Abstracts of the Scientific Programme of the XVI Canadian Congress of Neurological Sciences

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PARTICIPATING SOCIETIES

Canadian Neurological Society
Canadian Neurosurgical Society
Canadian Society of Electro Encephalographers, Electromyographers and Clinical Neurophysiologists
Canadian Association for Child Neurology
Canadian Association of Neurological and Neurosurgical Nurses

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XVI CANADIAN CONGRESS OF NEUROLOGICAL SCIENCES

Calgary, Alberta

PRE-CONGRESS COURSES

WEDNESDAY 24 JUNE, 1981

A. ANATOMICAL BASIS OF CONTEMPORARY NEUROSURGERY - Chairman - Dr. B.K. Weir

Dwight Parkinson	"The cavernous sinus and basal veins."
Sidney Peerless	"The incisura and basilar artery."
Donlin Long	"Anatomy of pain."
Harold Hoffman	"Developmental anomalies."
Normal Chater	"Scalp and surface arteries."
Jules Hardy	"Pituitary anatomy."
Douglas Miller	"Ventricular system and spinal fluid pathways."
Bennett Stein	"Posterior third ventricle and pineal region."
Gazi Yasargil	"The importance of cisternal anatomy for microneurosurgery."

B. NEUROENDOCRINOLOGY COURSE - Chairman - Dr. Leo Renaud

Joseph Martin	Neuroendocrinology: An Overview of its Development and Scope.
Robert Elde	Anatomy of the Endocrine Hypothalamus: Aminergic and Peptidergic Pathways.
Warren Veale	Role of the Hypothalamus in Thermoregulation and Febrile Convulsions.
Leo Renaud	Neurophysiology of the Endocrine Hypothalamus.
Quentin Pittman	Hypothalamic Peptides: Functional Correlates.
Otto Rorstad	Hypothalamic Regulation of Growth Hormone Secretion.
Michael Thorner	Prolactin Secretion: Physiology and Pathology.
Bernard Corenblum	Medical Approaches to the Management of Neuroendocrine Disorders.
Jules Hardy	Surgical Approaches to the Management of Neuroendocrine disorders.

C. ELECTROENCEPHALOGRAPHY - Chairmen - Dr. Warren Blume
Dr. Jean Reiher

Pierre Gloor	"Physiology of the E.E.G."
Pierre Gloor	"Principles of polarity and potential fields."
Gordon Blair	"I.C.U. recordings."
Jean Reiher	"Recognition of some difficult to recognize E.E.G. patterns."
Adrian Upton	"Artefactual pitfalls."
Don McLean	"Clinical significance of some major E.E.G. patterns."
Morton Low	"Introduction to evoked potentials."
Andrew Eisen	"Somatosensory Evoked Potentials."
Sherrill Purves	"Visual and Auditory Evoked Potentials."

For course registration write to:

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I - 1

IMMUNE REGULATION IN MULTIPLE SCLEROSIS (MS): T CELL SURFACE MARKER ANALYSIS

D. PATY, N. MORGAN, C. SYLVESTER
Vancouver

There is good reason to postulate that MS is a disorder of immune regulation. We have looked at T cell surface antigens using anti-human monoclonal antibodies Leu-1 (anti T-cell), Leu-2a (anti Suppressor cell) and Leu-3a (anti Helper cell, Becton Dickinson) with an indirect method via Biotin-Avidin conjugate with fluorescein. Analysis of cell numbers, viability, and fluorescence was by a FACS-IV (Becton Dickinson) fluorescence Activated Cell Sorter. The T cell subfractions found were as follows (% lymphocytes ± SD):

	T Cell	S Cell	H Cell
Normals (N = 18)	72.8 ± 8.2	21.1 ± 8.0	46.4 ± 10.9
MS: Stable (N = 30)	71.3 ± 8.9	20.1 ± 7.6	49.1 ± 8.8
Chronic Progressive (N = 16)	65.2 ± 14.6*	11.8 ± 6.0†	52.6 ± 10.0

The suppressor cell levels in chronic progressive disease were significantly low † ($p \leqslant 0.0005$) when compared to stable MS and normals. T cell levels were also significantly low*, ($P = 0.037$) but less markedly so than the S cells. The finding of low S cell levels is chronic progressive MS supports the concept that biological disease activity is related to a failure in regulator mechanisms. This failure in suppression could then allow the appearance and activity of autoreactive cytotoxic T cells.

We are following the stable MS patients and will re-test them within 24 hours of the onset of a new relapse. In this way the patients will act as their own controls.

I - 2

NEW BRAIN PARTICLES AND NERVOUS DISEASE

P. AVERBACK
Montreal

The relationships of so-called normal senile brain changes and pathological nervous system findings in presentile dementia, senile dementia, senile psychosis and senile brain changes in brain diseases other than Alzheimer's disease are complex, although it is generally acknowledged that qualitative overlap exists. Microscopic hallmarks of Alzheimer's disease such as senile plaques, neurofibrillary tangles and granulovacuolar degeneration are considered quantitatively more significant in that disease although the same degenerative changes may be found in normal aged brain, and may indeed be equally severe in conditions such as Parkinson's disease. An analogous concept has evolved for Parkinsonism and the Lewy body, which is less common. Newly recognized inclusion bodies are described as consistent findings in Alzheimer disease, Parkinson's disease, so-called senile brain atrophy and some forms of psychosis. The electron microscopic interpretation of these structures is most consistent with virus particles, virus precursor or virus product, and matching photographs with established agents are presented in detail to lend support to this interpretation. Homogeneity of size and pattern of large (ultrastructural) masses of the particles in involved cells and fine structural preservation of other cellular elements are shown in detail to support further and non-artefactual nature of the structures. Identical methods on suitable controls (i.e. absence of degeneration) have been thus far negative.

Patient data from 25 cases of Parkinson's disease, 25 cases of Alzheimer's disease, and 25 aged brains and senile psychosis, and matched controls, and methods are briefly summarized. It is suggested that these various diseases and some degree of "normal" aging may possibly be differing manifestations of a common underlying pathogen.

I - 3

C2 DEFICIENCY AND STROKE

D. SIMARD, C. PARENT, J-P. MATHIEU
Québec

A genetic deficiency of the second complement (C_2 deficiency) was first described in 1966 by Klemperer and al.

Since then, C_2 deficiency has been found in association with various connective tissue and immunological disorders (dermatomyositis, myasthenia gravis etc.) and even with multiple sclerosis.

However cerebrovascular events have not yet been reported in C_2 deficient patients. We had the opportunity of investigating in the past two years three young patients who presented with a dense neurological deficit and who were found to have a C_2 deficient vasculitis.

The clinical immunological and genetic aspects of this new neurological entity will be presented.

I - 4

TOPOGRAPHIC ELECTROENCEPHALOGRAPHIC STUDY OF TIAs IN ASYMPTOMATIC PERIOD

K. NAGATA, M. HIRANO, G. ARAKI, M. MIZUKAMI
Isesaki, Gunma, Japan

Does the brain function of the patients with transient ischemic attacks (TIAs) recover completely as the clinical symptoms disappear within 24 hours? Twenty-five patients with TIAs of internal carotid arterial system were studied by means of CT, cerebral angiography and computed mapping of electroencephalogram (CME) in the asymptomatic period. The CME is a microcomputer system to display scalp topograph of square roots of average powerspectra over each EEG frequency band on color television. CT demonstrated a basal ganglionic small low density area in two cases and no low density area in 23 cases. Fourteen patients showed no abnormality on angiogram, eight showed stenosis of the main trunks of cerebral arteries and two had occlusion of the internal carotid artery. One patient had an unruptured aneurysm in the left middle artery. The CME demonstrated normal symmetrical alpha activity in 2 cases, diffuse slow alpha activity in 2 cases, foci of theta activity in 8 cases, asymmetrical alpha activity in 12 cases and low voltage beta activity in one case. The correspondence of the lesion side on CME to the lesions suspected through the clinical symptoms was 68% (17 of 25). Especially, the correspondence was 82% in the patients with abnormal angiogram and it was as high as 88% in the examination within 2 weeks of the attacks. Using CME, a lesion side of TIA can be detected even in the asymptomatic period. Accordingly, it can be said that the brain dysfunction caused by an ischemic event such as TIA may persist for a long period though the clinical symptoms disappear completely.

I - 5

CEREBRAL ARTERIOVENOUS MALFORMATIONS IN CHILDHOOD

R. HUMPHREYS, E. HENDRICK, H. HOFFMAN, E. GARRIDO
Toronto, Philadelphia

Arteriovenous malformations (AVMs) of the brain are the commonest cause of hemorrhagic stroke in children. A review (which excludes venous circulation abnormalities) of 85 children from the Hospital for Sick Children, Toronto, reveals that the features of pediatric AVMs are different in children, than in adults. Almost 80% of children with AVM present with intracranial hemorrhage, and the mortality from the first hemorrhage is 24%. Chronic epilepsy occurs half as frequently in children as in adults, prior to the diagnosis of the malformation, and is usually halted with surgical excision of the lesion. The malformations in children are scattered throughout the brain, often in awkward locations; unlike adults, an unusual number are found below the tentorium.

Because of the child's susceptibility to repeat hemorrhage, *total excision of the malformation, wherever* possible must be attempted with the expectation that the child's brain vasculature may be more resilient to operative intrusion. The operative mortality is 11% and 65% of patients surviving operative care can be expected to be neurologically intact.

I - 6

CONSERVATIVE OR SURGICAL MANAGEMENT OF INTRA-CEREBRAL SPONTANEOUS HEMORRHAGE A RETROSPECTIVE CLINICAL APPRAISAL

J. CASTEL, J. DARTIGUES, J. ORGOGOZO
Bordeaux, France

The benefit of surgical versus conservative management of intra-cerebral spontaneous hemorrhage caused by arterial hypertension must be re-evaluated. The CT scanner offers a powerful new appraisal of location and volume of the hemorrhage. Medical resources to treat high intracranial pressure are also improving.

A clinical, retrospective study of 45 medically and 39 surgically treated patients with spontaneous hemorrhage in the basal ganglia (by CT scan) was done. Surgically treated patients had the largest hemorrhages and the higher mortality and disability at the first month.

We concluded that whatever may be the volume of the clot and the treatment, the evolution of the initial state of consciousness had the most predictive value for the final outcome of these patients.

I - 7

TREATMENT OF CAROTID CAVERNOUS FISTULA

P. ALLEN, R. DELEO
Edmonton

Experience with a technique of directly closing a carotid cavernous fistula by thrombus production is discussed. Patients with carotid cavernous fistulae, post traumatic, secondary to aneurysm rupture developmental, presented with intracranial bruit, unilateral exophthalmus and chemosis. Angiography confirmed the fistula and thrombosis occurred following insertion of fine copper or steel wires directly into the cavernous sinus. In each case, flow in the internal carotid artery remained normal, morbidity was minimal and the pre-operative signs and symptoms disappeared rapidly.

Thrombogenesis and direct closure of a fistula, within the cavernous sinus, is a preferable technique to arterial occlusion and is applicable to the treatment of other intracranial vascular problems.

I - 8

EXTERNAL CAROTID ARTERY STENOSIS AFTER EXTRACRANIAL—INTRACRANIAL ANASTOMOSES

H. SCHULTZ, M. CHIU, K. TERBRUGGE
Toronto

Fifty-seven extracranial to intracranial artery anastomoses were done in patients for inaccessible arteriosclerotic cerebrovascular disease. To assess the patency of the anastomoses, arteriograms were done three months after surgery. Severe stenosis of the external carotid artery had developed in seven patients, even though that artery had been clear of disease three months earlier. In these patients, the microvascular anastomosis was widely patent, and the superficial temporal artery had at least doubled in size. Three patients have suffered transient ischaemic events due to the new stenosis at the origin of the external carotid artery, requiring endarterectomy and/or patch grafts of the freshly diseased artery.

It appears that additional haemodynamic stress may predispose the proximal external carotid arteries to accelerated atherosclerosis. The transition of laminar to turbulent flow may be responsible for acceleration and accentuation of the arteriosclerotic process in predisposed *arteriopathic patients.*

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I - 9

MIGRAINE AND PARTIAL SEIZURES WITH COMPLEX SYMPTOMATOLOGY IN CHILDREN: AN ASSOCIATION?

J. REGGIN, S. SESHIA
Winnipeg

There is a recognized relationship between migraine and epilepsy in children but little attention has been paid to the types of seizures in this association. We therefore report on 20 children who had migraine and partial seizures with complex symptomatology. The median age at the time of presentation was 11.2 years (range 6-17 years), of onset of migraine 8.5 years (range 3-17 years) and of seizures 10.0 years (range 3-12 years). Seizures preceded the onset of migraine in 6 cases and both commenced simultaneously in 4. The types of migraine (W.F.N. classification) were: common in 17, classic in 3, hemiplegic in 2, basilar in 1; 3 cases had more than one type. Seizures were subtyped according to the International Classification: impaired consciousness alone (1), cognitive symptomatology (5), affective (3), psychosensory (5), psychomotor (2) and compound (4). Cases with EEG abnormalities in the temporal regions but whose attacks did not fulfill the clinical criteria of the International Classification were excluded. A family history of migraine was obtained in 70%, of seizures in 30% and of both in 25%. EEGs were normal in 5/19; 13 of the remaining 14 had focal abnormalities over one/both temporal regions: anterior temporal (5), mid-temporal (7), posterior temporal (9); abnormalities at more than one site occurred in 8 and were bilateral in 3. The abnormal wave forms were: sharp (9), spike-slow wave complexes (5), slow waves (3); 3 cases had more than one type of abnormal wave form in the same record. Generalised sharp slow-wave paroxysms occurred in 2. We suggest that our cases represent an association between migraine and partial seizures of complex symptomatology and that some of those with "migraine equivalents" may well be examples of this relationship.

II - 1

THE EFFECT OF PROSTACYCLIN IN EXPERIMENTAL TRAUMATIC PARAPLEGIA IN RATS

R. CHAN, J. SCHWEIGEL, G. THOMPSON
Vancouver

This is a preliminary report of the role of Prostacyclin-Thromboxane system in experimental spinal cord injury. Severe blunt trauma to the spinal cord was induced in 50 rats by dropping a 2-gram weight from a height of 25 cm onto an impounder. The impounder was set lightly on the exposed dura at T3-4 vertebral level. Ten rats were randomly selected for the control group and 40 rats were in the Prostacyclin group. Fifty micrograms of Prostacyclin per rat were administered intraperintoneally at time intervals of 15, 30, 60 and 90 minuted following the spinal cord injury. Ten rats were used in each of the time groups. The animals were followed for 4 weeks before they were sacrificed. Segments of the spinal cord were removed. The injury site and segments proximal and distal to the area of injury were examined histologically. All animals that received Prostacyclin 15 minutes after the spinal cord injury recovered with normal hind limb functions. Microscopically, small areas of central hemorrhagic necrosis was found at the level of injury only. The remaining Prostacyclin treated rats failed to show any significant improvement compared to the control rats.

III - A - 1

HEMIFACIAL SPASM: CAUSE AND CURE

D. FAIRHOLM, K. LIU, J. WU
Taipei, Taiwan

Compression of the root entry zones of cranial nerves can cause paroxysmal activity. We have now operated on 15 patients with progressing hemifacial spasm. They had many previous attempts at therapy ranging from Tegretol to Chinese herbal medicine. All patients had angiography and some had CT scans. All underwent unilateral posterior fossa craniectomy laterally to the sigmoid sinus. The cerebellum was elevated and the root entry zone of VII and VIII inspected. All had compression by arterial loops, usually AICA or its branches. The offending vessel was relocated using Teflon sponge. 14 of 15 patients were cured. 2 had transient facial weakness, 4 had decreased hearing, 3 had increased tinnitus, 1 had facial herpes, 1 had hiccups. The treatment of hemifacial spasm is relocation of the offending vessel and this can be done with low morbidity and mortality.

III - A - 2

COMPARISON OF MICROVASCULAR DECOMPRESSION AND PERCUTANEOUS TRIGEMINAL RHIZOTOMY IN THE SURGICAL TREATMENT OF TRIGEMINAL NEURALGIA — REPORT OF ADDITIONAL CASES AND FURTHER FOLLOW-UP

D. BRETT, G. FERGUSON, S. PEERLESS, H. BARR, J. GIRVIN
London

We are reporting our further experience and larger complete follow-up in twenty-four patients (average age 52 years) treated by microvascular decompression (MVD); and in fifty-four patients (average age 65 years) treated by percutaneous trigeminal rhizotomy (PTR).

In the MVD group (average follow-up 28 months), 17 patients (71%) have remained pain free with three (12%) immediate failures and four (17%) delayed recurrences.

In the PRT group (average follow-up 30 months), 29 patients (54%) have remained pain free with two (4%) immediate failures and 23 (43%) delayed recurrences.

Complications in this series, surgical pathology found, as well as the advantages and disadvantages of each procedure will be discussed.

The relative value of MVD and PTR in the surgical treatment of trigeminal neuralgia will only be determined by longer follow-up in this and other larger series.

III - A - 3

SELECTIVE PERIPHERAL DENERVATION FOR SPASMODIC TORTICOLLIS

C. BERTRAND, P. MOLINA NEGRO, S. MARTINEZ
Montreal

During the past 3 years, selective peripheral denervation has been used almost exclusively for the treatment of spasmodic torticollis. Originally it was an adjunct to stereotactic surgery. However, while blocking specific nerves or rami during electromyography, it became evident that peripheral denervation could produce marked or total relief of dystonic neck movements. In a typical case of laterocollis denervation would usually involve the sterno-cleido-mastoid muscle on one side and C1 together with the posterior rami of C2, C3, C4, C5 and C6 on the opposite side. In certain cases of retrocollis the posterior rami have been avulsed down to C5 on both sides.

It has many advantages over rhizotomy: 1) Only the muscles involved are denervated and the anterior cervical group remains intact so that rehabilitation is much easier and more complete. 2) There is no laminectomy so that the stability of the neck is not compromised. 3) Denervation may be carried down to C7 on one side if required since the phrenic nerve is not involved. 4) The operation can be tailored to the needs of the patient.

It is also a more dependable and a more benign procedure than a thalamotomy and/or pallidotomy although it is still used in conjunction with stereotactic surgery in the more severe forms of dystonia.

Out of 26 cases, 18 have had complete or almost complete relief of symptoms and the other 8 patients were improved markedly. In 3 cases of retrocollis there was marked improvement in 2 cases and only moderate improvement in the other.

III - A - 4

CORDECTOMY — CLINICAL-PATHOLOGICAL CORRELATION IN SEVEN CASES

Q. DURWARD, G. RICE, M. BALL, J. KAUFMANN
London

Cordectomy is rarely performed. Seven patients with varying clinical indications have undergone this procedure. History, neuropathological findings, and clinical results were correlated in each case.

