01
4) Benign MS (N = 3)	32.7	19.30	<0.02
5) Normals (N = 17)	62.1	14.76	

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 relapse. Reduced NSC activity is associated with increased mitogenic response. 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 MYOPATHY WITH COPIOUS LIPOFUSCIN ACCUMULATION IN THE CENTER OF MUSCLE FIBERS

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A 40 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 CPK was elevated less than twofold. Electromyography revealed some single fiber activity and some abnormally 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 myofbril-free zone (up to 35 microns in diameter) which was often filled with copious amounts of glycogen and/or typical lipofuscin. Both fiber types were equally affected by the central nucleation, and its associated abnormalities. There was impressive atrophy of the type I muscle fibers, and hypertrophy of the type II's. 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 central nuclei; this suggested a genetic etiology (probably autosomal dominant) for the disease. The pathological picture in the father's

biopsy differs from that of other forms of centronuclear myopathy, because it shows large clusters of central nuclei, and because of the copious central accumulation of glycogen and lipofuscin. The pathophysiology of these changes is presently unknown.

FACIO SCAPULO HUMERAL DYSTROPHY: THE INFANTILE VARIETY (IFSH)

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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 age 7 years. Outstanding are the peculiar shoulder set and lumbar lordosis. He is intellectually normal. Serum enzymes are markedly elevated and EMG studies show myopathic motor units. The muscle biopsy taken at 5 and 13 year old show end stage muscle with prominent fatty tissue infiltration. The patient's mother showed mild facial, neck and shoulder weakness in line with a low penetrance of the FSH phenotype. Her muscle biopsy showed type I fiber atrophy. This is more typical of myotonic dystrophy (MD) with which this family has little in common clinically. However like in congenital myotonic dystrophy most reported cases of IFSH had an early disabling myopathy most likely inheritated through an autosomal dominant gene from a very mildly affected parent. A major difference seems to be the lack of siblings with an intermediate disability between the two ends of the FSH spectrum unlike (MD). A movie will show the patient at the near end of his ambulatory period 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 EndPlate potentials and (2) other larger voltage negative positive potentials which frequently have pre-potentials 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 phrenic nerve 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 positive 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 positive potentials were clearly triggered by *contact of the concentric needle electrode and the muscle. 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 muscle fibre action potentials, probably pre-synaptically activated by mechanical irritation of the motor axon terminal and pre-terminal branches. The pre-potentials probably represent the End-Plate Potential. This evidence that these negative-positive potentials detected at the End-Plate zone represent muscle fibre action potentials not only clarifies a common misconception in human electromyography, but may lead to important new investigative methods in disorders of neuromuscular transmission.

QUANTITATIVE ANALYSIS *OF MOTOR FUNCTION*

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There is a need to quantitate motor function in clinical neurology in order to better define homogeneous groups of movement disorders and to document changes. The goal of the present investigation was to construct a series of computer-generated and analyzed tasks for the evaluation of fine motor movements. Five motor tasks were programmed using a Declab 11-40 on-line computer and each was presented visually on a Graphics Processor. Motor responses were made by manipulation of a lever which controlled the position of a marker on the screen. The tasks were designed to measure steadiness, initiation time, speed and accuracy of movement. Controls and patients with ataxia were tested.

Each of the motor tasks could be performed by all of the controls (age 7-15 yrs.) and all except the most severely affected patients (age 7-22 yrs.). Practice effects were seen for several of the tests but these effects were small compared to between-group differences. All four patients with Ataxia Telangiectasia had markedly impaired function on all tasks. Speed, accuracy and steadiness were more affected than movement-initiation time. These differences from control were highly significant on analysis of variance. A fifth patient with spinocerebellar degeneration had a different pattern of function. Each patient could serve as their own control in trials of drug therapy if the results before, during and after therapy were compared.

This study demonstrates the feasibility of quantitating motor function. This approach should be useful diagnostically to categorize different types of motor deficit and should prove effective in the documentation of changes in motor function with time and/or with therapy.