Four patients had progressive arm pain and neurological deficit caused by post-traumatic syringomyelia. Their cordectomy specimens revealed a syrinx lined by dense gliosis in three cases. Schwannosis and leptomeningeal fibrosis occurred close to the level of injury, but not further away. All cases improved post-operatively.

Two patients with post-traumatic paraplegia had severe spontaneous lower limb dysesthetic pain. Cordectomy specimens revealed a small syrinx in one case. Schwannosis, gliosis and leptomeningeal fibrosis were marked. Long-term pain relief was not achieved.

One patient with multiple sclerosis underwent complete lumbar cordectomy for uncontrollable leg spasticity. Schwannosis was absent, but leptomeningeal fibrosis and gliosis were prominent. The spasticity was cured.

The present of a syrinx cavity in the cordectomy specimen has correlated well with clinical improvement in patients with a syringomyelic syndrome. Gliosis and leptomeningeal fibrosis were present in different types of cord injury with different clinical presentations. Schwannosis, possibly representing an attempt at neural repair, was seen close to sites of traumatic cord injury. These last three features correlated poorly with the clinical indication for cordectomy or post-operative result.

III - A - 5

SEIZURES AS A MANIFESTATION OF TIA'S IN BILATERAL CAROTID STENOSIS

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Monterrey, Mexico

Early contributions on TIA pathogenesis laid stress on their possible hemodynamic origin. However, the artery-to-artery embolic theory which suggests that the platelet-fibrin thromboemboli arising in the large extracranial arteries and embolizing to the smaller cerebral branches satisfactorily accounts for 70 to 90% of all the cases of TIA. Focal neurological and retinal ischemic events are uncommonly the result of hemodynamic mechanisms; on the other hand, the symptoms of diffuse reduction in cerebral perfusion consist mainly of blurred vision, impaired consciousness, vertigo and seizures. By definition, these episodes have been excluded in the diagnosis of TIA. A series of eight patients with seizures as the only manifestation of TIA or associated with transient hemiparesis is the subject of this report. Five of these patients had bilateral kinks of the internal carotid arteries and three had bilateral obstructions of more than 50%. In all of the above cases, the clinical presentation was focal seizures and in three cases the syndrome of hemiconvulsion-hemiplegia epilepsy was present. Bilateral endarterectomies were preformed in three cases with carotid obstuction and resection of kinks with termino-terminal anastomosis in all the rest. At an average follow-up of 18 months seven of the eight patients were well and asymptomatic. One patient died of myocardial infarction 12 months after the second operation (endarterectomy); but none of the eight cases had seizures or TIA's postoperatively. We wish to call attention to the clinical picture of bilateral stenosis of carotid arteries and the occurrence of frequent seizures. The ictus lasted less than 30 minutes, and although sometimes postictal paresis occurred, this cleared completely in less than 24 hours. This symptom was completely alleviated by surgery which carried an operative mortality of zero and a morbidity of six percent.

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III - A - 6

PROPHYLACTIC CAROTID ARTERY REPAIR ON THE ASYMPTOMATIC SIDE CONTRALATERAL TO PREVIOUSLY OPERATED SYMPTOMATIC CAROTID STENOSIS

M. FAZL, W. TUCKER
Toronto

In the past 4 years, WST has performed 152 carotid endarterectomies in 123 patients. In 29 patients, bilateral endarterectomies were done, usually approximately 3 months apart. In 27 of the 29, the second side had been asymptomatic, but moderate or severe disease was noted angiographically. The management of asymptomatic carotid stenosis contralateral to a symptomatic lesion remains controversial, and it is this group of patients we wish to discuss.

The average age of the patients was 61.7 years (range: 42 to 71 years). There were 17 male and 12 female patients. There was no operative mortality. There was no serious morbidity following the first side. One patient suffered a persisting post-operative neurological deficit after repair of the asymptomatic second side.

These patients have been followed for an average of 21 months (range: 1 month - 41 months) since their second operation. There have been no further symptoms in the distribution of the originally symptomatic artery; symptoms developed in the distribution of the "asymptomatic" artery before it was repaired in 5 cases. Two patients had had bilateral symptoms at the time of inital presentation. None of the patients have had symptoms post-operatively in the distribution of the second artery.

We suggest that repair of major stenosis of the carotid artery contralateral to a symptomatic lesion is potentially beneficial if a low operative mortality and morbidity are maintained.

III - A - 7

THE NATURAL HISTORY OF THE ASYMPTOMATIC CAROTID BIFURCATION LESION: IMPLICATIONS FOR MANAGEMENT

Q. DURWARD, G. FERGUSON, H. BARR
London

Significant stenosis (>50%) and/or ulceration of the internal or common carotid artery at the bifurcation is an indication for endarterectomy in the patient with a T.I.A. or minor stroke in that vessel territory. A similar, but asymptomatic, lesion is frequently encountered in the opposite carotid artery. Controversy exists as to whether this lesion should be treated surgically.

We have followed 43 patients with angiographically identified asymptomatic stenosis and/or ulcer in the opposite carotid artery for periods ranging from six months to six years (average follow-up 2.5 years). During this time, nine patients have developed cerebral ischemic symptoms in this previously asymptomatic carotid territory (21%). However, in only one case did the ischemia manifest as a stroke. In eight cases, a T.I.A. occurred. These patients then underwent carotid endarterectomy.

During this follow-up period, seven deaths have occurred — four from cardio-vascular disease, two from stroke (one carotid territory, one vertebro-basilar territory) and one from cancer.

This data suggests that the asymptomatic carotid lesion is associated with a low incidence of stroke. This, curiously, contrasts with a recognized stroke incidence of 50% as the initial manifestation of cerebral ischemia in the general population. The data does not support the idea that asymptomatic carotid lesions should be operated on prophylactically. They should be kept under regular review and consideration given to endarterectomy if appropriate cerebral ischemic symptoms develop.

III - A - 8

PITUITARY BASOPHILISM 1981: TUMOUR AND "PSEUDOTUMOUR" IN CUSHING'S DISEASE

H. SMYTH
Toronto

The sella was enlarged in only one of ten patients with well-documented pituitary-dependent hypercortisolism. A basophil adenoma was found in four patients; two harboured non-adenomatous focal hyperplasia. One diffusely enlarged gland showed only Crooke's hyaline change, and focal Crooke's change was the only abnormality in another gland of normal size. Two patients underwent selective anterior median wedge resection of the adenohypophysis after negative explorations for tumour.

Four of the first eight patients have had sustained remission of disease. Of three who improved, two had early, and one a delayed recurrence of severe hypercortisolism. One patient has sustained remission following wedge resection alone, while the other shows progressive biochemical abnormality.

Of the four cases of Nelson's syndrome, the sella was enlarged in three, and basophilic adenomas were removed from all four. Two patients improved, and two have sustained recovery. Selective adenomectomy in one of these led to clinical and biochemical remission of both Nelson's syndrome and recurrent Cushing's Disease.

These findings support the hypothesis that pituitary basophilism is a varied response of corticotroph cells to an unknown extra-pituitary stimulus. The spectrum of pathological change, which includes enlargement, hyperplasia and adenoma formation, may represent a morphological continuum of progressive response to this stimulus.

III - A - 9

APOPLEXY, HAEMORRHAGE AND NECROSIS IN PITUITARY ADENOMATA

G. MOHR, F. ROBERT, M. FINLAYSON, J. HARDY
Montréal

Acute degenerations with massive haemorrhage in pituitary tumors causing dramatic life threatening compression of the hypothalamus, bilateral cavernous sinus involvement and sudden loss of vision with subarachnoid haemorrhage have been reported episodically by numerous authors since BROUGHAM's classical description of the pituitary apoplexy in 1950. Besides these full blown apoplectic clinical pictures, degenerations with haemorrhagic cysts and necrotic changes are frequently discovered at surgery in pituitary adenomas, even in microadenomas.

Haemorrhagic and necrotic degenerations have been studied in a series of 664 pituitary adenomas between 1962 and 1979, including one autopsy-case. Apoplectiform degenerations were found in 9.6% of the cases (64 patients): these included 37 prolactin-secreting adenomas (12.4% from 299 cases), 8 growth hormone-secreting adenomas (4.7% from 171 cases), 18 endocrine inactive adenomas (12.5% from 144 cases) whereas there was one among 50 cases of ACTH-LPH-secreting adenomas (2%).

Classical "pituitary apoplexy" was present in only four instances and the remaining 60 cases were operative discoveries, including 21 large adenomas with suprasellar expansion and 39 moderate and small sized adenomas.

Pathophysiologically, the syndrome of "acute intra-adenomatous apoplexy of the pituitary" was in two instances related to disorders of the blood clotting mechanisms. In prolactinomas particularly, there seems to be a causative relationship between hormonal treatment, especially when this is stopped abruptly, and intratumoral degenerations. Radiotherapy also seems to favor acute morphological changes in the microvasculature of the pituitary adenomas.

III - B - 1

ROLE OF CIRCULATING IMMUNE COMPLEXES (CIC) IN MULTIPLE SCLEROSIS (MS) AND AMYOTROPHIC LATERAL SCLEROSIS (ALS)

F. SALINAS, H. COUSIN, G. EBERS, D. PATY
Vancouver

In view of the clinical significance and incomplete understanding of how CIC correlate with disease progression and prognosis in MS patients, we have detected CIC in 12/21 (57%) MS patients' sera, in 11/21 (52%) matched cerebrospinal fluid (CSF) samples, and in 4/13 (31%) ALS patients' sera by the Raji cell assay. The results expressed as μg aggregated human IgG equivalent/ml of serum (μg/ml) showed significantly different mean $\pm$ s.e. values of 29.2 ± 6.3 and 19.2 ± 3.4 μg/ml for the MS serum and CSF respectively as compared to 7.1 ± 1.5 for 44 normal control sera ($p \leq 0.00001$, Mann-Whitney test). 6/11 MS patients with elevated IC in CSF had normal serum levels, suggesting that in some instances the IC are arising from within the central nervous system. These findings support the concept that MS is due to an autoimmune process. The difference between ALS and normals was also significant (17.0 ± 4.3 μg/ml, $p \leq 0.01$, Mann-Whitney test). In addition, a preliminary serial study of one MS patient over 6 months showed an elevated CIC pattern with cyclical fluctuations. The 4/5 elevated levels were all during periods of clinical activity. Correlations are now being made between all samples tested and levels of clinical activity.

III - B - 2

PLASMA EXCHANGE IN CHRONIC DEMYELINATING NEUROPATHY

J. HUMPHREY, K. SHUMAK
Toronto

Chronic inflammatory neuropathies of the Guillain-Barre type are distinctive demyelinating disorders felt to be immunologically mediated. Recent reports indicate plasma exchange (plasmapheresis) may be of value in the treatment of this disorder.

6 patients (5 male, 1 female, age 19 to 54) have been studied. All had features consistent with a chronic demyelinating neuropathy. Three had a progressive course while 3 had two or more previous relapses of weakness. Four patients were on immunosuppressive drugs without improvement. Three received a single course of plasma exchange (10-15 l) while 3 have received two or more courses of exchanges.

Rapid improvement of weakness following plasma exchange occurred in 5 of 6 patients. Two of these 5 relapsed within 4 weeks while another relapsed on immunosuppressive drugs 3 months later. Improvement occurred following re-exchanging and has been maintained in all 5 with continued immunosuppressive therapy. Electromyographic and nerve conduction studies have been monitored throughout the course of all patients. Nerve conduction studies showed little change immediately after plasma exchange. Subsequent relapse was associated with a deterioration of nerve conduction.

Plasma exchange produced useful, but usually temporary improvement in these patients. Subsequent maintenance with immunosuppressive drugs or repeated courses of plasma exchange appear necessary to maintain improvement.

III - B - 3

THE PREDICTIVE VALUE OF CSF ELECTROPHORESIS IN "POSSIBLE" MULTIPLE SCLEROSIS

D. MOULIN, D. PATY, G. EBERS
Vancouver

The presence of oligoclonal banding (O.B.) on CSF electrophoresis has been well established in patients with clinically definite Multiple Sclerosis (MS). However, the demonstration of O.B. is of less value in patients with clinically definite MS than in the common clinical situation in which the diagnosis is in doubt.

We reviewed 211 patients with "possible" MS in whom CSF electrophoresis had been performed at the initial diagnostic evaluation (in most cases).

All these patients had objective evidence of neurological disease and the majority presented with either a chronic myelopathy or optic neuritis. Of 97 patients with O.B. followed for a mean of 28.2 months (median - 24 mo.), 9 patients developed clinically definite MS. Of 114 patients without O.B. followed for a mean of 27.7 months (median - 16 mo.), 8 patients developed clinically definite M.S.

These findings in patients with possible MS indicate that O.B. (in the absence of disseminated lesions) is not of high predictive value for the subsequent development of clinical MS, at least over this period of follow-up.

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III - B -4

CHRONIC NEUROPATHY ASSOCIATED WITH IgM ANTIBODY REACTIVE TO PERIPHERAL NERVE MYELIN

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Montréal

A 64 year old male with an acquired chronic demyelinative sensorimotor polyneuropathy and "benign" monoclonal IgM elevation, has been followed for 5 years. Severe stocking-glove sensory loss involves all modalities; motor weakness and atrophy are less prominent. CSF protein = 78 mg% with normal electrophoresis; nerve conductions have demonstrated progressive slowing of motor conduction to 1—2 meters/second. Sensory potentials are absent. Blink reflexes show markedly prolonged early and late responses without clinical facial involvement. Denervation has recently developed in distal muscles. Serial sural nerve biopsies show progressive demyelination and onion bulb formation. Repeated investigations have not revealed an occult malignancy, multiple myeloma, Waldenstrom's macroglobulinemia or amyloidosis.

Quantitative immunoglobulin measurements of IgG and IgA are normal; IgM is elevated at 1200 mg/dl. Other routine immunological tests are normal. Serum antibodies to human peripheral nerve myelin (HPNM) measured by microcomplement fixation (CF) assay gives a titer of 1:12,800. The IgM fraction CF titer is 1:160. the IgG CF titer is nil. After HPNM absorption of patient's serum there is disappearance of the monoclonal peak.

Steroid therapy was ineffective. Plasmapheresis resulted in a reduction of serum IgM. Long term treatment with plasmapheresis and chlorambucil has been instituted.

III - B - 5

HLA TYPING IN MS SIBLING PAIRS

G. EBERS, J. CRIPPS, D. PATY, P. SELAND, R. NELSON
London, Vancouver, Calgary, Ottawa

Several studies have shown an association between MS and the HLA antigens B7 and DR2 (and in some studies in DR3). However, other associations appear to exist in selected populations. Family studies in MS have not produced evidence for the tight HLA linkage expected from the population data. Accordingly, the nature of the MS-HLA association remains uncertain.

Forty unselected sibling pairs with Multiple Sclerosis from London (32) Calgary (4) and Ottawa (4) were typed for HLA—A, B, C, & D antigens. The diagnosis of MS was based on clinical criteria with laboratory support in almost all. Unequivocal genotypes were ascertained by typing parents and other family members as necessary. Sharing of haplotypes (none, one, or two) was compared to that expected by chance. Although more HLA identical sibling pairs were observed (12) than expected (9), there were 9 sibling pairs who shared no HLA antigens. In one of these, autopsy confirmation of the diagnosis was available in one sibling. In the others, at least one sibling had had either optic neuritis, internuclear ophthalmoplegia or oligoclonal banding of CSF. These findings fail to support a tight linkage of susceptibility to HLA, although epistatic interactions, polygenes and high gene frequencies either singly or in combination are considerations. The findings in these well-documented sib pairs are significantly different from some previously reported data. Possible explanations for the discrepancy include heterogeneity in etiology, although selection bias in other reported sib pairs cannot be excluded.

III - B - 6

MULTIFOCAL CT ENHANCEMENT IN MS

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London, Vancouver

Review was made of 69 high resolution CT Scans on 59 hospitalized MS patients. 46% (17/37) of those done during an acute relapse have shown enhancing lesions, characteristically in other areas than those suspected to be active on clinical grounds. With high volume delayed scans during acute exacerbations 13/20 (65%) showed enhancing lesions. In 34% (6/18) enhancing lesions were shown during "active stages" (viz. progression or relapse within 3 months) while only 15% (2/13) were positive during clinically inactive periods.

Twenty scans showed low density lesions. Ten of these also showed enhancement. Even though some of these lesions had been suggested by clinical symptoms, the majority had not been suspected. Characteristically, patients with enhancing lesions had more than one abnormality usually a mixture of at least one low density lesions and several enhancing ones.

This study suggests that enhancing lesions on CT Scan in MS are an indication of biological activity of the disease and are mort likely to be seen during acute clinically active relapses. Furthermore, the findings concur with the clinical impression that acute relapses are commonly multifocal and are consistent with the notion that acute relapses are mediated by blood-borne factors.

III - B - 7

USE OF TRIGEMINAL NERVE STIMULATION TO EVOKE SEP's IN MULTIPLE SCLEROSIS SUSPECTS

A. EISEN, S. PURVES
Vancouver

Physiologically it is possible to elicit somatosensory evoked potentials (SEP's) by stimulation of almost any mixed or cutaneous nerve. Trigeminal nerve stimulation used to evoke SEP's, although potentially useful, has been infrequently reported. In 12 normal subjects aged between 19 and 66 years (mean 39.8 years), SEP's were recorded subsequent to stimulation of the mouth. In each subject the right and left angle of the mouth was sequentially stimulated, recordings being made from C5 and C6 scalp locations (international 10 - 20 system) respectively. The resultant SEP had a negative-positive-negative configuration. These components respectively measured: N14 = 13.6 ± 1.3 msec; P20 = 19.8 ± 1.5 msec; N30 = 29.6 ± 2.4 msec. The interpeak difference given by (P20 - N14) was 6.4 ± 1.8 msec, whilst that given by (N30 - P20) was 9.9 ± 1.7 msec. Side to side differences of the peak latencies of N14, P20, N30, and (P20 - N14) and (N30 - P20) measured 1.2 ± 0.84 msec and was never greater than 2.6 msec. In 11 multiple sclerosis (MS) suspects (four with progressive spinal MS and seven with optic neuritis only), abnormalities were found in six (55%). This is a significantly higher diagnostic yield than the 40% or lower incidence reported for MS suspects when limb nerves are stimulated to evoke SEP's. Furthermore, abnormalities of trigeminal SEP's clearly indicate an additional lesion(s) to those considered responsible for either optic neuritis or progressive spinal MS. This technique might also prove useful in documenting objective dysfunction of trigeminal pathways in other diseases such as trigeminal neuralgia.

III - B - 8

SERIAL PATTERN VISUAL EVOKED RESPONSE STUDIES IN MULTIPLE SCLEROSIS

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Calgary

In patients with Multiple Sclerosis (M.S.), Pattern Visual Evoked Response (VER) studies were repeated in 30 eyes with a known delayed positive peak latency 1 year after their initial study. None of these patients had clinical attacks of optic neuritis in the 1 year interval.

VER studies were done under identical conditions 1 year apart using a Nicolet CA-1000 signal averages, and a check size subtending a visual angle of 27.5 minutes of arc.

Positive peak latency changed little in most eyes over the 1 year period. No latency shortened by more than 7 msec. Only 4 eyes had a prolongation of over 7 msec. These 4 eyes had a latency increase of 10.0, 13.0, 14.5, and 16.0 msec. No patient had such a latency increase in both eyes over the 1 year period.

Ten normal subjects (20 eyes) were studied in an identical manner on two occasions 1 year apart. The mean change in latency in the group over the 1 year period was 2.0 msec. (S.D. + 1.5 msec.).

When VER's were repeated 1 year later in M.S. patients with a known delayed positive peak latency, 87% of eyes showed little change in latency (7 msec. or less). A small number (13%) showed a latency prolongation of 10 msec. or more. These patients may have developed new asymptomatic optic nerve plaques over the 1 year period.

The VER may enable us to monitor asymptomatic new plaque formation in the visual pathways of patients with M.S. This may help to better quantitate disease activity and may be useful in therapeutic trials in M.S.

III - B - 9

HUMAN GASSERIAN GANGLION IN TISSUE CULTURE

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Edmonton

Human gasserian ganglia can be removed from cadavers and grown in tissue culture as explantation monolayers. The resulting explantation monolayers provide an in vitro model for the study of Herpes simplex virus (HSV) reactivation. Reactivation of HSV occurred in 7 of 9 (78%) cases when the ganglia were explanted within 12 hours of death onto 250 cm² culture area. When the monolayers were initiated within 12 hours of death but onto 125 cm² rather than 250 cm² culture area the recovery rate dropped to 9 of 21 (43%) cases, and if the monolayers were initiated on 250 cm² culture area but not until 13-24 hours after death then the HSV reactivation rate was apporximately the same at 5 of 10 (50%) of cases. Reactivation of HSV occurred in 0 of 18 cases explanted 13-24 hours after death onto a 125 cm² culture area.

An in vitro model of explantation monolayers of human trigeminal ganglia can be divided into 3 phases: an initial phase of cell recovery from day 1 through 7, a second phase of reactivating virus from day 8 through 42, and a third phase of uninducible virus from day 43 onward. HSV does not reactivate during the phase of cell recovery. The rate of HSV reactivation during the second phase depends upon how quickly the explantation monolayers were initiated after death of the patient and the surface area available for growth of the cells. During the third phase uninducible virus can be detected by genetic probing of the cultured cells with thermosensitive mutants of HSV. This in vitro model provides a basis for future research.

IV - A - 1

IMPAIRED INSULIN EFFECT ON THE FIBROBLAST SURFACE MEMBRANE IN MYOTONIC DYSTROPHY

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London

Myotonic dystrophy patients develop hyperglycemia and hyperinsulinism that earlier studies have shown are probably a response to insulin resistance. In order to determine the nature of the insulin resistance insulin receptor binding and the effect of insulin on the transport of 2-deoxyglucose (2-DOG) and α-aminoisobutyric acid (AIB) in cultured human skin fibroblasts from 10 myotonic dystrophy (MyD) patients and 10 age and sex-matched control subjects were investigated. The initial insulin binding capacity was decreased 62% from 5.04 ± 0.13% in the MyD group ($p<0.01$) due to a marked reduction in the receptor affinity, with no significant decrease in receptor number. Basal 2-DOG and AIB transport were the same in MyD and control fibroblasts but insulin-stimulated 2-DOG transport was decreased 56% from 11.34 ± 0.22 nmoles/mg protein/2 min in the control to 9.16 ± 0.18 nmoles in the MyD group ($p<0.01$). This was due to a reduction of insulin effect on the transport V_{max} (59%) with no significant decrease in transport K_m. Insulin-stimulated AIB transport was decreased 54% from 34.62 ± 0.70 nmoles/mg protein/20 min in the control to 27.80 ± 0.62nmoles in the MyD group ($p<0.01$). This also was a result of a reduction of insulin effect on the transport V_{max} (54%) with no significant decrease in transport K_m. The findings indicate that insulin resistance in MyD is caused by a reduction in the affinity of the insulin receptors and an associated glucose and amino acid cell surface membrane transport defect.

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IV - A - 2

PREVALENCE AND INCIDENCE OF MUSCULAR DYSTROPHY (MD) IN ALBERTA, CANADA

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Edmonton

Studies in other countries have focused on the prevalence and incidence of Duchenne MD rather than the less common limb-girdle and facio-scapulo-humeral dystrophies. This report describes the prevalence of four major forms of MD, including dystrophia myotonica, in Alberta, Canada on January 1, 1979. The rates per 100,000 were: Duchenne - 9.50; Becker II - 1.72; limb-girdle MD - 3.29; facio-scapulo-humeral MD - 1.52; and dystrophia myotonica - 5.61. Rates established for Duchenne, limb-girdle and facio-scapulo-humeral MD in Alberta in 1962 had all been lower.

The report also describes the incidence of each MD type during 1950-74. The overall Duchenne rate per 100,000 was 26.17. By five-year periods the rate increased gradually to peak at 35.43 during 1965-69, then declined to 19.37 in 1970-74. Part of the decline may be due to improved genetic counselling, part to cases not yet diagnosed. The incidence of Becker II MD was consistently low (overall rate—1.71), but indicated a decrease in each decade. By ten-year periods there were also consistent declines in the incidence of limb-girdle, facio-scapulo-humeral MD, and dystrophia myotonica; the overall 1950-74 rates per 100,000 for these MD's in Alberta were 2.77, 1.44 and 4.46 respectively. Increased prevalence, despite declining incidence, for each of the muscular dystrophies may indicate improved medical care and greater longevity for affected youngsters.

IV - A - 3

RECENT ADVANCES IN MENTAL RETARDATION THE FRAGILE X CHROMOSOME

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London

About 30% more males than females are affected with moderate or severe mental retardation. Gillian & Brian Turner from Australia suggested this was caused by X-linked retardation frequently associated with a marker X chromosome characterized by a fragile or attenuated site at the end of the long arm q. 27. This unusual chromosome only appears in leukocytes grown in primitive culture media and has been overlooked in a headlong drive to develop more sophisticated media for identification of individual chromosomes by banding techniques in the nineteen sixties and seventies.

Adults males with a fragile X chromosome may be easily recognized clinically by the large testes, this physical sign may be absent before puberty but the young children often have a high birth weight, large forehead, prominent jaw and big ears. Speech and language development is delayed. Seizures may be present in about half. Carrier females may be mildly retarded though typical stigmata are not described.

Five local families are presented to illustrate the importance of appropriate case finding and genetic counselling which carries the potential of preventing severe mental retardation and seizures in males and mild retardation in females.

The importance of the discovery of the fragile X ranks with the finding of trisomy 21 in Down's syndrome as a major step in mental retardation research.

IV - A - 4

X — LINKED HEREDITARY ATAXIC DIPLEGIA AND PELIZAEUS-MERZBACHER DISEASE: THE BORDERLAND REVISITED

H. DUNN, L. BECKER, M. THOMPSON
Vancouver, Toronto

In a previous report details of a family were given in which ataxic diplegia appeared to be inherited as a sex-linked and probably recessive condition occurring in 3 males in successive generations (Dunn et al. Canad. J. Neurol. Sci. 1974; 1:226). The ataxic diplegia was associated with mild to moderate mental retardation, congenital nystagmus and significantly small stature. While the nystagmus tended to subside, associated extra-pyramidal features were liable to become more marked with age. Three reports of a similar familial condition were found in the literature. Data concerning the Xa blood factor in the family suggested that the loci of genes for X and for the sex-linked disorder might be within calculable distance from each other on the X chromosome. No evidence of linkage with other X-linked disorders was demonstrable. It was concluded that the differential diagnosis between a form of cerebral palsy and Pelizaeus-Merzbacher disease could not be made with certainty in such a family in the absence of neuropathological findings.

In 1980 the oldest affected male in the family died at the age of 52 years. Autopsy showed patchy areas of demyelination in both cerebral hemispheres, particularly in the centrum semiovale and hippocampus, and also in the cerebellum and postero-lateral columns of the spinal cord, consistent with the diagnosis of Pelizaeus-Merzbacher disease. The surprising amount of myelin preservation in some areas suggested a very slow demyelinating process. Some neuronal loss and axonal degeneration were also noted, particularly in the end folium of Ammon's horn, and senile plaques were found in the cortex.

It is concluded that Pelizaeus-Merzbacher disease may progress so slowly as to mimic cerebral palsy.

IV - A - 5

MOTOR SIDE EFFECTS WITH PEMOLINE IN MINIMAL BRAIN DYSFUNCTION SYNDROME

H. DARWISH, H. MOGHADAM, M. FLINTOFF
Calgary

Psychostimulant drugs used in the treatment of minimal brain dysfunction have been reported to produce tics, chorea, & aggravation of the signs of the Gilles de la Tourette syndrome in children.

Pemoline has been shown, in earlier studies, to have a more sustained effect on the symptoms of MBD than shorter acting agents, such as methylphenidate. We studied 25 children in a double-blind design using performance on tasks measuring attention & learning, and parent ratings to assess efficacy of pemoline. Following a single dose (m = 1.72 mg/kg, SD 0.50), 10/25 children developed motor side effects. These consisted of choreiform movements of the upper limbs, buccolingual dyskinesia, & facial tics. In 2 children, the involvement of the limbs with choreatic movements was consistently unilateral. The mean onset of these side effects was 4.17 hrs (SD = 1.61 hrs) after administration of one dose of pemoline. These symptoms continued to worsen, reaching a peak at a mean of 10.50 hrs (SD = 3.16 hrs). Spontaneous resolution occurred at a mean 17.72 hrs (SD 6.96 hrs).

The mean dose of pemoline in 5 children with motor side effects considered favorable responders on learning and behavior ratings, was only 1.64 mg/kg. The mean dose was 2.09 mg/kg in 8 favorable responders without motor side effects. In the 5/25 favorable responders who developed motor side effects, a mean dose of 0.99 mg/kg (SD = 0.25, N = 5) was sufficient to maintain positive behavioral and learning performance without side effects. When the children who had previous treatment with methylphenidate were compared to those naive to psychostimulant on entry to the study, 5/18 patients with previous stimulant therapy developed motor side effects; 6/7 naive patients did. The probability of obtaining such results by chance are remote (P less than 0.01).

We conclude that some children are predisposed to pemoline motor side effects after a single dose administration. These resolve spontaneously, and do not recur on a lower dose, which preserves the positive behavioral response. The onset coincides with the expected peak blood levels and maximal effect on learning and behavior. The possible protective effect of previous methylphenidate medication was not expected. Chronic dopaminergic stimulation is usually thought to make dopamine receptors hypersensitive.

IV - A - 6

POST-HEMIPLEGIC DYSTONIA: A CLINICAL AND COMPUTERIZED TOMOGRAPHIC (CT) CORRELATIVE STUDY

J. GRIMES, M. HASSAN, A. QUARRINGTON
Ottawa

Correlation of movement disorders with specific basal ganglia lesions is difficult. Delayed onset dystonia is a rare occurrence following hemiplegia. Pathological studies have shown that this movement disorder requires major lesions in the caudate nucleus and putamen, secondary degeneration of the thalamus, and relative preservation of the corticospinal system.

Four patients with post-hemiplegie dystonia have been documented by clinical examinations, axial and coronal CT scans and videotape. One patient (age 69) had developed a sudden hemiplegia at age 9. The other patients (ages 32, 51 and 68) had had more recent cerebral infarctions. All patients had had severe motor and sensory deficits, but now show only a minimal hemiparesis. The dystonic movements began about 6 months after the hemiplegia in 2 cases, and after an indeterminable period in the others. The movements are marked in 3 patients and minimal in one.

CT scans illustrate unilateral low-density areas compatible with cerebral infarctions involving maximally the dorsal caudate nucleus and the putamen, with lesser involvement of the internal capsule and the anterior thalamus. 3 patients had minor unilateral cortical atrophy and one had moderate posterior frontal and insular atrophy.

Treatment in 2 patients included haloperidol, levodopa, benztropine and baclofen with no significant improvement. The study of these patients is unique in that it has allowed correlation in life of clinical signs with basal ganglia pathology.

IV - A - 7

ADRENOLEUCODYSTROPHY

P. WALSH, M. ERDOHAZI, L. BECKER
Toronto, London (U.K.)

Clinical and laboratory data from 24 patients with adrenoleukodystrophy (ALD) are reported. Only 3 of these cases have previously been documented in the literature as having ALD. The age of onset ranged from 1 year to 42 years. In 20 out of 24 cases the onset was between 4 and 10 years of age. Length of neurological illness ranged from 8 months to more than 8 years with an average duration of 3.04 years. No females were found to have ALD. Nine cases from 6 separate families had a family history of the disease. In 3 of these families the onset of neurological illness occurred at different ages. Two unusual family histories will be described. Presenting symptoms and neurological signs are detailed with emphasis on psychiatric manifestations. Only 3 types of laboratory tests in this series were useful. These were EEG's with evoked potentials, CT and isotope brain scans, and adrenal function tests. ACTH stimulation tests and serum ACTH levels were found to be very useful tests. The latter test appears more sensitive. Measurement of urinary hydroxycorticosteroids and/or a.m. and p.m. serum cortisols may be misleading. The relevance of brain and conjunctival biopsies, together with the role of steroid therapy will be discussed.

IV - A - 8

FURTHER STUDY OF DIETARY MIGRAINE

W. PRYSE-PHILLIPS
St. John's

In order to ascertain the influence of trigger factors in the precipitation of headaches, 42 patients with tension headaches and 250 with vascular headaches (the latter group including 95 with classic migraine) have been studied for periods of up to 8 years (mean 39 months). Headache categorization followed the guidelines of the ad hoc committee. Patients were required to search for food and other headache precipitants over a 6-8 week period after which a modified MAOI drug diet was prescribed, nitrites and glutamates also being reduced or forbidden.

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The frequency of headaches was assessed initially and repeatedly during the study period using patient diaries.

Highly significant differences between the responses of the tension and of the vascular groups were noted. Food precipitants of headaches were detected by less than 15 percent of the tension group but by three quarters of the vascular headache group. Satisfactory headache control (complete relief or a reduction in their frequency to less than 50 percent of the initial frequency, no prescribed drugs required) was attained in 7.2% of the tension group and 71.5% of the vascular group. It is concluded that, like mental stress, starvation, smoking, etc. a diet high in phenylalanine encourages noradrenaline production and platelet aggregation, with consequent intravascular release of vasoactive agents and arterial vasoconstriction with later changes in the vessel wall and hyperalgesia; and that dietary measures should be taken in all patients with vascular headaches before drug therapy is contemplated.

IV - B - 1

SUPRASELLAR ARACHNOID CYSTS: INVESTIGATION AND MANAGEMENT

H. HOFFMAN, E. HENDRICK, R. HUMPHREYS
Toronto

Since the advent of CT scanning in our institution in 1974 we have seen seven patients with suprasellar arachnoid cysts. These patients have been compared to the 41 patients with suprasellar arachnoid cysts that have been described in the literature.

Patients with suprasellar arachnoid cysts typically present with evidence of hydrocephalus, frequently have impaired visual acuity, and occasionally display the "bobble-head doll syndrome", isosexual precocious puberty and hypopituitarism.

The CT scan characteristically shows ventriculomegaly and the appearance of a cyst filling the anterior third ventricle which can easily be mistaken for a dilated third ventricle. Following this initial investigation, with the suspicion that a suprasellar cyst is likely present we have investigated our patients typically by initially inserting a *ventriculoperitoneal shunt and then injecting metrizamide* through the shunt reservoir into the ventricle and thus confirming the presence of a non-filling suprasellar cyst on a metrizamide CT scan.

Based on the experience in the literature and our own experience with these lesions we feel that these cysts if large are best exposed by the transcallosal route which allows one to easily unroof the cyst and allow free communication of the arachnoid cyst with the ventricular system thus effectively obliterating the cyst.

With the now ready availability of CT scanning in the evaluation of the child with hydrocephalus, suprasellar arachnoid cysts, although rare, will be found with increasing frequency. Since these suprasellar arachnoid cysts can mimic the appearance of a dilated third ventricle they can easily be confused with the far more common condition of congenital aqueduct stenosis. Treatment of such a patient by a by-pass VP shunt will drain one lateral ventricle only and thus lead to dilatation of the opposite lateral ventricle as well as the cyst.

IV - B - 2

CONTINUOUS INTRACRANIAL PRESSURE RECORDING AND INTRAVENTRICULAR INFUSION STUDIES IN NORMAL PRESSURE HYDROCEPHALUS

D. ROWED, C. EKONG
Toronto

Twenty patients with normal pressure hydrocephalus (NPH) were investigated with continuous intracranial pressure (ICP) monitoring and intraventricular infusion studies to attempt to determine who will benefit from cerebrospinal fluid shunting. They were then followed up clinically and with Neuropsychological assessment for at least six months and up to 2 years after shunting. Sixteen patients improved and 4 did not. The baseline intracranial pressure was within normal limits in all patients and not significantly higher in those that improved. Seventeen patients had frequent B waves and 14 of them improved after shunting. Three patients had infrequent B waves and *none of them improved. There were 3 patients with* frequent B waves who did not improve after shunting. Intraventricular infusion studies revealed less compliance and delayed return to baseline ICP among those who improved. The compliance of those who improved compared to those who did not will be discussed. We feel that continuous ICP monitoring and intraventricular infusion studies are helpful in determining which patient with NPH would benefit from shunting.

IV - B - 3

VENTRICULAR DRAINAGE FOR CEREBELLAR INFARCTION

C. POLYZOIDIS, M. KHAN
Saskatoon

Massive cerebellar infarction with progressive brain stem compression is a neurosurgical emergency. The treatment recommended is resection of the infarcted tissue to relieve brain stem compression.

We present three cases of massive cerebellar infarction, two of which developed acute brain stem compression with associated obstructive hydrocephalus. The third case did not develop hydrocephalus and improved on conservative therapy including Decadron. Both patients with acute hydrocephalus had a dramatic recovery with temporary external ventricular drainage and Decadron.

The incidence of brain stem compression and acute hydrocephalus with large cerebellar infarcts is high (75%) resulting in death if left without surgical intervention. However, the operative mortality following cerebellar decompression is also high (25-66%), probably because of the associated pathology, age of patient and magnitude of the surgery in a relatively moribund patient.

We seriously question the benefit of such major surgery, i.e. cerebellar decompression — in such patients and suggest it be preceded by external ventricular drainage. The subsequent course of the patient may determine the need for cerebellar decompression.

We propose that acute hydrocephalus complicating cerebellar infarction enhances cerebellar and brain stem ischemia and its early relief reduces the need for major surgery.