PREDICTION OF RESPONSE OF HYPERACTIVE CHILDREN TO METHYLPHENIDATE

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Royal Ottawa Hospital

Follow up studies of hyperactive children have demonstrated persistant 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 longterm investigation of 96 boys age 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 deficit 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 neuropsychologic 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 mth-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 relationships between clinical variables (examination, 2-12 hrs. of the onset of coma) 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; 76% with generalized seizures 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: 88% with normal motor patterns were normal. All those flaccid and arreflexic died.
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. Respiration: 62% with normal respiration and 31% who required assisted ventilation were normal.
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 body temp. died.

Using a stepwise discriminant procedure utilizing clinical variables, 67% cases could be classified correctly in one of the five outcome groups; 90% were correctly classified using two outcome groups.

These data suggest that early discrimination of outcome based on clinical variables is a practical possibility. Such discrimination will (i) not only improve prognostication but (ii) will also provide baseline data to evaluate therapeutic measures.

SOCIO-ECONOMIC AND RELATED LIFE-EFFECTS IN 180 PATIENTS WITH NARCOLEPSY-CATAPLEXY FROM POPULATIONS IN NORTH AMERICA, ASIA AND EUROPE COMPARED TO MATCHED CONTROLS

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A questionnaire survey of some 160 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 similarly located. The life effects were attributed by the patients to their primary symptoms of excessive daytime drowsi*ness, sleep attacks, cataplexy, vivid hypnagogic hallucinations,* and also to other frequently documented 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 (66%), 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 sleep occurring at work or in the home (49%) or related to smoking (49%) were much more frequent in patients. There were also deleterious effects on education, recreation and personality related to the symptoms. Narcolepsy can therefore produce a variety of serious side-effects probably more serious and pervasive than, for instance, those of epilepsy. This emphasizes the importance of adequate and early diagnosis and treatment.

SPECTRAL ANALYSIS OF THE BASIC ACTIVITY FOR CLINICAL APPLICATION OF THE EEG APPLIED IN PATIENTS WITH SPACE OCCUPYING LESIONS

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This method has been applied to 49 EEGs recorded in 37 patients with intra-cranial space occupying lesions (24 tumors, 9 intracerebral hematomas, 3 extracerebral hematomas and 1 large aneurysm). It will be shown that in this group of cases the method served to detect, localize and quantify EEG abnormalities. In the majority of cases this also led to a correct localization of the lesion, but there were exceptions, which, however, corresponded usually with those found on visual interpretation and revealed functional disturbances at a distance from the lesion. A comparison will be made with the findings in a series of 40 patients with acute unilateral cerebral ischemia and 20 controls. These cases were studied in detail by Van Huffelen (thesis, 1980).

PRIMARY CNS LYMPHOMAS: 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, Parkinson's disease and encephalitis respectively (three cases), and hemiparesis (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 diffuse meningeal 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 and electronmicroscopy, and immunoperoxidase technique for gamma globulins and lysozyme. The tumors were diagnosed as poorly differentiated - PD (two), immunoblastic non-cleaved (one) and lymphoplasmocytic (two). Light microscopy and immunocytochemistry were diagnostic in three cases but both PD were negative for immunological markers and required EM for unequivocal diagnosis. None of the tumors was clearly monoclonal.

Biopsied cases received cranial irradiation and chemotherapy and were followed for up to 12 months. Because of the potentially good therapeutic response of brain lymphoma, importance of early biopsy with the aid of electronmicroscopy and immunocytochemistry is stressed in cases with obscure diffuse or multifocal CNS disease which may represent lymphoproliferative disorder.

CLINICAL APPLICATIONS OF SUBCORTICAL SOMATOSENSORY EVOKED RESPONSES

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Clinical application of subcortical somatonsensory evoked responses has been limited by difficulties in the evaluation of the origin of individual components.

We have had the opportunity to correlate the result of depth electrode (DBS) recording with those obtained simultaneously from surface electrodes (C_z to contralateral clavicle) in patients after stimulation of the median nerves at the wrists and after peroneal nerve stimulation at the knees. After wrist stimulation the "thalamic" component can be recorded at latencies of 13 to 18 msec. With surface electrodes, peak positive components are seen at peak positive latencies of 9.6 ± 0.6, 12.2 ± 0.8, 14.6 ± 0.9 and 18.6 ± 0.6 msec. in 30 normal volunteers.