IV - B - 4

EXPERIENCE WITH LUMBOPERITONEAL SHUNTING

M. SCHWARTZ
Toronto

Twenty-two patients have undergone lumboperitoneal shunting using polyethylene tubing with an internal diameter of .047 inches. The tubing is introduced into the lumbar subarachoid space using a 14 gauge Touhey needle. Thirteen patients had idiopathic normal pressure hydrocephalus, five patients had hydrocephalus after a subarachnoid hemorrhage. The remaining four had diverse conditions: decompensated hydrocephalus after a posterior fossa decompression for Arnold Chiari Malformation and syringomyelia, hydrocephalus and a bulging bone flap after excision of a meningioma, aqueduct *stenosis and insufficient reabsorption following a Tor*kildsen shunt and CSF rhinorrhea following proton beam irradiation for an acidophil pituitary tumor. The average followup period was eighteen months. In thirty "shunt-years" there were seven blocked shunts. Blockage occurred on the average, ten months after insertion. Four patients complained of back pain and sciatica intermittently, but no patient developed weakness of the lower limbs nor lost myotatic reflexes. In contrast to the experience in children, lumbar arachnoiditis is not a problem. In the group with normal pressure hydrocephalus, nine of thirteen, or 69 percent had a good early response to shunting. In this group, however, six patients developed subdural hematomas, and one, hygromas that required surgical evacuation and removal of the shunt. There were no shunt infections.

From this experience I conclude that in the group with normal pressure hydrocephalus, at least, the use of valveless shunts is contraindicated. The rate of blockage is higher than in published series' describing other types of shunt. The infection rate in this series and in others is better than for other types of shunts. Silastic tubing with valves seems to offer the same ease of insertion and *effectiveness with a lower incidence of subdural hema*toma and perhaps fewer revisions.

IV - B - 5

MANAGEMENT OF SYRINGOMYELIA — A PATHPOHYSIOLOGICAL APPROACH

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London

Five varieties of syringomyelia can be differentiated. These include communicating, idiopathic or non-communicating, post-traumatic and those associated with arachnoiditis and tumour.

Thirty-six patients representing all varieties of syringomyelia have been treated with sixty-four surgical procedures. Seventeen patients required more than one procedure.

In fourteen cases of communicating syringomyelia, neurologic deterioration has been halted in thirteen (follow-up two months to twelve years). Foramen magnum decompression with IVth ventricular C.S.F. diversion was performed in twelve patients. Eight of these patients required additional syrinx cavity drainage procedures.

Seventeen patients had non-communicating, post-traumatic or post-arachnoiditis syringomyelia. Improvement resulted from drainage procedures (including syringo-subarachnoid and syringo-peritoneal shunts, and cordectomy) in fifteen of these seventeen patients.

Five cases of tumour-associated syringomyelia had improvement in their syringomyelic symptoms following resection (total in three, partial in two), of the tumour (follow-up two months to five years).

Complications of the sixty-four procedures included transient neurological deterioration (9 cases), increased upper limb pain (6 cases), and failure of maintenance of patency of drainage procedures (3 cases).

Modern neuroradiologic investigation including metrizamide myelography, delayed computerized spinal tomography with visualization of the cavity, and occasionally direct puncture, has remarkably improved our ability to understand the hydrodynamics of syringomyelia.

Based on this experience, we would recommend an aggressive surgical approach to syringomyelia. Identification of the etiology and pathogenesis of the syrinx should indicate the initial surgical procedure.

IV - B - 6

PERIPHERAL NERVE STIMULATION FOR RELIEF OF CHRONIC PAIN

R. VANDERLINDEN
Toronto

Electroanalgesia has become an established method of treating chronic pain. Since 1968, papers by Sweet, Long, Picaza, Hashold and Law have reported success in 60% of patients treated by implanted peripheral nerve stimulation after 2 year follow up.

Twelve patients have been implanted with peripheral nerve stimulators for treatment of chronic pain from post-traumatic neuropathy since 1971. The criteria for implantation were aching or burning pain in the distribution of a traumatized peripheral nerve that had failed to respond to physical therapy, transcutaneous stimulation sympathectomy and neurolysis. The cuffed electrodes were implanted proximal to the site of nerve damage on the following nerves: ulnar 4, median 4, sciatic 3 and brachial plexus 1. One patient has been lost to follow up and the single brachial plexus implant was removed on account of infection. Of the remaining 10 patients, 7 have experienced excellent pain relief requiring no analgesics, 2 patients report partial relief and one patient complains of aggravation of pain after two years of satisfactory relief.

The implantation of a peripheral nerve stimulator is a *simple procedure that is now probably the treatment of* choice for chronic pain due to post-traumatic neuropathy.

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IV - B - 7

CEREBRAL BLOOD FLOW (CBF) IN PROFOUND INTRA-OPERATIVE HYPOTENSION: CORRELATION WITH PRE- AND POST-OPERATIVE MEASUREMENTS

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London

We have examined the progression of changes in CBF and neurological status prior to, during, and following the use of profound hypotension (mean arterial blood pressure (MABP) 30-40 mmHg) at surgery in twelve patients with intracranial aneurysms. Intra-operatively, CBF was measured in the parietal region contralateral to the craniotomy using a modification of the 133-Xenon intravenous injection technique. Hypotension was induced by halothane alone, or a combination of halothane and sodium nitroprusside. Pre- and post-operatively, CBF was measured bilaterally using the 133-Xenon inhalation method.

Nine patients (Group I) demonstrated intact autoregulation intra-operatively. Mean CBF increased by 5 ± 6% (mean ± SEM) as MABP was reduced to 49 ± 2 mmHg. At extreme hypotension (39 ± 1 mmHg), average CBF was 79 ± 6% of control although three patients showed continued autoregulation to a MABP of 30 - 43 mmHg. Three patients (Group II) had impaired autoregulation and CBF was reduced to 51 ± 9% and 58 ± 8% of control flow at a MABP of 49 ± 1 and 40 ± 1 mmHg respectively.

Four patients in Group I and all Group II patients had both pre- and post-operative CBF studies. The patient groups were indistinguishable in terms of neurological status. Pre-operatively, the Group I patients had normal CBF (48 ± 1 ml/100g/min) and none had arterial spasm, whereas those in Group II had mildly reduced CBF (42 ± 3 ml/100g/min) and two had arterial spasm (unknown in the third patient). Post-operatively, mean CBF was reduced by approximately 3 - 5 ml/100g/min bilaterally in both groups of patients. Two Group II patients developed ischemic neurological deficits related to spasm in the post-operative period which improved with hypertensive therapy. Although three of the nine Group I patients developed post-operative spasm, there were no associated ischemic symptoms.

Based on the above data, we have concluded that severe reductions in CBF for brief periods are well-tolerated under halothane-induced hypotension intra-operatively. We were also able to identify a group of patients who showed impaired cerebrovascular reserve at all three stages of the investigation. These patients were at high risk of developing late-onset neurological deficits post-operatively. *Arterial spasm would appear to be a major* contributing factor to the observed reductions in cerebrovascular reserve.

IV - B - 8

INTRACRANIAL ENDODERMAL SINUS TUMORS

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Calgary

Endodermal sinus tumors (EDST) are a rare form of germ cell tumor but they have been recognized in the parapineal area with increasing frequency since Teilum identified their germ cell origin.

Four cases of histologically confirmed intracranial EDST have been treated in Southern Alberta in the last four years. Their ages range from 10 to 16 years. Two were parapituitary (both female); two were parapineal (both male). All four presented with headache and visual disturbance; two had the suggestion of growth retardation. All four had augmenting lesions on CT scanning; three had hydrocephalus. Serum alpha-fetoprotein was elevated in the two cases in which it was measured. Subtotal tumor extirpation was carried out in three cases; one had a VP shunt as the only primary surgical treatment.

One case displayed inexorable post-operative progression; sequential CT scanning revealed tumor spread throughout the intracranial cavity. He died three months after presentation. Three cases received whole brain radiation (5000 rads tumor dose). One received prophylactic spinal radiation and one intra-thecal methotrexate. They had clinical evidence of recurrence 2, 3 and 10 months after primary treatment and died at 5, 9 and 12 months after presentation. All three had histological confirmation of extracranial metastases — 2 spinal intradural, 1 hepatic. Post-mortem findings, results of immunoperoxidase staining and a review of the literature will be presented.

We conclude that the EDST is a very aggressive form of germ cell tumor, that intra and extra-cranial spread is to be anticipated and that alpha-fetoprotein measurements in serum and/or CSF are a valuable aid in the pre-histological diagnosis and post-treatment follow-up. Direct surgical extirpation of the tumor likely aids dissemination and may not be required for diagnosis if elevation of alpha-fetoprotein is demonstrated.

VI - A - 1

DIFFERENCES IN SEXUAL AND GENITAL ICTAL PATTERNS SUGGEST SEXUAL DIMORPHISM IN TEMPORAL LOBE EPILEPSY

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A sexual aura was encountered in nine patients with temporal lobe epilepsy and four also had a sensation of orgasm. Only one had sexual automatisms. All patients were female as were eleven others previously described. We have studied only one female patient with a genital but non-sexual aura.

Only three male patients with a sexual aura are mentioned in the literature and the descriptions unlike those of female patients are incomplete. By contrast we and other groups have found that nonsexual, often unpleasant, genital sensation as an aura is more frequent in males.

Apparent dimorphism of sexual mechanisms in limbic structures may explain these strongly diverging clinical manifestations in patients with partial complex seizures. Interictal behavioural trends such as increased frequency of hyposexuality and homosexuality in males with temporal lobe epilepsy may be related to the same underlying mechanisms.

VI - A - 2

VALPROIC ACID (VPA); STEADY-RATE PHARMACOKINETICS IN 43 PEDIATRIC *PATIENTS WITH SEIZURES*

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VPA is used commonly to treat seizures in children, yet little information exists on its pharmacokinetic behaviour in this age group. We present data from a study of 43 pediatric patients after steady-state VPA concentrations had been attained with daily doses of 15 - 10 mg/kg/day (Table I).

Table I
VPA STEADY-STATE PHARMACOKINETIC VARIABLES

	HALF-LIFE t½ (hrs)	ELIMINATION RATE Ke (hrs. - 1)	CLEARANCE (Cl) ml/min/kg	ml/min/m²	VOL OF DISTRIBUTION V_D (L/Kg)
$\bar{x}$	7 9 ± 2 4	0 095 ± 0 026	0 33 ± 0 15	8 4 ± 3 4	0 21 ± 0 06
Range	4 4 - 15 2	0 046 - 0 159	0 11 - 0 71	3 6 - 17 4	0 10 - 0 34

Cl was significantly higher in children receiving phenobarbital plus phenytoin than those not receiving these drugs (0.43 ± 0.16 ml/min/kg vs. 0.29 ± 0.13 ml/min/kg $p < 0.01$) as was Vd (0.25 ± 0.06 vs. 0.19 ± 0.05 1/kg $p < 0.001$). Cl, t½ Ke, and Vd were age dependent. (Table II).

Table II
AGE EFFECTS ON VPA PHARMACOKINETICS

	Age (years) 0.9 - 9 n = 27	10 - 20 n = 16	P Value
Cl (ml/min/kg)	0.39 ± 0.15	0.24 ± 0.07	< 0.001
t½ (hrs.)	7.2 ± 1.9	9.1 ± 2.6	< 0.02
Ke (hrs. -1)	0.104 ± 0.027	0.080 ± 0.018	< 0.01
Vd (1/Kg)	0.22 ± 0.06	0.18 ± 0.05	< 0.02

Peak and trough concentration ranges were 45-206 μg/ml and 13-101 μl/ml respectively, the variability in those concentrations emphasizes the need for specifying time of sampling in therapeutic monitoring. Our study also draws attention to: 1) dependence of pharmacokinetic variables on concomitant anticonvulsant therapy and age, 2) the wide interpatient variability in clearance. These pharmacokinetic differences may be important in the clinical management of epileptic children on VPA.

VI - A - 3

EFFECTS OF CINROMIDE, A NEW ANTICONVULSANT, ON CORTEX — AND AMYGDALA — KINDLED SEIZURES IN THE RAT

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Toronto

Recent experiments have indicated that "kindled" seizures in the rat are highly responsive to the anticonvulsant drugs clinically employed in the treatment of tonic-clonic and complex partial seizures. Of particular interest is the fact that focal as well as generalized seizure activity may be monitored in this model and that amygdala-focal activity, like human complex partial seizures has proved to be refractory to drug suppression. Cinromide is presently undergoing clinical testing in the treatment of various seizure types and efficacy is suggested. The present study reports the effects of the new anticonvulsant Cinromide (Vumide®) on: 1) amygdala-focal seizure activity; 2) cortex-focal seizure activity; and 3) generalized consulsive seizures triggered from either focus. Dose-response experiments were conducted in 20 rats with well established cortical (10 animals) or amygdala (10 animals) seizures. The data indicate that Cinromide, like anticonvulsants previously tested, is more potent against generalized seizure activity ($ED_{50} \simeq 80$ mg/kg, regardless of the focus of origin). The doses required to suppress focal activity were higher and were often associated with measurable levels of toxicity. Cinromide, however, appeared to be relatively more effective against focal activity, than some of the currently used anticonvulsants (eg. phenytoin, phenobarbital). The results suggest that Cinromide should be clinically useful against tonic-clonic seizures. At higher (and possibly toxic) doses it should also be active against partial seizures.

VI - A - 4

EWALD'S SECOND LAW AS A BASIS FOR THE LOCALIZATION OF UNILATERAL LABYRINTHINE LOSS IN HUMANS

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Ewald postulated that, ampullopetal endolymph flow in the lateral semicircular canal is a more efficient stimulus than ampullofugal flow (1892). The confirmation of this theory by Goldberg and Fernandez's (1971) primary canal afferent data and by Baloh, Honrubia and Konrad's (1977) observation of rotatory nystagmus, suggested to us that normal compensatory eye movements during head movements should be asymmetrical for patients with unilateral labyrinthe loss and furthermore, that this asymmetry could indicate the side of a peripheral lesion in the course of our new diagnostic quantification technique for the vertibulo-ocular reflex (VOR).

The VOR of patients with complete unilateral labyrinthectomies was measured using sinusoidal stimulation (0.5 - 3 Hz); 1) in darkness, 2) when the patient suppressed the VOR while fixating a target moving with him and 3) while the patient fixated a stationary target, thus supplementing the VOR with visual tracking. Marked asymmetrical responses towards the labyrinthectomized side were observed under all conditions. The surprising low frequency asymmetry during visuo-vestibular interaction indicates an insufficiency of the visual input to generate compensatory eye movements, a conclusion which is in agreement with Baarsma and Collewijn's work (1975) on rabbits. In all the patients tested, fixation suppression gains were less than normal gain values (Istl, Hyden and Schwarz, 1981). It may be concluded that, asymmetry according to Ewald's second law permits side detection of unilateral labyrinthine loss.

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VI - A - 5

LATERAL DECUBITUS AND SUBSIDENCE OF SLEEP-INDUCED APNEAS (SIA): THERAPEUTIC IMPLICATIONS

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Diet, pharmacologic agents and tracheostomy have been used with variable success in the treatment of S.I.A. In the search for an effective, simpler and less invasive treatment of this condition, our own observations reported elsewhere (Dorsal and lateral decubitus in detecting and abolishing S.I.A., Eastern American EEG meeting, Feb. 81, Ste-Adèle) might be of relevance.

The present study was undertaken to assess how consistenly and to what extent apneas and symptoms of S.I.A. could be effectively modified by strict avoidance of dorsal decubitus.

In a consecutive series of 21 patients with S.I.A., repeated daytime or all-night polysomnograms were obtained in both dorsal and 50° lateral decubitus positions.

Apneas, best detected in dorsal decubitus, subsided immediately with lateral decubitus in all patients but one. The effect of lateral decubitus in S.I.A. was documented repeatedly throughout the follow-up period. However, even after years of sleeping in lateral decubitus, apneas immediately reappear upon resuming the dorsal decubitus position.

14 patients have complied with instruction to sleep in lateral decubitus for 6 months or more. All 14 have reported reduction in snoring and daytime sleepiness.

Despite some inconvenience, sleeping in lateral decubitus have obvious advantages over the current forms of treatment; furthermore, the fact that it is immediately effective for central apneas as well as for obstructive and mixed apneas may shed light on the physiopathology of S.I.A.

VI - A - 6

SLEEP APNEAS AND IMPAIRED PERIPHERAL CHEMOSENSITIVITY IN AN ADULT PATIENT WITH HINDBRAIN MALFORMATION

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A 45 year old patient presented with syncope. Symptoms suggestive of sleep apnea syndrome were elicited. Bilateral lower cranial nerves signs were conspicuous. Mild hypoxemia and hypercapnea were observed at rest and made worse by exercise. Radiological studies, then surgery, disclosed Chiari malformation, some degree of basilar impression and a Dandy Walker type of cyst. All night and daytime polysomnograms were done before, then 3 weeks, 4 and 10 months after surgery. Initial recording showed an apneic index of 60 with a percentage of sleep time spent in apnea of 66%. The apneas were predominantly of the obstructive type. Three weeks after sugery, apneas were much less frequent; after 10 months, they were no longer seen. Respiratory response measured before, one and four months after surgery, was normal to hypercapnea from the beginning and remained so. On the other hand, it was absent to hypoxemia preoperatively and improved stepwise on subsequent studies. Medroxy-progesterone was given orally during 10 days before operation. It did not improve either the apneic index or the impaired chemosensitivity.

Sleep apneas have been unfrequently documented in adults with hindbrain malformation before surgery, their precise mechanism remains unclear. The blunted response to hypoxemia in this case probably results from distorsion in afferent pathways for hypoxic drive in the ninth cranial nerves in agreement with conclusion from one previous report.

VI - A - 7

RESPIRATORY INHIBITORY APRAXIA: CLINICAL AND COMPUTERIZED TOMOGRAPHIC STUDY

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Ottawa

Respiratory Inhibitory Apraxia (R.I.A.), the inability to stop breathing on command, was previously described by Atack and Suranyi, 1975. On the basis of clinical assessment, technetium brain scan and angiography as well as one autopsy examination, they concluded that this entity was always secondary to a deep-seated minor hemisphere lesion. We dedided to further study R.I.A. with a view to a clear definition of the clinical picture, the natural history and the anatomical localization with the assistance of C.T. Scanning.

All patients seen by the authors in a 14-month period with a diagnosis of stroke were thoroughly assessed. 15 patients were identified as having R.I.A. over the 14-month period. These patients were all right-handed and once again as in our original study all the patients had a minor hemisphere lesion with left hemiplegia or hemiparesis. No patients with major hemisphere lesions had R.I.A. Of the patients with R.I.A., 13 had an ischaemic lesion with infarction and 2 had a hemorrhagic lesion. 13 had motor impersistence, 11 had anosagnosia, 10 had left homonymous hemianopia, 10 had cortical sensory impairment in the left extremities, 10 had parietal lobe inattention of the left, 7 had impairment of pain sensation on the left side. Follow-up was possible in 13 of the 15 cases with R.I.A. and 12 developed impersistence for respiratory inhibition before gradual return to normal ability to inhibit respiration. One patient whose stroke had occurred 7 years before continued to have R.I.A. 7 years after this was first observed. C.T. Scanning was carried out on all patients and in each case showed a right hemisphere lesion involving the internal capsule or the deep part of the corona radiata. We postulate, therefore, that the essential lesion in these cases is deep-seated in or very near the internal capsule and that this lesion has disconnected fibres projected from the cortical centres for learned inhibition of respiration to the brain stem respiratory centres. We feel that Respiratory Inhibitory Apraxia is an example of the disconnection of an inborn movement pattern from cortical centres which are capable of influencing it voluntarily, in this case in regard to inhibition. It would seem that the minor hemisphere has an important role to play in this regard.

VI - A - 8

THE BLOOD BRAIN BARRIER AND FOLATE DEFICIENCY

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Montréal

On the basis of animal experiments, it has been postulated that even in severe folate deficiency, folate levels in the "brain are well maintained" (Spector and Lorenzo, 1975).

Twenty-three patients (average age: 63.2 ± 14.9 (x ± SEM) ranging from 28 to 83 years) with low or borderline cerebrospinal fluid (CSF) folate and low serum folate underwent two lumbar punctures, i.e. before and after three weeks of folate therapy.

The rise in CSF folate content in the whole group after replacement therapy was significant by both the *Lactobacillus casei* ($p < 0.05$) and radioisotope ($p < 0.005$) methods of folate determination. In patients with folate-responsive neurological disorders, the rise of CSF values after replacement therapy was definitely higher ($p < 0.07$ and $p < 0.025$ respectively) than in the group of patients in whom folate-deficiency was not related to the actual clinical picture.

A comparison has also been made between the rise in CSF folate in those subgroups of patients with less *versus* those with more than 12 ng/ml respectively, value which can be considered as definitely low. There was a statistically significant rise ($p < 0.001$) in folate content in the subgroup with low initial CSF folate values.

Contrary to previous statements, the present study revealed that there is not an absolute blood-brain barrier for folate because the lower the CSF folate level was, the more rapid and spectacular the increase in CSF folate after replacement therapy was found.

VI - B - 1

PORPHYRIN NEUROTOXICITY IN VITRO

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Neurological abnormalities characterize the clinical presentation of the acute intermittent and variegate forms of hepatic porphyria. It is commonly thought that the abnormalities are caused by the porphyrin precursors delta-aminolevulinic acid and/or porphobilinogen. However, Sima et al (Can. J. Neurol. Sci., in press, 1981) demonstrated that various porphyrins as well as delta-aminolevulinic acid can produce a dose-dependent decrease in motor nerve conduction velocities when injected into mice, and that severe axonal abnormalities accompanied by myelin breakdown may develop several weeks following the injection of certain synthetic porphyrins.

We present evidence that naturally occurring and synthetic porphyrins can produce dose-dependent toxicity to neurons *in vitro*. The assay for toxicity is based upon a single neuronal cell biological assay for Nerve Growth Factor (NGF) modified by Riopelle and Cameron (J. Neurobiol., in press, 1981). NGF produces a dose-dependent neurite outgrowth response from chick embryo sensory neurons *in vitro*. In the presence of porphyrin, there is a dose-dependent inhibition of NGF-induced neurite outgrowth with frank neuronal killing at high concentrations. In the micromolar range, the naturally occurring porphyrins protoporphyrin IX, coproporphyrin I, and uroporphyrin I suppress NGF-induced neurite outgrowth by approximately the same degree if comparison is made on a molar basis. However, the porphyrin precursor delta-aminolevulinic acid is not neurotoxic up to millimolar concentrations. The synthetic porphyrins tetraphenylporphine sulfonate (TPPS) and hematoporphyrin derivative (HpD) are also neurotoxic.

These *in vitro* experiments support and extend the *in vivo* observations of Sima et al (1981). Since delta-aminolevulinic acid rapidly forms porphyrins when injected *in vivo*, but does not do so in the neuroblast culture system, we therefore suggest that the abnormalities of neuronal function characteristic of patients with porphyria are produced by the direct action of naturally occurring porphyrins on neurons, rather than by the porphyrin precursor delta-aminolevulinic acid. Finally, this *in vitro* assay system may prove useful for rapid quantitative screening for neurotoxicity of chemotherapeutic agents and radiation sensitizers of potential clinical value. Both TPPS and HpD are used as sensitizers in photoradiation therapy of cancer.

VI - B - 2

INTRACEREBRAL ARTERIOLAR PERMEABILITY TO LANTHANUM

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Kingston

Our previous studies of angiotensin-induced acute hypertension demonstrated that focal segments of intracerebral arterioles situated in the second and third layers of the temporal and parietal rat cortex develop increased permeability to horseradish peroxidase (HRP). In the present study the morphology of these vascular segments in control and acutely hypertensive animals was further studied using colloidal lanthanum solutions as a marker.

Hypertension was induced in Wistar-Furth rats using angiotensin II (Ciba) infused at the rate of 20 μg/minute for 2 minutes. Animals were perfusion fixed 3 minutes after the onset of hypertension following which colloidal lanthanum solutions were perfused for periods varying from 5 - 40 minutes. Control animals were subjected to the same experimental procedures as the hypertensive animals.

Lanthaum was demonstrable in arteriolar walls and the surrounding brain of control animals circulated with this tracer for periods varying from 12 - 40 minutes. The two principal mechanisms associated with tracer extravasation into the brain was diffuse passage through endothelial cytoplasm and via interendothelial spaces bypassing tight junctions. The latter finding has not been previously reported and suggests that the tight junctions between endothelium of intracerebral arterioles are a meshwork of closely arranged maculae occludentes rather than, complete circumferential occluding bands as was previously believed.

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In hypertensive animals lanthanum was demonstrable in arteriolar walls, and the surrounding neuropil following only 5 minutes of circulation. Passage of tracer through vessel walls occurred by the same routes as observed in controls. Numerous pinocytotic vesicles were also observed and those in contact with the luminal or abluminal plasma membrane were labelled with tracer confirming our previous studies using HRP that increased vesicular transport occurs in cerebral arteriolar endothelium in acute hypertension.

VI - B - 3

EXPERIMENTAL ELONGATION OF MAMMALIAN SPINAL AND MEDULLARY NEURONS

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Montréal

The elongation of axons from spinal and medullary neurons was studied using a new experimental model which employed PNS grafts as "bridges" to connect the spinal cord and the brain stem. In a series of adult Sprague-Dawley rats an autologous segment of sciatic nerve (3.5 cms long) was grafted so that one end of the graft was inserted into the dorsolateral medulla and the other was introduced into the dorsolateral region of the upper thoracic spinal cord. The span of these "bridges" was placed extraspinally. Light and EM examination 7 - 9 months after surgery showed the grafts to be well innervated by Schwann cell-ensheathed axons that had traversed the 3.5 cm long PNS "bridges". The location of the cell bodies and terminal arborizations of these axons was determined by transecting the regenerated grafts and applying horseradish peroxidase to the cut ends. After allowing for retrograde and anterograde transport of the enzyme, frozen sections of the medulla and spinal cord were incubated in TMB and H_2O_2. Retrogradely labelled neurons were found in the medulla and spinal cord near the sites of insertion of the graft. Anterogradely labelled fibers coursing within the graft were shown to penetrate the CNS for approximately 1 mm.

There results indicate that following CNS injury a conducive glial environment does allow spinal and medullary neurons to elongate axons for distances that are often greater than those that they usually occupy in the intact animal. Thus, the success or failure of regeneration in the mammalian CNS may depend largely on changes in the glial environment rather than on intrinsic properties of the neurons themselves.

This new animal model may also help to devise ways for the experimental reconstruction of certain neuronal circuits in the damaged spinal cord.

VI - B - 4

AXONS FROM NEURONS IN THE CEREBRAL HEMISPHERES OF ADULT RATS GROW IN PNS GRAFTS

M. BENFEY, A. AGUAYO
Montréal

These experiments were carried out to determine if axons from neurons in the cerebral hemisphere of adult rats are capable of elongation after focal injury to the brain and introduction of a peripheral nerve graft. An approximately 2 cm long autologous segment from the sciatic nerve was introduced through an opening in the skull overlying the lateral surface of the hemisphere and advanced into the region of the internal capsule and basal ganglia. The other end of the nerve graft was sutured to the temporalis muscles. Approximately two months after grafting, the external (intramuscular) stump of the graft was re-exposed and horseradish peroxidase (HRP) was applied locally to retrogradely labelled neurons innervating the graft. After two days each animal was killed and cryostat sections (20 μm) of the brain were obtained and examined. HRP-labelled nerve cells were found in different regions of the basal ganglia and also in the cortex neighbouring the graft.

Thus, this type of graft arrangement: i) results in the unidirectional growth of axons from central neurons towards muscle, ii) permits the use of the distal end of the nerve graft for the application of the HRP tracer and, iii) has demonstrated that in response to injury the neurons within the hemisphere itself are capable of axonal elongation when their CNS glial environment is substituted by one which resembles that normally found in the PNS.

VI - B - 5

INTRACRANIAL INTERNAL CAROTID ARTERY STENOSIS

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London

There has been no report dealing with the natural history and prognosis of atherosclerotic intracranial internal carotid artery (ICA) stenosis. Although extracranial-intracranial arterial bypass has been performed for this condition, the results cannot be evaluated adequately because of lack of controls. A study has been initiated to determine the prognosis of symptomatic intracranial ICA stenosis. This report concerns a follow-up of 21 patients averaging 31 months. These 21 patients were admitted to University Hospital, London during the 8-year period (1972-1980). Fifteen patients presented with a stroke in the appropriate cerebral hemisphere with or without associated TIA's. Six patients presented with TIA alone including 3 patients with amaurosis fugax. Cerebral angiography revealed a lesion at the supraclinoid portion in 9, cavernous portion in 10, pre-cavernous portion in 4 and petrous portion in 1. Six patients had a moderate stenosis of the ICA at the origin as well as the intracranial lesion, two of whom underwent carotid endarterectomy. Both patients continued to experience ischemic events postoperatively. Five patients had repeated angiograms after 7-26 months and in all of them, progression of disease was demonstrated as either increased stenosis or irregularity. In one patient, significant stenosis went on to complete occlusion in 7 months. Sixteen patients were on platelet anti-aggregants, 3 on anticoagulants and 2 patients received neither. Eight patients (38%) experienced a persisting ischemic event in the territory supplied by the stenotic ICA and 3 patients (14%) had recurrent TIA's in the same vascular territory. TIA's or strokes in other vascular distribution occurred in 4 patients. Six patients were free from any cerebrovascular events. Eight patients died during the follow-up period. The causes of death include 3 myocardial infarctions, 1 pulmonary embolus, 1 heart failure, 1 ruptured aortic aneurysm, 1 possible brain stem stroke and 1 brain stem stroke as an angiographic complication.

These results indicate that intracranial ICA stenosis is a very serious lesion in terms of the recurrence of cerebrovascular events as well as a risk factor for other systemic vascular disease. This prognostic study will continue. The question of whether surgical treatment is beneficial still remains to be answered in the ongoing collaborative EC/IC Bypass Study.

VI - B - 6

OXYGENATED FLUOROCARBON PERFUSION AS TREATMENT OF ACUTE SPINAL CORD COMPRESSION INJURY

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Hamilton, Cleveland

Experiments were conducted to determine the relative therapeutic value of durotomy only, saline perfusion or perfusion with oxygenated fluorocarbon following acute spinal cord compression injury.

Unlesioned control dogs which had durotomy, subarachnoid catheter placement and saline irrigation for 4 hours did not have any significant lasting neurological deficit.

A series of 41 dogs received an acute spinal cord lesion using an epidural balloon inflated to a pressure of 160 mm of mercury and maintained for 1 hour.

Perfusion with either saline or oxygenated fluorocarbon at room temperature significantly improved motor function recovery over dogs which received a durotomy only. Perfusion with oxygenated fluorocarbon improved motor recovery significantly better than perfusion with normal saline.

The possible mechanisms underlying the beneficial effects of perfusion with either perfusate are discussed.

VI - B - 7

FURTHER OBSERVATIONS ON THE ROLE OF BARBITURATE PROTECTION IN GLOBAL CEREBRAL ISCHEMIA IN THE MONGOLIAN GERBIL

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Vancouver

Previous work in our laboratory using the Mongolian gerbil, failed to show unequivocal protection by barbiturates to an insult of 15 minutes of global cerebral ischemia. This could possibly have been due to the barbiturate induced hypotension.

To clarify this issue, a further series of animals was studied with ventilatory and circulatory support. After anaesthetic induction with 50 mg/Kg intramuscular ketamine, the animals underwent tracheostomy and controlled ventilation on a Harvard respirator. Femoral intra-arterial and intravenous lines were inserted for blood pressure monitoring and support with aramine.

The experimental animals were divided into 2 groups: Group A (n = 12), received 50 mg/Kg of nembutal intraperitoneally either 2-15 minutes before (n = 6) or 15 minutes after (n = 6) reversible carotid occlusion of 15 minutes duration. Group B (n = 6) received 25 mg/Kg of thiopental intravenously either 15 minutes before (n = 3) or 15 minutes after (n = 3) the same ischemic insult. Adequate controls (n = 6) were included. After 4 hours of recirculation the animals were sacrificed by perfusion-fixation.

Detailed histological examination did not reveal any protection in either of the barbiturate groups, regardless of the time of administration.

We conclude that there is no evidence to support the efficacy of barbiturates in global ischemia.

VI - B - 8

AN ASSESSMENT OF SUPERFRACTIONATION RADIOTHERAPY IN A MURINE MODEL SYSTEM AND A PROSPECTIVE CLINICAL TRIAL

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Calgary

Superfractionation (SF) radiotherapy is the administration of more than one fraction per day. We have assessed the effect of SF radiotherapy in a murine brain tumor model and in a prospective, randomized clinical study of malignant astrocytomas.

C57B1/6J mice with subcutaneous tumor were irradiated to a total of 800 ret(NSD). They received 5 or 15 fractions in 5 days, 10 or 30 fractions in 12 days and 15 or 45 fractions in 19 days. The effect of the radiation therapy on survival fraction was assessed by the end-point dilution assay method of Hewitt. In the conventional fractionation (CF) groups, the survival fractions were 0.263, 0.182 and 0.132 respectively. In the SF groups, the survival fractions were 0.148, 0.0279 and 0.00629, respectively. The % decrease in survival fraction for 5 versus 15, 10 versus 30, and 15 versus 45 fractions delivering the same NSD(800 rets) was 78%, 552% and 1999%, respectively. We conclude that SF is superior to CF in this model system.

67 patients with intracerebral malignant astrocytomas or glioblastoma multiforme were entered into a prospective randomized clinical trial from January 1979, to December 1980. 33 patients were randomized to CF (3400 rads in 17 fractions in 3.5 weeks, whole brain and 1600 rads in 8 fractions in 1.5 weeks, local boost, NSD 1573 ret). 34 were randomized to SF (4000 rads in 45 fractions in 3 weeks whole brain and 1000 rads in 5 fractions in 1 week, local boost, 1276 ret). Age, pathology, tumor location and size, degree of surgical removal, performance status and steroid requirement were comparable in the two arms. Both arms received a course of CCNU. The one and two year actuarial survival rates for the CF group was 41% and 9%, respectively. A trend suggesting the superiority of SF radiation has been identified and merits further elucidation.

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P - 1

ADULT MEDULLOBLASTOMA

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Calgary

Medulloblastomas are highly radiosensitive tumors which occur most commonly in a midline cerebellar or vermal location, in the pediatric age group. They are unusual in adults. We have encountered five adults with medulloblastoma in the course of the last decade. Their ages range from 22 to 45 years. All five were male. All presented with headache and gait disturbance. Primary treatment in each case consisted of suboccipital craniectomy with subtotal tumor extirpation followed by total neuraxial radiation.

Two are alive and neurologically well 6½ and 4 years after treatment. Three have histologically confirmed recurrence. One recurred as a spinal intradural tumor three years after primary treatment. One recurred in the posterior fossa after 3¾ years. One recurred in the posterior fossa 4½ years after primary treatment; he died six months after the posterior fossa recurrence with diffuse spinal intradural spread. Radiographic features and histology will be reviewed.

It has been suggested that adult medulloblastomas are less aggressive than their pediatric counterpart. Our experience suggests that medulloblastoma in the adult is an aggressive tumor with a propensity for recurrence.

P - 2

THE CHALLENGE OF INTERNATIONAL NEUROSURGICAL EDUCATION

D. FAIRHOLM
Taipei, Taiwan

The personal perspectives and experiences of a Canadian neuro-surgeon working in Taiwan will be used to suggest ways in which Canadians can help the developing world overcome its problems through education.

P - 3

SIMULTANEOUS EPIDURAL AND CISTERNAL PRESSURE MEASUREMENTS IN DOGS

L. IVAN, S. CHOO
Ontario

To assess the accuracy and reliability of the Ladd epidural sensor, simultaneous eipdural and cisternal pressure measurements were done in eight dogs. All animals were anesthetized with I.V. Nembutal, ventilated with a volume controlled respirator. Through a small craniectomy, the sensor was inserted in the epidural space connected to a Ladd monitor and the pressure was recorded with a multi-speed Omni-scribe recorder. Through a guage 20 needle, and using a Statham transducer, the cisternal pressure was monitored on a Grass polygraph.

In order to compare different pressure levels, the intracranial volume was increased by injecting 1 ml of physiological saline into the cisterna magna every 5 minutes, until a total of 8 ml fluid was infused. The correlation between the epidural and cisternal pressure was established statistically using the least squares method as well as plotting the two pressures against each other at corresponding time intervals during the decay of the peak pressure response. The results show that the slope of the regression lines lies between 0.82 — 1.18 with an intercept in the range of 0.1 to -7.2. The correlation coefficient lies between 0.96 to 1 indicating a high degree of correlation between epidural and cisternal pressure. A slight time lag was noted in the development of the epidural peak pressure, the significance of which will be discussed.

P - 4

POST SURGICAL EXTRADURAL SPINAL CYSTS

B. BENOIT, N. RUSSELL
Ottawa

The dura mater is occasionally traumatized during surgery but the literature makes scant reference to the subsequent occurrence of extradural cysts. Our recent experience with such a case, presenting one year after lumbar discectomy, led to a review of the subject.

Two types of post surgical extradural cysts have been described. One type represents a true diverticulum of arachnoid, which herniates through a dural opening and gradually enlarges. Symptoms result from compression, distortion or entrapment of adjacent neural structures. The diagnosis is suggested by the development of progressive neurological symptoms beginning some months after a spinal operation. These may mimic recurrence of the original lesion. In the other type, the arachnoid has also been breeched and a fistulous connection exists between the subarachnoid space and paraspinal tissues. These manifest themselves early in the post operative period, the cardinal signs being postural headache and a fluctuant mass, which can be seen or felt beneath the surgical scar.