The "thalamic" component is often hard to identify in patients with multiple sclerosis. The responses have been delayed in patients with spinal cord tumors, posterior fossa tumors, Miller Fisher variant of the Guillain Barré syndrome, multiple sclerosis, brainstem cerebrovascular lesions, brainstem trauma, Reyes syndrome and polyarteritis. The latencies have been normal in patients with dystonia musculorum deformans, torticollis and Parkinson's disease.

Greatest clinical and diagnostic value of these subcortical somatosensory evoked responses is achieved by correlation with subcortical auditory, cortical somatosensory and visual evoked responses. These responses provide an atraumatic and practical method of assessing function of somatosensory pathways in a wide range of medical and surgical conditions.

PLASMA EXCHANGE AND MYASTHENIA GRAVIS

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Plasma exchange was used to treat thirty-seven patients (age 16 to 74 years) with moderate to severe myasthenia gravis. Three groups of patients were studied: 18 immediately pre- or post-thymectomy; 13 with relapses of myasthenic weakness; and 6 with chronic weakness. All patients were assessed clinically before and after plasma exchange. Serum anti-bodies to acetylcholine receptor were measured in all patients before plasma exchange. During each 2 to 2.5 L of plasma were removed and replaced with a combination of N-saline, stored and fresh frozen plasma.

Seventeen of the 18 patients undergoing thymectomy had normal or improved muscle strength following plasma exchange and 16 have remained in remission (on immunosuppressive drugs) for 3 to 20 months. One of these patients relapsed with bulbar weakness and respiratory failure following discharge after thymectomy.

Nine of the 13 patients in myasthenic relapse following thymectomy on Prednisone improved following plasma exchange and with adjustment and continuation of immunosuppression have remained improved or in remission for 4 to 18 months.

Five of the 6 patients with chronic weakness had immediate improvement with plasma exchange and 4 remain improved following multiple course of plasma exchange and immunosuppression.

The complications of plasma exchange in these 37 patients included urticaria, citrate toxicity and transient hypotension but serious problems were not encountered. Plasma exchange has a role in preventing significant bulbar and respiratory weakness and allowing uneventful introduction of Prednisone therapy in myasthenia patients undergoing thymectomy. Plasma exchange may be useful in preventing respiratory failure and prolonged hospitalization among myasthenic patients in relapse with chronic weakness, but continued improvement depends on adequate immunosuppression.

FAILURE OF PLASMAPHERESIS IN POST-THYMECTOMY MYASTHENIA GRAVIS

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Five patients are presented with myasthenia confirmed by clinical, electrical and serological criteria. All were taken to early thymectomy with no other anti-myasthenic therapy being employed. These patients experienced persistent ocular myasthenic features (diplopia and ptosis) following thymectomy, despite resolution of proximal muscle and bulbar weakness. When ocular symptoms failed to resolve during the ensuing 12 months, plasmapheresis was performed. These patients were consecutively studied Tensilon 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 normalization of the antibody titer suggests that another mechanism may be operational. The striking response to steroids in preference to plasmapheresis suggests that these agents may operate by different mechanisms.

THE CEREBRAL DYNAMICS OF ALBUMIN AND WATER FLUX AFTER INTRACAROTIC INJECTION OF HYPEROSMOLAR SOLUTIONS

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This study was carried out to determine the influence of infusion solution osmolarity on the flux of Evans BLue-albumin complex, isotope labelled serum albumin and H_20 into rabbit brain and CSF. The experimental model used was the ipsilateral infusion of a determined volume (4 cc/kg) of hyperosmolar saline solution (37°C) at a known pressure and varying only the osmolarity of the solutions from 0.29 Osm to 2.5 Osm.

No alteration in albumin permeability either into specific brain regions or CSF was found after infusion of iso-osmolar NaCl. Increasing the solution osmolarity to 0.5 Osm increased albumin penetration into CSF but not into cerebral regions. Progressive qualitative and quantitative increases in albumin flux were seen after infusion of 0.83 Osm, 1.6 Osm and 2.5 Osm solutions.

Following the changes in albumin flux and H_20 content after infusion of 0.83 Osm solutions demonstrated a marked decrease in albumin penetration into both brain and CSF 5 - 15 minutes after infusion with normalization of many brain regions 1 hour after infusion. A significant increase in specific ipsilateral regional brain H_20 content was found 5 minutes following infusion which was no longer present 25 minutes after infusion.

Our study demonstrates that increasing the osmolarity of cerebral infusion solutions results in an increase flux of albumin and H_20 into brain regions which reaches a maximum in the first 5 - 15 minutes after hyperosmolar infusion and then shows progressive normalization over 1 h.

THE EFFECTS OF MANNITOL ON THE CEREBRAL, CARDIAC AND PULMONARY CIRCULATIONS IN NEUROSURGICAL PATIENTS

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Neurosurgical patients undergoing craniotomies were specifically selected for this study by their need for intravenous mannitol. Informed consent was obtained from all patient for the placement of internal carotid catheters for $Xenon^{133}$ injection and cerebral blood flow measurement. Anaesthesia was induced with thiopentone, the trachea intubated, muscle relaxation produced by pancuronium and anaesthesia was maintained with N_2O O_2 and enflurane. Artificial ventilation produced moderate hypocarbia. A transfemoral internal carotid catheter monitored arterial blood pressure, enabled blood gas and hematology sampling and served as an injection site for $Xenon^{133}$. A Swan Ganz thermodilution cardiac catheter was advanced into the pulmonary artery for determination of central venous pressure, cardiac output, pulmonary artery pressure and pulmonary capillary wedge pressure. All cerebral, cardiac and pulmonary parameters were measured prior to, during and post mannitol infusion at an infusion of 1.0 to 1.3 Gms·Kg of body weight. The significant features of this study were the large increases in cardiac output and cerebral blood flow; the maintainence of normal blood pressures and the marked changes in cerebral and peripheral vascular resistance due to vascular dilation.

MECHANISMS OF GROWTH OF CHRONIC SUBDURAL HEMATOMAS — IS A FLUID SHIFT INVOLVED?

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It has been hypothesized for some time that chronic subdural hematomas grow in volume by a differential osmotic or oncotic pressure between the hematoma contents and the adjacent vascular or CSF compartment. Oncotic pressure is that which results across a membrane whose pores are sufficiently large that they are impermeable only to huge organic molecules such as albumin but allow many smaller molecules in addition to water to pass freely. With the recent development of membrane oncometers with fast response times it has become feasible to measure oncotic pressures of biological samples. It was felt that the direct simultaneous measurement of oncotic pressure in hematoma fluid and blood would provide an opportunity to test the oncotic hypothesis of hematoma growth. The oncotic pressure in the blood of 14 patients with chronic subdural hematomas was 25.9 ± 1.7 mm Hg., the hematoma fluid had a pressure of 27.5 ± 1.9 mm Hg. There was therefore no significant difference. On the other hand in six cases of subdural hygromas there was a significant difference with blood having a pressure of 20.3 ± 2.2 and hygroma fluid being 7.2 ± 3.2. This finding fails to support the theory that chronic subdural hematomas grow and produce symptoms after latent intervals because they attract fluid from the blood via dural vessels.

"REDUCED ANESTHESIA REQUIREMENT IN PATIENTS AFTER PERIAQUEDUCTAL GRAY STIMULATION"

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Electrical stimulation of the periaqueductal gray region of patients with intractable pain has produced relief concommitant with elevation of CSF B-endorphin levels. Accumulation of norepinephrine occurs in the same periaqueductal area in the rat during halothane or cyclopropane anesthesia. It is hypothesized that anesthetic requirement may be affected by selective stimulation or inhibition of specific brain regions.

To determine whether stimulation of the periaqueductal gray area changes anesthetic requirement, we studied the minimal alveolar concentration (MAC) of halothane and 60% nitrous oxide (amount of anesthetic needed to prevent movement in response to skin incision in 50% of patients) in 22 patients, ages 29 to 65, with previously implanted periaqueductal gray electrodes who were undergoing 23 procedures for electrode internalization. Patients had neither used their stimulators for 24

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 determined by a modification of the Dixon "Up and Down Method". End tidal halothane concentration was monitored through analysis of expired gas by mass spectrometry.