The first type requires myelography to confirm the diagnosis. Treatment of each consists of closure of the dural defect and excision of the cyst.

P - 5

INTRADURAL EXTRAMEDULLARY SPINAL METASTASIS

R. PERRIN, K. LIVINGSTON
Toronto

Intradural extramedullary spinal metastasis is infrequently encountered and rarely reported. We have encountered six such cases among 150 neurosurgically treated patients with symptomatic spinal metastasis.

Four men and two women, ranging in age from 39 to 65 years (mean 54 years) each presented with a characteristic clinical syndrome including pain, weakness and sensory loss. The site of predilection for intradural extramedullary metastasis was L2-3 (four cases). Culpable primaries included lung (2 cases), melanoma (2 cases), breast and Hodgkins. Following surgical decompression 50% reported some pain relief and all patients were ambulatory.

Cerebral metastases declared themselves in three cases, three to six months following spinal decompression.

Intradural extramedullary spinal metastasis confirmed at surgical decompression may be an indication for post-operative systemic chemotherapy and/or total neuraxis radiation.

P - 6

UNIVERSITY OF TORONTO HEAD INJURY STUDY

M. SCHWARTZ
Toronto

Head injuries are a major cause of death and disability in Canada today. Some patients die or are disabled because of primary mechanical damage to the brain on impact, but many patients develop increased intracranial pressure as a result of cerebral swelling that is severe enough by itself to cause death. The best method of treatment for increased intracranial pressure is unknown. The major drugs are in use at the present time are mannitol and pentobarbital. To *determine which is more effective, half of our patients,* chosen by random assignment, are given mannitol initially. If this fails to relieve the intracranial pressure, then they immediately receive pentobarbital. The other patients receive pentobarbital initially, but are immediately given mannitol as well if the pentobarbital is inadequate to control increased intracranial pressure. Only patients with intracranial measuring devices and intracranial hypertension are included in the study. All patients are mechanically ventilated. The administration of both mannitol and pentobarbital are governed rigidly by a protocol, as are the other aspects of the patient's care.

In the first seven months of operation, eight patients have received mannitol as their initial therapy. Three of the eight patients required barbiturates as well for the control of intractable intracranial hypertension. Four of the eight patients survived.

Twelve patients have received pentobarbital as their primary drug. Of these, nine required mannitol in addition to pentobarbital for the control of intractable intracranial hypertension. Five of these twelve patients have survived.

Adherence to the protocol seems good. Those patients receiving pentobarbital as the initial treatment require mannitol in addition to the barbiturate for the control of intracranial hypertension more frequently than the patients who get mannitol as the first treatment require *barbiturate. At this point, despite randomization, the two* groups are not perfectly comparable. On the average, the pentobarbital patients were seven years younger than those who received mannitol but had, on the average, a slightly lower Glasgow Coma score on admission. It is estimated that a total of one hundred patients will be required before firm conclusions can be drawn. Hence, it is wise to mistrust nonrandomized trials that have at best historical controls.

P - 7

ATLANTO-AXIAL TRAUMA WITH SPINAL CORD INJURY

M. SCHWARTZ
Toronto

It is a widespread misconception that patients with atlanto-axial fractures either die of their injury or survive with the spinal cord intact. In the past five years at Sunnybrook Medical Centre, among more than forty patients with atlanto-axial fractures, we have treated eight patients, or roughly twenty percent of the total who suffered partial spinal cord injuries. All had transient tetraplegia. One patient, a sixty-five year old woman with ankylosing spondylitis, had a mild hemiparesis affecting her arm and leg equally, without sensory loss. The other seven patients with partial injuries illustrated a characteristic pattern. In each case, whether the patient had a fixed tetraparesis or a fixed hemiparesis, the arm was always weaker than the corresponding leg. There were no cases of cruciate palsy with one arm and the contralateral leg affected, although one might predict such a syndrome from the anatomy. Spinal sensory deficits were observed but invariably were mild and were overshadowed by the motor weakness.

As with neurologically intact patients with atlanto-axial fractures, the primary treatment was reduction and immobilization of the fracture by external means. *Operative fusion was reserved for inadequate reduction* or for non-union after an adequate period in a halo-vest apparatus. As those patients with spinal cord injury tended to have less stable fractures than those without, the percentage of patients requiring operative fusion was higher in the group with spinal cord injuries than in those without.

P - 8

SPONDYLOSIS IN ACUTE CERVICAL SPINAL CORD AND SPINAL COLUMN INJURIES

D. ROWED
Toronto

Sixteen (21%) of 77 consecutive patients admitted with acute cervical spinal cord injuries had radiographic evidence of pre-existing spondylosis. The mechanism of injury was hyperextension in 14 (88%). Six (43%) of the hyperextension injuries showed a clinical picture of central cord injury syndrome and 5 (36%) of transverse myelopathy.

Twelve of the 16 patients (75%) also sustained acute *spinal column injuries consisting of fracture, dislocation,* fracture-dislocation, rupture of the anterior longitudinal ligament or intravertebral disc herniation. Nine (56%) required immobilization, with or without reduction, by means of halo devices, and 6 (38%) had myelographically demonstrated persistent spinal cord compression with the neck in a relatively neutral position.

Four patients (25%) were subjected to surgical decompressive procedures (3 anteriorly and 1 posteriorly) within 18 days of the time of injury. All improved postoperatively, but the difference in final neurological grade between surgically and conservatively treated patients does not permit any conclusion regarding the influence of early decompression on clinical outcome.

Spinal column injury and persistent spinal cord

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compression appear to result more commonly from cervical spondylosis with hyperextension than has been previously believed. This suggests that a more aggressive approach to early management may be indicated.

P - 9

CEREBRAL BLOOD FLOW ASSESSMENT OF PATIENTS WITH EXTRACRANIAL-INTRACEREBRAL SHUNTS

M. VILAGHY
Toronto

Twelve patients with extracranial-intracerebral (EC-IC) anastomoses were assessed for cerebral blood flow changes prior to and after the surgical procedure by the intra-arterial 133_{Xe} injection technique, initial slope index method.

The mean cerebral blood flow (MCBF) was found to be increased by 35% (41.4 56 ml/min/100g) in 5 patients (41.7%), unchanged (36.4 36.1 ml/min/100g) in another 5 and decreased by 17% (51.2 43.8 ml/min/100g) in 2 (16.7%). The immediate (3-17 days) postoperative condition of the patients was unchanged in 10. Two other suffered strokes on the 2nd day following anastomosis surgery. Long term (1-3 years) follow-up data revealed gradual deterioration of stroke-related deficits in 3 of the 12 cases. No neurological improvement was attributed to the surgical procedure in the other 9. Contralateral ICA or CCA injections proved to be a much better way of injecting 133_{Xe} than via the ipsilateral ECA's both pre- and post-operatively.

Our data suggests that: 1) EC-IC anastomosis surgery results in increased CBF in less than half of the operated cases. 2) There is no correlation between the surgery-induced blood flow changes and post-operative neurological condition of the patients.

P - 10

MORBIDITY, MORTALITY AND RECURRENCE RATES OF SUBDURAL HEMATOMAS IN CT ERA

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Edmonton

One hundred and sixty subdural hematomas, from January 1975 to December 1980, with no other evidence of space-occupying lesions and diagnosed by computerized axial tomography were analyzed. 71% patients were males, 29% were females. 46.2% were acute, 36.9% subacute, and 41.1% chronic subdural hematomas. 16.2% patients had cardiopathies, 8.1% neuropathies, 5.6% were diabetic, and 36.9% alcoholic. 14.4% used antihypertensive medication, 15% anticoagulants, 20.6% other types of medications. 55.7% had burr holes as treatment, 26.6% underwent craniotomy, 3.8% subdural taps, and 13.9% did not receive any treatment. 22.5% cases had less than 50 cc. of subdural hematoma drained, 47.5% less than 100 cc., 15% less than 150 cc. and 15% 150 cc. or more drained. No drain was used in 56.2% of cases, a red rubber catheter was left in 16.2% of cases, Jackson-Pratt drain in 22%, Penrose drain in 2.5% and Hemovac drain in 3.1% cases. Two cases of post-operative infection were recorded, both with the use of the Jackson-Pratt drain. Removal of the drain was within the first 24 hours of insertion in 94.3% of the cases. 11.6% of the patients had recurrences, most of them occurring during the first two weeks postsurgery. 84.4% patients required only one operation. Outcome by groups was as follows: acute subdurals — 55.6% died; 20.8% cured; 12.5% severely disabled; and 11.1% moderately disabled. Subacute subdural: 5% died, 90% cured; 5% moderately disabled. Chronic subdurals: 5.8% died; 84% cured; 2.9% severely disabled; and 7.3% moderately disabled. Significant factors in prognosis will be discussed and comparison made to historical series.

P - 11

CT SCAN AND SERIAL CEREBRAL BLOOD FLOW (CBF) MEASUREMENT IN THE MANAGEMENT OF CEREBRAL ISCHEMIC COMPLICATIONS FOLLOWING SUBARACHNOID HEMORRHAGE

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London

In 31 patients with acute subarachnoid hemorrhage, CT scans within seven days of the initial hemorrhage were reviewed and the patients were divided into three groups: Group I — no blood in the basal cisterns — 9 patients, Group II — blood in the basal cisterns — 17 patients, Group III — intracerebral or intraventricular hemorrhage — 5 patients. Serial CBF measurements using the 133-Xenon inhalation technique were performed in each group of patients and correlation with subsequent CT scans, cerebral angiography and clinical status were studied.

Moderate to severe vasospasm was demonstrated in 11% of group I patients, 82% of group II patients, and 40% of group III patients. The lowest value of mean CBF among serial measurements was 37 ± 6 ml/100g/min in group I patients and 36 ± 5 ml/100g/min in group III patients, both of which were significantly higher than the value of 30 ± 5 ml/100g/min in group II patients ($p < 0.01$). Fluctuation of serial CBF measurements (the difference between highest and lowest CBF in the same patient) was 13 ± 6 ml/100g/min in group II patients. This was significantly greater than the fluctuation (5 ± 3 ml/100g/min) observed in group I patients ($p < 0.001$), suggesting an unstable hemodynamic status in patients with blood in the basal cisterns. Observation of sequential change in CBF in group II patients indicated that the flow fell towards the second week after hemorrhage and recovered by the third or fourth week. As expected, cerebral infarction demonstrated by CT scans and neurological deficit was common in group II patients.

Our results support the concept that the presence of subarachnoid blood on the initial CT scan following subarachnoid hemorrhage is a predictor of cerebral vasospasm. Serial CBF measurements indicate an unstable hemodynamic status in such patients. Wide fluctuation of CBF in this group of patients emphasizes the need for serial monitoring of CBF to obtain useful and reliable information as to the risk of cerebral ischemia and the timing of surgery.

P - 12

GLIOBLASTOMA MULTIFORME WITH PULMONARY, OSSEOUS AND SPINAL EPIDURAL IMPLANTATIONS

E. BERGER, A. NEAGA
Montréal

Distant spread of glioblastomata has occasionally been reported in the literature although doubts were expressed in a few instances concerning the true histological nature of the pulmonary seeding.

It has been postulated that spread occurs through lymphatics and veins outside the nervous system.

A 28 year old male underwent a right parietal craniotomy for removal of a para sagittal tumour attached to the falx. The lesion was partly cystic and histologically a glioblastoma multiforme infiltrating the dura. Post-operative radiotherapy was given.

Three months later the patient was re-admitted for progressive left arm weakness with paresthesia. A myelogram revealed a space occupying lesion from C6 to D2 and at laminectomy an epidural tumour was removed which on histological examination proved to be a glioblastoma multiforme.

Death occurred three weeks later. The post mortem examination showed microscopical evidence of pulmonary infiltration and macroscopic involvement of the vertebrae from C5 to D2, the adjacent spinal epidural space, the brachial plexus, the superior longtitudinal sinus, but no residual tumour at the original site.

P - 13

MULTIPLE CEREBRAL LESIONS AND METASTASES IN C.T. SCAN: EXPERIENCE OF 106 CASES

J. BLAIN, J. LEGER, J. LAMOUREUX
Montréal

The hope that computerized tomography of the skull would obviate and simplify the diagnosis of tumoral lesions has to be reviewed.

One hundred and six consecutive cases of patients with a diagnosis of cerebral metastases made on the basis of the CT Scan are reviewed. The diagnosis was made on the basis of a multiplicity of lesions, the presence of well circumscribed lesions associated with edema and lesions which usually enhanced with the infusion of contrast media.

Fourteen patients or 12% of these cases were proven not to harbor metastatic disease from other organs. Of these, 5 had proven primary brain tumors either at craniotomy or at autopsy. One of these patients was thought to have a metastatic lesion at biopsy but this later proved to be a malignant glioma.

Three patients with occlusive vascular disease and a stroke syndrome were also included in this group. One patient suffered from cryptococcal meningitis and abscess formation. Two patients, still alive, have no definitive diagnosis.

Other pathologies include one patient with primary cerebral lymphoma, one with probable demyelinating disease and one patient with a cerebral hamartoma.

The clinical significance and correlations of multiple lesions on CT Scan are discussed in the light of other paraclinical investigations, in particular, the EEG, technicium and gallium brain scans and angiography.

P - 14

TOPOGRAPHIC ELECTROENCEPALOGRAPHIC STUDY OF OCCLUSIVE CEREBROVASCULAR DISEASE FOLLOWING EC/IC BYPASS OPERATION

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T. KAWASE
Gunma, Japan

As an objective evaluation of EC/IC bypass operation, we studied eight patients with occlusive cerebrovascular disease using computed mapping of electroencephalogram (CME) following EC/IC bypass operation. The CME displays scalp topograph of square roots of average powerspectra over each EEG frequency band on color television. The angiography demonstrated an occlusion of the internal carotid artery (ICA) in 4 cases, an occlusion of the middle cerebral artery (MCA) in 2 cases, a stenosis of the ICA in one case and a stenosis of the MCA in one case before the operation. The preoperative CME findings were, delta foci on the side of the lesion in 4 cases and asymmetrical alpha activity in 4 cases. The CME findings were well correlated with the clinical symptoms. After the operation, the clinical symptoms were gradually improved and the anastomosis was patent in 7 of the 8 cases. The CME demonstrated disappearance of delta foci and increase of alpha activity on the side of the bypass, but on the other hand, theta foci appeared at the site of the operation in 5 of the 8 cases. There was an improvement of delta and alpha activity on CME following EC/IC bypass operation and it well reflected the improvement of the clinical symptoms. To evaluate the efficacy of the bypass operation, 6 of 8 patients were studied by compressing the superficial temporal artery (STA) at the proximal portion to the bypass. In 3 of the 6 cases, an increase of slow components during compression. Changes on CME by compression of the bypassed STA will be helpful in evaluation of EC/IC bypass operation.

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P - 15

CHANGES IN GRAY MATTER DENSITY FROM CRANIAL COMPUTED TOMOGRAPHY IN SEVERE HEAD INJURY

A. ADEGBITE, J. HOWIE, J. McQUEEN
Saskatoon

Emphasis has been placed on CT density changes in the white matter in the past because of the prominence of vasogenic edema in the traumatic setting. We analyzed white and gray matter density changes from multiple sites in 36 consecutive patients with severe head injury (Glasgow Coma Scores < 8). These were divided into one group with GCS 3-4 and GCS 5-8 and were compared with 20 normal adults.

Fifteen patients in an age range from 17-65 years demonstrated evidence of generalized brain swelling on the initial CT scan. A GCS of 3-4 was found in 7 and a GCS of 5-8 was found in 8. A statistically significant increase in the mean gray matter densities was found in the group with GCS of 5-8. Gray matter density means in patients with GCS of 3-4 and white matter density means in both groups were within normal limits. Only 2 of 36 patients showed evidence of classical white matter vasogenic edema on visual inspection. It is postulated that the augmented density reflects hyperemia and that this may be seen early in the gray matter of such patients. One explanation for the normal densities in the GCS 3-4 patients will be given.

P - 16

TRACKING DOWN THE OBLIQUE ORBITAL (INNOMINATE) LINE

R. PERRIN, J. McEWAN
Toronto

The oblique orbital line is a prominent feature on an AP radiograph of the normal skull. This distinct line is projected obliquely (superolateral to inframedial) across the outline of the lateral orbit — hence "oblique orbital line". The precise anatomical basis for this line has been the subject of some debate — hence "linea innominata".

It was postulated at one time, to be due entirely to the squamous temporal bone. Others maintain that the innominate line had no specific anatomical counterpart, but represented "depression in the lateral cortex of the sphenoid bone, forming the medial wall of the temporal fossa". Still others believe it to be an image created when the X-ray beam strikes the junction of the anterior and lateral aspects of the greater sphenoid wing tangentially.

Our interest in the subject was stimulated by a patient who had been treated with bilateral orbital decompression for proptosis complicating Grave's disease. Following ostensibly identical procedures at the right and left orbits with excellent clinical result, the follow-up skull X-ray revealed elimination of the oblique orbital line on one side.

The operative procedure was repeated using the dried skulls and with the aid of serial radiographs we determined that portion of bony removal which was critical in eliminating the oblique orbital line.

It appears that the anatomical substrate for the oblique orbital line involves the junction of the squamous and zygomatic components of the lateral (squamozygomatic) surface of the greater sphenoid wing.

P - 17

IDIOPATHIC DYSGRAPHIA (WRITER'S CRAMP)

R. McLACHLAN
Oshawa

The cause of writer's cramp is controversial. Both psychological and organic etiologies have been proposed. Five patients whose main complaint was slowly progressing writing difficulty are presented. All are males age 21 to 75 years who had symptoms for at least three years. In all patients, attempts at writing induced tremor followed by increased tone and discomfort in the hand and arm. Examples of their writing will be presented. None of the patients exhibited any psychopathology. Four patients had evidence of postural tremor apart from their writing difficulty and one developed a Parkinsonian syndrome. None improved on Propanolol or other medications. Biofeed back was used in two patients. Investigation failed to define any specific abnormality.

The findings in these patients suggest an organic basis for their dysgraphia which appears to be an early sign of defective motor control.

P - 18

CEREBRAL BLOOD FLOW IN BILATERAL CAROTID OCCLUSION

M. VILAGHY, P. DAVIS, J. NORRIS
Toronto

Cerebral blood flow measurement (CBF) is often used as an index of cerebral function when considering patients with severe bilateral carotid artery disease for vascular surgery. 6 patients with angiographically proven bilateral carotid artery occlusion (BCAO) and 4 with severe bilateral (greater than 95%) carotid artery stenosis (BCAS) were evaluated with cerebral blood flow studies prior to potential cerebrovascular surgery. CBF was measured by the intracarotid 133Xe method in 5 cases and by the inhalation 133Xe in 5 patients.

Mean cerebral blood flow was decreased in 3/6 of those with BCAO and 1/4 of those with BCAS. Regional cerebral blood flow was decreased in 5/6 patients with BCAO and 2/4 with BCAS. The remaining patients had either normal or higher than normal CSF values. Only 5 of a total of 10 patients had major clinical disability (3/6 BCAO and 2/4 BCAS) while the remaining 5 cases had minimal or transient symptoms only.