MAC of halothane and 60% nitrous oxide for patients stimulated immediately before surgery was 0.10% which is significantly less by the Waud test than 0.50%, the MAC for patients in the unstimulated group. Ventricular CSF B-endorphin levels (pgm/ml) measured in one patient were 276 before stimulation, 966 at 15 minutes after stimulation, 1011 at 30 minutes, and 1065 at 60 minutes. Thus, anesthetic requirements may be reduced by periaqueductal gray stimulation. The duration of analgesia is apparently less than 24 hours so that stimulation must be renewed for continued effect.

BLOOD-BRAIN BARRIER (BBB) DYSFUNCTION FOLLOWING EXPERIMENTAL SUBARACHNOID HEMORRHAGE (SAH)

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The pathophysiology of clinical deterioration in some cases of SAH is still unexplained. A BBB dysfunction was postulated to be a possible mechanism involved.

Experiments were performed with 35 cats. Metaraminol-induced arterial hypertension or carotid perfusion of 6.0×10^{-5M} solution of mercuric dichloride ($Hg\ C1_2$) were used to cause BBB breakdown. The SAH was simulated by cisternal injection of autogenous blood. The BBB breakdown was detected by the extravasation of Evans blue into the extravascular space leading to staining of the brain. The specimens were further examined under the fluorescent microscope.

The animals were divided in the following groups: (1) controls, (2) arterial hypertension, (3) $HgC1_2$, (4) SAH, (5) SAH followed by arterial hypertension, and (6) SAH followed by $HgC1_2$ perfusion. Animals in which arterial hypertension had been induced and those treated with $HgC1_2$ showed well defined areas of BBB breakdown. Animals submitted to SAH failed to display BBB leakage. BBB breakdown was neither seen in animals made hypertensive nor in animals injected with $HgC1_2$ following SAH. Therefore, SAH did not cause BB8 breakdown by itself. On the contrary, it prevented the BBB breakdown induced by the damaging agents. The mechanism of action of these agents has been postulated to involve increased pinocytosis through the capillary endothelium. Our results suggest that SAH interferes with this active pinocytotic process.

Pinocytosis inhibition has been reported by others during the acute stage following cerebral ischemia.

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), analagous 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.

Alterations in the BNB were assessed by both fluorescent tracing of intravenously-injected bovine serum albumin labelled with Evans' blue and by electronmicroscopic tracing of I.V. -injected horseradish peroxidase. The right sciatic nerve of 4 groups of 25 animals were subjected to either 1) nerve injection injury with a variety of antibiotic, steroid and local anaesthetic agents, 2) nerve transsection followed by epineurial suture, 3) nerve transsection with neuroma formation, or 4) crush injury. The left sciatic nerve was used as the control throughout. The nerves were studied at varying periods from 1 hour to 8 weeks following injury.

Results revealed that in the normal nerve the intravenously-injected tracer proteins were confined to the lumen of the endoneurial microvessels and epineurium 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.

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 fibre 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 week, spread throughout the distal segment undergoing Wallerian degeneration. Regeneration was followed closely by restitution 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. Included 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 favourably 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 by increase in water content of the brain but by acute congestion of the brain, vasodilatation and hyperaemia. Studies of cerebral blood flow, cerebral blood volume and CT scan density are all in favor of this concept. In the second group of children with immediate onset of unconsciousness, 60% developed evidence of extracerebral 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 neurological 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 INTERNAL NEUROLYSIS IN PERIPHERAL NERVE SURGERY

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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 dangerous procedure which carries risk of injury to nerve and of damage to the microcirculation, resulting in intrafascicular and perineurial scarring. Paucity of laboratory work on the subject prompted the present study designed to examine morphological and physiological alternations in nerve fibres following internal neurolysis.

Internal neurolysis was performed on the right sciatic nerve of 108 Wistar rats using the operating microscope and microsurgical techniques. Groups of 9 animals were studied from 1 hour 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 tracing of intravenously-injected Evans' blue albumin and by electronmicroscopic tracing of horseradish peroxidase.

Carefully performed internal neurolysis does not result in any significant nerve fibre damage as assessed by both light and electronmicroscopy. Connective tissue stains revealed only minimal degree of epineurial and perineurial fibrosis and no evidence of intrafascicular scarring. Bloodnerve barrier studies showed transient increased permeability of perineurium and endoneurial microvessels to tracer proteins at 1, 24 and 48 hours, no longer evident at 1 week. Electrophysiological studies revealed transient early 20-40% drop in nerve conduction velocity in the operated as compared to the control nerve 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 perineurium.