CBF findings correlated poorly both with the clinical state of the patient as well as with the severity of carotid stenosis.

P - 19

ENHANCED DETECTION OF CAROTID STENOSIS BY DOPPLER USING SPECTRAL ANALYSIS

J. NORRIS, B. SHURVELL, F. SILVER
Toronto

The ability to detect carotid lesions by the Doppler ultrasound technique should be increased by the addition of real-time frequency analysis. To evaluate this we compared various indices of accuracy of the Doppler examination to the results of carotid angiography in two groups of patients, using the angiographic findings as a reference standard.

In the first group, 135 carotid arteries (67 patients) were evaluated by Doppler without spectral analysis and compared to the second group of cases consisting of 69 carotid arteries (36 patients) examined with this additional technique. We found: —

1. Stenoses < 35% were not detected accurately in either group.
2. With stenoses 35 - 80% overall accuracy (83%) and specificity (93%) were unchanged but sensitivity rose from 70% to 96%.
3. With stenoses > 80% none of the indices were significantly affected, accuracy remaining high at 92%.

The indices of accuracy of carotid Doppler show a high correlation with carotid angiography with or without spectral analysis in patients with severe carotid stenosis (> 80%). However with lower grade stenoses this additional technique considerably enhances the detection of carotid lesions in the neck.

P - 20

CREUTZFELDT — JAKOB DISEASE PRESENTING AS CONVERSION HYSTERIA

R. RUTHERFORD, J. BILBOA, A. GORDON
Toronto

A 60 year old woman developed left limb ataxia while attending her sister's funeral. Initial investigation including a CT scan, EEG and LP were unremarkable, while neuropsychological and psychiatric evaluations suggested an hysterical component to her illness. The emergence of a paranoid psychosis followed by mutism with catatonic posturing led to atrial of ECT. The diagnosis of Creutzfeldt — Jakob disease was suspected when the patient developed myoclonus, with subsequent confirmation by serial EEG's and brain biopsy.

This patient demonstrates that Creutzfeld — Jakob disease may have psychiatric presentation and the difficulty in differentiating "organic" from "functional" psychiatric disease.

P - 21

ISOLATION OF HERPES SIMPLEX VIRUS FROM THE CENTRAL NERVOUS SYSTEM (CNS) OF LATENTLY INFECTED MICE FOLLOWING IMMUNOSUPPRESSION

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Vancouver, Philadelphia

In addition to acute disease such as encephalitis, Herpes simplex virus can produce persistent infections in man. It is this feature of herpetic infection which makes its' study pertinent to investigation of chronic neurological diseases suspected of being viral in origin.

A murine model of acute and persistent HSV infection was developed based on known aspects of primary infection in man. The spread of virus from the peripheral site of inoculation to the CNS was documented and two stages of infection defined. Studies with different inbread strains of mice suggest that resistance to acute CNS infection is inherited and probably immunologically mediated.

Preliminary studies suggested an important role for T-cell lymphocytes in limiting the primary infection. T-cell ablation in persistently infected animals was produced by thymectomy, lethal dose irradiation, and reconstitution with anti-Thy 1.2 plus complement treated bone marrow cells ('B' mice). Infectious virus was isolated transiently from the CNS of a number of animals but was not necessarily lethal.

This study suggests that under certain conditions of immunosuppression, viral antigens can be present transiently in the CNS of animals harbouring a persistent infection.

P - 22

FACTORS ASSOCIATED WITH RELAPSE OF MYASTHENIA GRAVIS AFTER PLASMA EXCHANGE THERAPY

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Toronto

Almost 60% of patients with Myasthenia Gravis (MG) who undergo plasma exchange (PLX) have a relapse of clinical symptoms following PLX. To identify factors associated with relapse, chemical and immunological parameters were analyzed in 41 patients with generalized MG, who had received PLX and who had been followed for at least 12 months or until the first relapse. The patients fell into 3 categories: Group I - no relapse within 12 months (n = 16); Group II - relapse within 2 months (n = 22); Group III — relapse within 6 - 12 months (n = 3). Group I patients represent patients without factors predisposing to post-PLX relapse, while Group II are presumed to have these factors.

Results showed that Group II (relapse) patients had significantly longer mean duration of disease ($P < 0.01$) — especially disease duration greater than 40 months ($P < 0.025$); but less frequently had a thymoma ($P < 0.05$). There was no significant difference between the two groups in mean age, sex, pre or post PLX clinical grades or severity, prior relapses, or frequency of other autoimmune disease ($P > 0.1$). There was no difference in either the mean pre-PLX or the mean immediate post-PLX steroid or azothiaprine doses ($P > 0.1$). No difference existed between the two groups in the volume or duration of PLX ($P > 0.1$). The frequency of occurrence of other autoantibodies (ANF, RF, thyroid, mitochondrial, smooth muscle, skeletal muscle) did not differ in the two groups ($P > 0.1$). Neither the mean pre-PLX anti Acetylcholine Receptor Antibody (anti-AC_h-R Ab) titres, nor the mean post-PLX anti-ACh-R Ab titres were found to discriminate between the two groups ($P > 0.1$). Moreover, while there was a tendency for Group II (relapse) patients to less

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frequently achieve a significant change in anti-ACh-R Ab titres after PLX (≥ 1 S.D. of normal mean), this difference did not reach statistical significance (n = 12). Preliminary studies of cell mediated immunity, ennumerating the ability of peripheral blood T-Lymphocytes to form E-rosettes, and their modulation by nicotinic cholinergic agonists did not discriminate between the two groups.

P - 23

PERIPHERAL NERVE GRAFTS TO THE OPTIC NERVE IN RATS

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Montréal

A series of grafts and cross-unions between peripheral nerve segments and the retinal stump of the optic nerve were performed intracranially in rats. The results were assessed one week to four months later with standard light and electron microscopic techniques and also with radio-autography after intraocular injection of tritiated amino acids. Although axons were found in most of the cross-united nerve segments and in some of the grafts, none of them were identified to come from the optic nerve. Near the tips of the cut optic nerves, a marked central necrosis developed rapidly. In cross-sections of the optic nerve near the globe, approximately 10% of the normal number of myelinated axons were present four weeks after transection. At four months, a few myelinated axons were still seen and also some unmyelinated axons, probably representing limited axonal outgrowth. In these experiments, neither the survival nor regrowth of cut fibres in the optic nerve was enhanced by grafted peripheral nerve tissue. The lack of response may be due to intrinsic neuronal properties, poor local tissue perfusion or both.

P - 24

A CLUSTER OF AMYOTROPHIC LATERAL SCLEROSIS

C. KREIGER, C. MELMUD
Montréal

Amyotrophic lateral sclerosis (ALS) is an uncommon disorder with a prevalence of 4 to 6 per 100,000 population. Age adjusted incidence is 1.8 per 100,000 in white Americans. The epidemiology is poorly understood — minor sex and racial differences exist. Occasional reports have described geographic clusters of a few cases of unrelated individuals developing the disease at roughly the same time.

Three cases of ALS were discovered within an 18 month period, all living in separate apartments in the same building in Montreal. All patients were between 57 and 63 years of age when diagnosed, of Ashkenazi Jewish extraction and not related. The first patient had autopsy proven ALS, after a progressive two year course. The second had bulbar symptoms leading to generalized disease and death within 12 months of diagnosis. A third is still living (her diagnostic has been supported by serial EMG's and muscle biopsy).

This cluster of patients represents an extremely unusual occurrence which may be due to chance but which may provide clues to the etiology of this disease.

P - 25

THE ETIOLOGY OF SCIATIC NEUROPATHIES

D. GENDRON, J. STEWART
Montréal

We have reviewed the sciatic nerve lesions seen over a 3 year period in an electromyography laboratory. Disorders of the lumbo-sacral plexus or combined lesions of the posterior tibial and common peroneal nerves were excluded. Twenty-two patients had neuropathies of the main trunk of the sciatic nerve.

The causes were as follows: Post-hip replacement, 6 (5 early and 1 delayed); hip fracture with dislocation, 5; compression following coma, 4; other extrinsic compression, 2; mono-neuritis multiplex, injection, herpes zoster, endometriosis, hematoma, 1 of each.

The clinical picture was predominantly motor. The increased susceptibility of the lateral trunk of the sciatic nerve, regardless of the etiology, was confirmed. Foot drop was therefore the commonest presenting feature, and common peroneal palsy the commonest misdiagnosis. Electromyography was valuable in localizing the site of the lesion in many cases.

P - 26

INFLUENCE OF MUSCLE STRENGTH ON SOMATOSENSORY EVOKED POTENTIALS IN THE CAT

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In cats under Pentobarbitone anesthesia (30 mg/kg) six silver ball electrodes were placed epidurally over somatosensory area I. A mechanical stimulation was applied to the contralateral Achilles tendon and 100 evoked potentials were averaged and recorded for each electrode and each of the five selected positions of the tibio calcaneal joint (70°, 90°, 120°, 160°).

Characteristics of the evoked potentials were reliable for each experimental situation (six cortical points and five ankle positions were explored in four cats with a total of 120 experiments). Amplitude and pattern of evoked potentials were related to the angular position of the joint and to localization of cortical points. The highest amplitude was recorded in the post cruciate gyrus with the joint positioned at 160°. These results indicate a close reciprocal influence between the cerebral motor cortex and musculature which should be evaluated when analyzing motor cortex functions.

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FUKUYAMA'S DISEASE

J. McMENAMIN, L. BECKER, E. MURPHY
Toronto

Congenital muscular dystrophy with central nervous system involvement has been reported in Japan. We report a male infant born following a normal pregnancy and delivery to Japanese parents. He presented with hypotonia and muscle weakness from birth. Muscle enzymes were markedly elevated, electromyography was normal and muscle biopsy showed great variation in fibre size, central nuclei, necrotic muscle fibres and proliferation of endomysial connective tissue compatible with muscular dystrophy. Histochemistry was normal. CAT scan demonstrated areas of hypodensity throughout the white matter of the cerebral hemispheres. On follow-up the child has become more progressively weak, is mentally retarded and has a seizure disorder. The clinical and pathological features of this case will be compared to our experience with 24 cases of congenital muscular dystrophy followed at The Hospital for Sick Children, Toronto over a period of 15 years.

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CONGENITAL STRUCTURAL MYOPATHIES

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Toronto

The incidence of congenital structural myopathies is unknown. We have reviewed 1077 muscle biopsies performed in the Hospital for Sick Children, Toronto, during the 20 year period between 1961 and 1980. Eighteen patients with congenital structural myopathies were recognized. Eight had nemaline myopathy, 5 myotubular myopathy, 4 congenital fibre type disproportion, and 1 had central core disease. During the same period, approximately 200 cases of Duchenne Muscular Dystrophy were diagnosed.

The sex distribution in our cases is approximately equal, except in the myotubular group where all 5 patients were male.

Seven patients had associated joint contractures.

Eleven patients presented with neonatal hypotonia, (4/8 nemaline, 4/5 myotubular, 2/4 congenital fibre type disproportion, and the one patient with central core myopathy). As five of this group had also had perinatal asphyxia, the etiology of the hypotonia was initially unclear. Both E.M.G. and serum C.K. levels were uniformly unhelpful. We will therefore propose clinical criteria for muscle biopsies in hypotonic infants. Fourteen patients had no family history of neuromuscular disease; but, there were two sets of siblings. A muscle biopsy of the father of one of the children with nemaline myopathy showed characteristic abnormalities while the mother's biopsy was normal.

The clinical and pathological features of our cases will also be presented.

P - 29

LUETIC MIDDLE CEREBRAL ARTERY STENOSIS

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Ottawa

With the development of cerebral revascularization procedures, the etiology and especially the prognosis of middle cerebral artery (MCA) stenotic lesions is of great interest. In cerebrovascular syphilis, the MCA is the most commonly involved vessel.

A 67 year old woman presented with confusion, dysphasic speech, mild right hemiparesis and Argyll-Robertson pupils. Angiography showed a tight (1 mm) left MCA stenosis. The VDRL was positive in blood but negative in CSF. Blood FTA-Abs was positive. CSF contained 2 WBC and 44 mg % of protein. She was treated with 13 million units of penicillin.

Because of the tight MCA stenosis, the uncertain prognosis of MCA stenotic lesions, and the indefinite CSF findings, a prophylactic left STA-MCA anastamosis was performed. Brain biopsy from the left superior temporal gyrus showed normal cerebral cortex; the meninges showed chronic inflammatory and vascular changes compatible with meningovascular syphilis.

Repeat angiography 3 years later documented that the previous tight left MCA stenosis was greatly improved (3.5 mm). The bypass remained patent. Her blood VDRL is now negative and this finding, combined with the positive meningeal biopsy and the follow-up angiographic improvement, indicate that the MCA stenosis was luetic rather than arteriosclerotic. This experience suggests that luetic MCA stenosis may be managed with anti-syphilitic therapy alone.

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NORMALIZATION OF THE SOMATOSENSORY EVOKED POTENTIAL IN INCOMPLETE SPINAL CORD INJURIES

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Toronto

The cortical somatosensory evoked potentials in patients with incomplete lesions of the spinal cord normalize as clinical sensory and motor function improve. Statistical verification of this statement was obtained as follows.

Cortical somatosensory evoked potentials in response to left and right tibial nerve stimulation were obtained at admission and discharge in twenty-five patients with incomplete cord injuries at varying levels of the neuraxis. In addition, composite scores of motor and sensory neurological function distal to the level of lesion were obtained for right and left body sides for each patient at admission and discharge. The latencies and amplitudes of each peak of the patient's tibial evoked potentials were compared with those of normal evoked potentials to produce an index of abnormality for each potential.

In general, it was found that:

(1) neurological status improved between admission and discharge,
(2) the normality of tibial evoked potentials increased between admission and discharge,
(3) the better the neurological status, the greater the normality of the tibial evoked potential, and finally,
(4) the increase in normality of the tibial evoked potentials between admission and discharge was directly proportional to the improvement in neurological status.

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QUANTITATIVE EMG OF THE APB

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Quantitative EMG of distal hand muscles should identify patients with neurogenic disorders to be of diagnostic use. The number of turns per second, mean amplitude between turns, and ratio of turns to mean amplitude were measured in the abductor pollicis brevis muscle at three force levels: maximum, 30% of maximum, and an absolute 0.3 kg force. Fifteen controls showed no relation between turns at the three force levels and the maximum force — unlike results in large limb muscles. Twenty-one patients were studied: 10 with the carpal tunnel syndrome, 9 with polyneuropathy, and 2 with motor neuron disease. Seventy-five per cent had increased mean duration of motor unit potentials when measured singly. At the 0.3 kg force, turns were decreased in 7 patients, mean amplitude increased in 3, and the ratio of turns to mean amplitude was decreased in 13 giving a cumulative diagnostic yield of 2/3. At 30% of maximum, or maximum force, the yield was only 1/2. The most information resulted when values at 0.3 kg and maximum force were combined. Turns were decreased in 14 patients, and 85% of the total patient group was identified as having neurogenic involvement. These results show that this form of quantitative EMG is valuable when investigating patients with suspected neurogenic disease.

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HALO DEVICES IN THE TREATMENT OF ACUTE CERVICAL SPINAL CORD INJURY: THE SUNNYBROOK EXPERIENCE

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Toronto

Fifty-nine patients with acute cervical spinal cord injury (25 complete, and 34 incomplete) were treated by immobilization in a halo device. The age of the patients ranged from 12 to 86 years. The bony injury consisted of a variety of dislocations, fractures, and fracture-dislocations. Fifty of the patients were followed up adequately (over 6 months) to determine the results of bony fusion and neurological improvement. Seven died early mainly from respiratory causes, and 2 were lost to follow-up. Successful healing with stability at the fracture site was achieved in 43 cases (86%) as a result of the initial treatment, and after late fusion in the remaining 7. Improvement in the neurological deficit was noted in 44 patients. No patient was made worse by the halo device. The average duration of hospitalization was 43 days for incomplete cord injury patients, and 63 days for complete cord injury patients. Complications related directly to the halo devices were few and minor and included scalp infection, pressure sores, and loosening halo pins — none of which necessitated discontinuation of halo treatment. In our experience there are no absolute contraindications to the use of halo devices. In the presence of continuing spinal cord compression, the device may be used after initial cord decompression, and where the bony injury is excessively unstable and frequent redislocations occur, it may be used after an early surgical fusion. The major advantage of halo devices is that they allow one to externally manoeuver various bony injuries in all three dimensions and to fix them when acceptable reduction is attained without resorting to early surgical procedures with their attendant complications in these already very ill patients. In addition they allow early mobilization of patients with spinal cord injuries, and early discharge from the acute hospital setting.

P - 33

CHARACTERISTIC LONG-LATENCY REFLEX ABNORMALITIES IN DYSTONIA AND RIGIDITY

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Toronto

Reflex activity in response to wrist displacement, imposed by a torque motor, was studied in a normal human population and compared to patients with Dystonia Musculorum Deformans and Parkinsonian rigidity. Normal reflex responses consist of 2 distinct EMG peaks, previously termed M1 and M2/M3. The latency and duration of these peaks was constant despite widely varying amplitudes (20-50 degrees) and initial velocities (20-250°/sec) of the displacement. Normal values for flexor carpi radialis (FCR) are:

	M1	M2/M3	
latency	31.7 ± 3.0	56.7 ± 3.5	ms.
duration	21.5 ± 4.2	38.0 ± 3.5	ms.

The reflex magnitude was quantitated as a percentage of the EMG activity during a maximum isometric contraction. Magnitude was shown to increase in a non-linear fashion as the initial velocity was increased and was best described by a family of power functions dependent on the level of background EMG activity. For example, with background activity equal to 8.6%.

M1 (%) = 5.1 *ln (velocity) - 9.7 (r = .99)
M2 (%) = 10.2 *ln (velocity) - 27.5 (r = .99)

In the Parkinsonian and dystonic patients the latency, duration, and power functions describing the reflex magnitude of the M1 were within normal limits. The M2/M3 component displayed characteristic and diagnostic differences from normals. The Dystonic M2/M3, although normal in latency and magnitude had a prolonged duration, directly related to the duration of the imposed displacement. That is, the reflex activity persisted as long as the imposed movement continued, whereas that in the normals terminated after a constant duration despite continued movement. The Parkinson M2/M3 displayed normal latency and constant duration, however, the magnitude was 2-5 times normal for given initial velocity. The power functions for the Parkinsonian M2/M3 effectively saturated at low velocities. Using three-dimensional plots it can be shown that the abnormal M2/M3 in Parkinsonians and Dystonics cannot be explained by increased levels of resting motorneuron activity and is dependent on characteristic defects in central control. Such quantitiative analysis provides a sensitive means for diagnosis of these conditions and the evaluation of various therapeutic measures.

P - 34

SPONDYLOSIS IN THE ETIOLOGY AND OUTCOME OF CERVICAL SPINAL CORD INJURIES

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Toronto

Cervical spondylosis is a common antecedant cause of spinal cord injury. The patient population and mechanism of injury differ from cervical cord injuries in general and the outcome is better when identical principles of management are followed.

Sixteen (21%) of 77 consecutive acute cervical spinal cord injuries showed radiographic evidence of spondylosis and the remaining 61 did not. Salient features of comparison follow.

The mean age of the spondylosis patients was 65 ± 10* years compared with 32 ± 15 years. Falls were the cause in 9 (56.3%), compared with 18 (29.5%)**. The mechanism of injury was extension in 14 (87.5%), as opposed to 1 (1.6%)***.

Only 2 patients (12.5%) sustained complete motor and sensory loss distal to the level of injury, compared with 28 (45.9%) of the others**. The mortality in spondylosis patients, however, was quite high. Three (18.8%) died, all from respiratory complications, compared with 7 (11.5%) of the others.

At final follow-up (12 months), 5 (38.5%) of the survivors in the spondylosis group could walk unassisted, as opposed to 17 (31.5%). Only 2 (15.4%) remained unable to bear weight, compared with 30 (55.6%)**, whereas 6 (46.2%) had returned to full or part-time work, compared with 10 (18.5%)**. Nine (69.2%) were living independently, compared with 19 (35.2%).

* one standard deviation
** $P < 0.05$
*** $P < 0.001$

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MOTOR LEARNING IN DIFFERENT KINDS OF MOVEMENT

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Toronto

The quantitative measurement of movement has been used to follow motor development and the natural history of a movement disorder and to document responses to treatment. In order to be useful these measurements must be reproducible and variables which may influence the results need to be recognized. The present study investigates the influence of handedness, practice or learning and schedule of testing on several computerized tests of hand movement. Tasks measuring speed, accuracy, initiation and steadiness of movement were presented and quantitated on-line using a Declab 1140 computer system.

In normal adults there was little difference in motor function between the dominant and non-dominant hand for most tests. There was however, a distinct advantage of the dominant hand for speed in tracing which, however, diminished with practice. Most tests showed improvement on repetition thereby demonstrating motor learning. In a reaction test both the time taken to initiate movement and the time required to converge on the target decreased with practice. For the initiation of movement, practice with one hand improved the performance of the second hand, i.e. there was transfer of learning. In contrast, for convergence on the target there did not appear to be any influence of the first hand's practice on the second hand's function.

Thus these results demonstrate motor learning in several different tasks of hand movement. Moreover, they suggest that there are at least two kinds of motor learning, namely those in which learning can be transferred from one hand to the other and those in which learning remains lateralized. These presumably have different anatomical and physiological correlates and theoretically could be selectively altered in specific disorders of movement.

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SOMATOSENSORY EVOKED RESPONSES IN PATIENTS WITH UNILATERAL CEREBRAL HEMISPHERE LESIONS

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Calgary

Somatosensory evoked responses (SER) to stimulation of the median nerve at the wrist were recorded from 20 patients with infarcts or tumors involving one cerebral hemisphere. Responses from the abnormal side were compared with those recorded over the opposite hemisphere and with SER's from a group of normal control subjects. Abnormalities in the SER's were correlated with findings on clinical examination of sensation and with the results of CT scanning.

Marked suppression of all components of the SER was a consistent finding in patients with extensive sensory loss and large lesions demonstrated by CT Scan. In two patients with sensory inattention and unilateral neglect the SER was almost absent over the affected hemisphere. However disruption of the SER was also observed in some patients with no abnormality on the CT Scan and in one patient with pure motor hemiplegia and no sensory deficits on clinical examination. In contrast, the SER was entirely normal in a patient with a lateral medullary infarct causing unilateral loss of pain and temperature sensation.

In two patients with "cortical sensory loss" due to localized lesions of the post-central cortex there was preservation of the early negative wave of the SER (N1 - latency 18-20 msec.) with suppression of the later components. This suggests that N1 may be generated by subcortical structures such as the thalamus.

It is concluded that the SER is a potentially useful adjunct to the clinical examination and the CT Scan for assessing functional integrity of higher order components of the somatosensory system.

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LOW BLOOD AND CEREBROSPINAL FLUID (CSF) THIAMINE VALUES IN PHENYTOIN TREATED EPILEPTICS

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Montréal

Blood and cerebrospinal fluid (CSF) thiamine contents were assayed by *Lactobacillus fermenti* microbiological assay.

Normal controls were considered those who i) did not display any gastroenterological or metabolic disease; ii) had not any evidence of neuropsychiatric manifestations which could be attributable to a nutritional deficiency and iii) had a rather normal or borderline normal dietary assessment.

The normal thiamine values were compared with the values of epileptic patients receiving phenytoin (PHT) for more than 5 years. Epileptic patients taking alcohol were ruled out from the study. There were no significant differences for age amongst the two group. The whole blood thiamine values ($x \pm S.D.$) in normal controls (89 S_t) was 66.69 ± 12.77 ng/ml while in epileptic patients (15 S_t) was 45.69 ± 6.04 ng/ml ($p < 0.001$).

The CSF thiamine values in 23 normal controls ($x \pm S.D.$) was 44.03 ± 6.9 while in treated epileptics (9 S_t) was 19.86 ± 11.49 ($p < 0.001$).

These preliminary findings could open new views on the possible side effects of PTH; further vertifications are required.

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FACIAL NERVE PARALYSIS

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Toronto

This study is based on a series of over one hundred patients suffering from facial nerve paralysis due to various etiologies seen in a facial nerve clinic. These patients were fully assessed (clinical assessment, audiological studies and, where appropriate radiological and electrophysiological studies) and closely followed.

Based on this experience the prognostic value of various clinical features and investigative test results will be discussed and recommendations made with respect to treatment.

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EFFICACY AND TOXICITY OF VALPROIC ACID IN PATIENTS WITH COMPLEX PARTIAL SEIZURES

J. BRUNI
Toronto

Twenty-four adult outpatients with poorly controlled complex partial seizures were treated with valproic acid in conjunction with their previous anticonvulsants. The period of follow-up varied from 3 months to 36 months (average 8 months). Valproic acid dosage was adjusted accordingly to response, toxicity, and plasma levels. Plasma levels of the other anticonvulsant drugs were maintained within the therapeutic range. Plasma valproic acid concentrations ranged from 49.7 μl/ml to 105.0 μg/ml. Five patients (21%) obtained long-term excellent seizure control (greater than 75 percent seizure reduction); seven patients (29%) achieved good seizure control (greater than 50 percent reduction) for a variable period (3 months to 18 months). Twelve patients (50 percent) achieved good to excellent control initially. Twelve patients (50 percent) failed to show any significant response. Responders and non-responders did not differ significantly with regards to seizure frequency, age and sex, and duration of seizure disorder. Clinically and laboratory side effects were generally mild and did not require discontinuation of therapy. The side effects will be reported. The data suggest that valproic acid has a long-term beneficial effect in only a small number of patients with this seizure type. A significant number of patients develop tolerance to its initial anticonvulsant and/or placebo effect. Further controlled trials are required to determine the efficacy of valproic acid in less refractory patients with complex partial seizures.

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THE EFFECT OF SINGLE ANTICONVULSANTS ON FASTING PLASMA AMMONIA AND AMINO ACIDS IN CHILDREN

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Vancouver

Hyperammonaemia has been noted in patients on multiple anticonvulsants including Valproic Acid and it has been suggested as a cause of the stupor sometimes found in these patients. Infants on Phenobarbital or Primidone have also been noted to have elevated fasting plasma and CSF glutamine and ornithine levels and it was suggested this might reflect an alteration in ammonia metabolism.

We studied 10 children on Phenobarbital (10 months - 16 years; mean 4.4 years), 10 children on Carbamazepine (6.5 - 15 years; mean 8.5 years), and 8 children on Valproic Acid (14 months - 9 years; mean 4.5 years). All children were on a single anticonvulsant and had normal liver function tests.

Two children on Phenobarbital had a mild elevation of plasma ammonia at 45 and 53 micromoles per litre (normal up to 40 micromoles per litre) but plasma glutamine and ornithine levels were not elevated in any of the 10 patients. Two children on Carbamazepine had slightly elevated plasma glutamine levels of 820 and 845 micromoles per litre (normal range 360 - 740) but ammonia and ornithine levels were normal. Plasma ammonia, glutamine and ornithine levels were normal in all patients on Valproic Acid. Glycine levels were significantly higher in patients on Valproic Acid.

The anticonvulsants studied, given singly, had little effect on plasma ammonia, glutamine or ornithine in these children.

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IS ICTAL FEAR A FEATURE OF GENERALIZED SEIZURES?

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Two patients had seizure disorders of unknown etiology in which the feeling or the expression of fear was a prominent feature. EEG features were, however, suggestive of a generalized seizure disorder.

A nine year old girl had absence attacks with 3/sec. spike and wave. Ethosuximide abolished these for 14 months. She then developed spells for which she was amnestic consisting of a fearful expression, sweating and tachycardia, coincident with a 10/sec. bifrontal rhythm which quickly generalized. This was preceded by generalized spike and wave or generalized or bitemporal complexes. A videotape will be shown.

A 23 year old woman had a nine year history of seizures beginning with disorientation and fear followed by aimless walking about and partial responsiveness. 3/sec. spike and wave was the only EEG abnormality.

Neither patient had features implicating temporal lobe origin. Physical and neurological examinations were normal.

The distinction between partial and generalized seizure disorders is sometimes blurred and fear by itself may not be a reliable differentiating feature. The EEG is valuable in distinguishing the seizure type.

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VALPROIC ACID (VPA): FIRST-DOSE AND STEADY-STATE PHARMACOKINETICS IN CHILDREN WITH SEIZURES

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Winnipeg

There is little information on the pharmacokinetics of VPA in children and none comparing pharmacokinetic parameters obtained after a single dose with those at steady-state. We therefore present the results of such a study done in seventeen children:

	Half-life ($t^1/_2$) hrs.	Elimination rate (Ke) hrs. $^{-1}$	Vol. of Distribution (V_D) L/kg	Clearance (Cl) ml/min/kg	ml/min/m²
Single dose n = 17	7.7 ± 2.5	.100 ± .035	.17 ± .05	29 ± 13	7.4 ± 2.8
Steady-state n = 17	7.3 ± 1.2	.098 ± .020	.22 ± .07	37 ± .16	9.7 ± 3.7
P	NS	NS	< 0.02	< 0.05	< 0.05

There was a significant difference between the Cl and V_D of VPA between the two study periods.

	$t^1/_2$ (hr.)		Ke (hr. $^{-1}$)		Cl (ml/min/kg)		V_D (L/Kg)	
	Single Dose	Steady State	Single Dose	Steady State	Single Dose	Steady State	Single Dose	Steady State
0.75 - 9 years	7.5 ±	7.0 ±	.105 ±	.103 ±	.33 ±	.42 ±	.19 ±	.24 ±
n = 11	2.7	1.4	.039	.023	.13	.18	.06	.08
10 - 20 years	8.2 ±	7.7 ±	.090 ±	.091 ±	.22 ±	.28 ±	.14 ±	.19 ±
n = 6	2.2	0.8	.026	.008	.07	.05	.01	.04
P	NS	NS	NS	NS	< 0.05	< 0.05	< 0.02	NS

Cl of VPA was significantly different in the two age groups at both study periods whereas V_D was significantly related to age only in the single dose phase. Our study suggests that: 1) changes frequently occur in the pharmacokinetics of VPA in individual patients between the first dose and at steady-state (which exists during maintenance therapy), 2) pharmacokinetics of VPA are age-related. These findings should be taken into account in individualizing VPA therapy and assessing its efficacy. This study was supported by the White Cross Guild, Children's Hospital Research Foundation and the Manitoba Medical Services Foundation Incorporated.

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TEMPERATURE EFFECTS ON HUMAN COMPOUND ACTION POTENTIALS: COMPUTER SIMULATION TO DEFINE MECHANISMS

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London

In a previous study, we showed that rising temperature over a physiological range caused a progressive reduction in the latency, duration, amplitude and area of the human sensory compound action potential (CAP). The summated effects of changes in the conduction velocities of various-sized myelinated fibers, and changes in the duration and amplitude of the action potentials of single fibers, are postulated mechanisms. We used computer simulation studies to determine the role of each mechanism.

Previous electrophysiological and morphological studies of nerves provide estimates of the range and relative numbers of various-sized myelinated fibers contributing to CAP, and computer studies by Stegeman et al (1979) provide an estimate of the size and shape of the action potential of a single myelinated fiber recorded at the skin surface. Our computer simulation study produced the closest match to CAP changes observed in humans during rising temperature under the following conditions: a considerable reduction in the duration of the single fiber action potential, but only a mild reduction in amplitude, and a rise in the conduction velocity of various-sized myelinated fibers according to a semi-logarithmic relationship of temperature to velocity.

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VALPROIC ACID (VPA) PRODUCING A REYE-LIKE SYNDROME

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Ottawa

Although several cases of acute hepatic failure associated with the administration of VPA have been reported, the pathogenesis of this problem remains unclear. The purpose of this communication is to report another case with several features similar to those of Reye's Syndrome, suggesting the possibility of common pathogenesis.

A 40 month old male with chronic seizure disorder on VPA and phenytoin for two years was admitted after a prolonged seizure. During the next 72 hours, he showed progressive neurological deterioration and hepatorenal failure (SGOT > 2500; Ammonia 264; PT/PTT elevation;

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bilirubin > 1; BUN 52; CBC, platelets & blood sugar normal). On day four ICP monitoring was started because of early papilledema (initially 66 mm Hg). On day nine a percutaneous liver biopsy showed early post necrotic fibrosis and diffuse fatty changes. Despite aggressive management of ICP, patient developed refractory intracranial hypertension and died on day 20. Postmortem examination showed severe brain swelling with diffuse hypoxic-ischemic changes and resolution of hepatic abnormalities.

As with previous reports of hepatic toxicity with VPA, this patient received several potentially hepatotoxic agents; however, acute fatal hepatic failure has been associated more often with VPA. Considering the many features in this case similar to Reye's Syndrome and the experimental evidence implicating short chain fatty acids in the pathogenesis of this disorder, it is suggested that VPA, a short chain fatty acid, may produce hepatic failure by impairment of mitochondrial functions.

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CAN THE VESTIBULO-OCULAR REFLEX (VOR) BE ISOLATED FOR QUANTITATIVE DIAGNOSIS?

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Toronto

VOR measurements have traditionally been employed in the diagnosis of vestibular and brain stem disorders, however, it has not been possible to eliminate the contribution of non-vestibular mechanisms to compensatory eye movements. In order to achieve this, a new rotary test has been developed in our clinic which involves measuring the VOR within the dynamic range of normal head movement (0-5 Hz) thus excluding contamination by the visual reflexes operating at low frequencies (approx. 0-2 Hz). In addition, a pseudo-random oscillating stimulus prevents predictive motor commands. Theoretically, therefore, only vestibular imput should be operational in the higher frequency range. To examine the possibility of visual contamination, normal subjects were tested under 2 conditions: 1) Subjects were instructed to fixate a small stationary visual target, a task in which the VOR is supplemented by vision. The resulting VOR gains were found to be 1 (0 dB) for the entire frequency range tested, indicating perfect compensation. 2) The contribution of visual reflexes was quantified by a VOR suppression task in which subjects were instructed to fixate a small visual target moving with him. VOR suppression to -3 dB at low frequencies (0-1 Hz) verified the assumption of an interaction between the smooth pursuit system and the VOR. Above 3 Hz, the gain rose to 4 dB at approximately 4.5 Hz. The high frequency gain difference in the visuo-vestibular co-operation task and the suppression task implies that, extra vestibular factors (eg. central motor programmes) are present at the high frequencies previously believed to be representative of only vestibular input. In view of the fact that a similar high frequency gain increase has been observed in spike data from the vestibular nerve (Fernandez & Goldberg, 1971) and in sinusoidal VOR data in darkness (Benson, 1970; Keller, 1978), it is reasonable to propose that high frequency performance during visual suppression test, best represents the pure vestibular drive.

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SCIATIC NERVE PALSY DUE TO ENDOMETRIOSIS

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Montréal

A 33 year old woman presented with a 3 year history of increasing pain in the right upper posterior thigh associated with weakness of that leg for 3 months. The right leg was maintained in flexion and external rotation, and all movements of the hip exacerbated the pain. Clinical examination combined with electromyography and nerve conduction studies showed lesions involving the sciatic and inferior gluteal nerves. Plain radiographs of the pelvis, hips and lumbar spine, and a myelogram, were all normal. A CT scan showed a mass in the region of the right sciatic notch. In addition, the pyriformis and gamelli muscles were scarred and compressing the nerve.

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CHRONIC VASOSPASM IN MONKEYS AFTER LARGE SUBARACHNOID HEMORRHAGE

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Edmonton

The onset of delayed neurological deficit in patients with subarachnoid hemorrhage (SAH) and severe vasospasm (VSP) appears to relate to the volume of subarachnoid blood apparent on computed tomography scans (CT). A large volume of autogenous arterial blood (1.3 to 1.7 ml/kg body weight) was injected within 15 min. into the chiasmatic cistern in 12 anesthetized cynomolgus monkeys. CT, cerebral angiography and cerebral blood flow (CBF) by ^{133}Xe clearance were performed before and within 5 hr. post-SAH and at 7, 14 and 21 days after. Neurological assessment was made daily during this time. The CT showed thin to fairly thick layers of SAH in all animals, and subfrontal hematoma in 3. Animals with the largest CT-SAH had more VSP acutely and at 7 days. Mild to moderate VSP occurred acutely and at 7 days (average of 24% in 12 of 12 animals and 15% in 7 of 8 respectively). Only mild VSP occurred at 14 and 21 days (average of 12% in 5 of 6 animals and 6% in 4 of 5 respectively). None of these animals showed more severe VSP at 7, 14 or 21 days than was seen acutely. Post-SAH CBF changes were inconsistent but generally decreased, at the acute stage and day 7, 14 and 21 respectively average decreases of 31% occurred in 8 of 8 animals, 5% in 7 of 7, 17% in 6 of 6 and 6% in 5 of 5. Neurological deterioration (frontal lobe syndrome) developed in one animal persisting from day 17 to 21. Findings indicate: (1) large SAH in the monkey can cause chronic mild/moderate VSP; (2) chronic severe VSP has not occurred to date using this model; (3) VSP is maximal immediately after SAH, and vessel caliber tends to normalize by 21 days; (4) the degree of VSP induced was not associated with delayed detectable neurological deficit. In conclusion large SAH did produce chronic VSP without obvious delayed neurological deficit in the monkey. The reasons for this discrepancy to the clinical situation might be: (1) insufficient collection of blood around the circle of Willis in the monkey due to small size of subarachnoid space compared to humans; (2) species difference; (3) our inability to detect subtle neurological deficits.

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