It is concluded that experimental internal neurolysis per se does not result in significant lasting morphological and physiological alterations in normal rat peripheral nerve. Nevertheless, care must be taken to avoid damage to the perineurium 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), giving fractionated treatments to the whole head. Groups of 14-20 rats received total doses of 2300, 4600, or 5750 rads 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 4600 and 5750 rads doses were significantly more effective than 2300 rads. In other series of experiments, BCNU chemotherapy (10 mg/kg) was combined with 2300 and 4600 rads radiation therapy. Synergism of therapies was demonstrated. Methyl prednisolone acetate (2 mg/kg twice weekly over 4 weeks) alone did not affect survival curves and its combination with 4600 rads radiation therapy negated the prolongation of survival with 4600 rads alone.

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 principal mechanism resulting in increased permeability is enhanced pinocytosis. In order to exclude the possibility that the enhanced pinocytosis was a direct effect of angiotensin, cerebrovascular permeability alterations were studied in rats with nonpharmacologically induced hypertension.

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Rats received horseradish peroxidase (HRP) intravenously, following which hypertension was induced by clamping the abdominal aorta. Animals were sacrificed 2 ½ 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 focal segments of penetrating arterioles in the temporal and parietal cortex showed increased permeability to HRP. Permeable vessels showed increased numbers of pinocytotic vesicles 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 neurones of adult mongols' brains. The area (and hence volume) of six cortical "zones" surveyed was measured with a digitizer and programmable calculator.

In decreasing magnitude of affliction, the rank order for neurofibrillary tangles was: entorhinal > subiculum > H1 > endplate > H2 > presubiculum. That for granulovacuolar change was: subiculum > H1 > H2 > endplate > entorhinal > presubiculum.

These striking regional predispositions suggest that a common neurotransmitter deficit may underlie the local selectivity of such lesions in mongoloid brains; and the predilections' marked similarities to ranking orders already noted both in normal aged subjects and especially in senile dements of the Alzheimer type (Ball, M.J., Acta Neuropath. 42: 73-80, 1978) enhance the importance of the Down's syndrome neuraxis as a key to the puzzle of Alzheimer's dementia.

AKINETIC MUTISM: CONTRIBUTING DYNAMIC FACTORS

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6 cases presenting a state of akinetic mutism are reviewed. They presented a normal or low pressure hydrocephalus of various etiology; aqueductal stenosis, previous posterior fossa surgery for cerebellar hemangioblastoma and atresia of the foramen of Magendie, subarachnoid hemorrage and cystic tumor of the third ventricle.

Surgical treatment consisting in aspiration, partial excision and radiotherapy for the cystic tumor of the third ventricle and CSF shunting procedures in the other cases was successfull with clinical recovery.

The authors emphasize the role of mechanical factors as a cause of the state of akinetic mutism in their patients. The importance of recognizing hydrocephalus, even if mild and of the low-normal pressure type is discussed.

There seem to be a common denominator in these cases which is the enlargement of the 3rd ventricle associated with hydrocephalus or intraventricular cystic tumor. It is postulated that this interferes with the function of periventricular structures through direct pressure, oedema or reduction in blood flow to cause this syndrome.

NEUROFIBROMATOSIS AND MACROCRANIA - MEGALENCEPHALY

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Macrocrania as a feature of neurofibromatosis has not been much emphasized in the past. Sixty-one children with neurofibromatosis, half of whom were familial cases were assessed regarding head size. All had measurements of the occipital frontal circumferences (O.F.C.) and standard skull x-rays. Six children had pneumo-encephalograms and 25 had CT-Scans to investigate macrocrania or confirm the presence in nine patients of optic nerve or chiasm gliomas.

Head size by O.F.C. was skewed toward upper percentiles for age,(15 of 61 at or greater than the 98%ile) while stature (height) was skewed towards the lower percentiles for age. There was a low order correlation between macrocrania and other features including intellectual deficit, motor deficits, and EEG abnormalities.

From plain skull radiographs, the cranial capacity was estimated and found to be above the 95%ile 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, 619-635).

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.