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HERBERT JASPER PRIZE

Effects of Stimulus Shape on Visual Evoked Potentials

by Sherrill J. Purves, Vancouver, B.C.

The recording of cerebral evoked potentials has been widely used as a method of studying correlates of sensory processes in human subjects. In the visual system it has been established that certain physical parameters of the stimulus including intensity, focus and size of the checks in a checkerboard pattern have significant effects on the configuration of the evoked response recorded from the occipital region. Another, as yet unreplicated study has shown that evoked responses to certain geometric forms are different, and it has consequently been suggested that the evoked responses reflect higher perceptual processes rather than ones related to physical properties of the stimuli.

Evoked responses to four geometric shapes (a square, circle, ϵ and ω) were recorded from multiple scalp locations. There were significant differences between the evoked responses in the occipital region to the square and ϵ shapes and between those to the circle and ω shapes. These differences were demonstrated by the three measurement techniques; performance of the discriminant functions computed by SWDA in classifying single trail responses, a ratio statistic called λ and amplitude differences in the N₂ and P₂ components at 148 and 219 msec. latency.

The evoked potential differences found were believed to be due to two classes of variables; the physical characteristics of the stimulus, specifically in the central 1.5° of the visual field, and the symbolic meaning of the stimulus; and these affected earlier and later parts of the waveform respectively.

K. G. MCKENZIE MEMORIAL AWARD NO. 1

The Effect of Amphotericin B on the Survival of Brain Tumor Bearing Mice Treated with CCNU

Paul J. Muller

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The nitrosoureas are very effective in prolonging survival in experimental brain tumors; however, clinical results have been disappointing. It has been suggested that the reduced efficiency of nitrosoureas may be related to their failure to reach some parts of the cerebral tumor in adequate concentration in spite of their excellent blood brain barrier penetration properties. In vitro studies by a number of investigators have shown that amphotericin B (AMB) increases cell permeability and potentiates or enhances the effects of various agents. In vivo studies have shown that AMB potentiates the therapeutic effects of nitrosoureas in AKR leukemic mice and a subcutaneous murine tumor.

We have studied the effect of CCNU with or without AMB pretreatment in our transplantable intracerebral mouse ependymoblastoma. Survival studies showed that CCNU IP 5mg/kg administered on the 5th day after intracerebral (IC) tumor implantation was ineffective in prolonging survival; 10 mg/kg was moderately effective and 20-30 mg/kg was highly effective. The addition of AMB IP on the 4th day did not improve survival in CCNU treated brain tumor bearing mice. This failure of response to AMB IP is likely a failure in drug delivery since AMB crosses the blood brain barrier very poorly.

To circumvent the drug delivery problem, AMB was administered IC, directly into the tumor bearing hemisphere. With AMB IC 0.5 mg/kg 24 hours prior to CCNU IP 10 mg/kg, the percent increase length of survival (%ILS = 61) was significantly ($p < .05$) increased over control when CCNU alone did not significantly alter the survival (%ILS = 17). When 0.2 or 1.0 mg/kg AMB IC preceded CCNU IP 20 mg/kg the %ILS was

significantly ($p < .01$) increased over the group receiving CCNU 20 mg/kg alone. When treatment was delayed for two weeks the AMB IC - CCNU IP combination groups did better than the CCNU alone group although the %ILS was small. We have confirmed the potentiation effect of AMB IC on CCNU IP by modifying a subcutaneous tumor growth and point dilution technique for the assessment of in vivo therapy of experimental brain tumors.

We may conclude that direct, local (IC) administration of the membrane active polyene, AMB, enhances the therapeutic effect of CCNU in experimental brain tumors.

Cause of Death in Acute Stroke

V. Bril, J. W. Norris, V. C. Hachinski

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Ninety-seven of 413 patients admitted to an Intensive Care Stroke Unit died; 85 of these died within the first month which represents a 21% mortality rate. The death rate in 265 patients with cerebral hemispheric infarction was 18% compared to 94% of 36 patients with cerebral hemispheric hemorrhage. Analysis of the 44 autopsied cases revealed high diagnostic accuracy for hemispheric lesions (90% for cerebral infarction and 82% for cerebral hemorrhage).

Twelve (18%) of 68 patients with brain stem lesions died mainly from complications of their strokes. Autopsy correlation indicates a much lower incidence of diagnostic accuracy than in patients with hemispheric lesions.

The major cause of death within the first three days of stroke was brain stem compression by cerebral pressure cone. This was the main cause of death in the cerebral hemorrhage cases. Thereafter, patients died from the complications of stroke mainly pneumonia. This was the main cause of death in the cerebral infarction group. In 4 of 17 autopsied cases with cerebral infarction acute myocardial infarction played a major role in the mortality.

Although in many stroke patients a fatal outcome appeared to be inevitable, the pattern of mortality indicates that medical or surgical treatment available through an Intensive Care Unit may prove life saving in selected cases.

Lumbar Disc Disease in Children

Eddy Garrido, Robin P. Humphreys, E. Bruce Hendrick and Harold J. Hoffman

Division of Neurological Surgery, The Hospital for Sick Children, University of Toronto, Toronto, Ontario

The clinical experience with lumbar disc disease in 39 patients at a large children's hospital is reviewed.

All patients were treated initially with prolonged bedrest for several weeks. Thirty-one patients underwent surgery because of failure of conservative management. Three of the surgical patients proved to have posteriorly displaced epiphysis as the cause of the disc syndrome. Ninety-four per cent of the operated group were cured or markedly improved on follow-up. Seven patients (18%) obtained long lasting relief with nonsurgical treatment. The average follow-up for the whole group was one year.

The clinical presentation, surgical aspects and main differential features of disc disease between adults and children are reviewed.

Specificity of Afferent Information Carried via Eye Muscle Proprioceptors to the Visual Cerebellum

D. W. F. Schwarz and R. D. Tomlinson

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Extraocular proprioceptive input to cerebellar vermis, lobule VI, was investigated in cats under N₂O analgesia by recording neuronal responses to eye muscle stretch. Both optic tracts were transected and the periorbital skin and conjunctiva were locally anaesthetized. Eye rotation within the physiological range was achieved by applying a pull of predetermined length and tension to each of the eight musculi recti at their insertion to the globe. Within lobule VI only small patches of cortex receive stretch receptor afferents. The information made available by

these afferents corresponds to a change in eye position. Minimal responses were dependent upon angular deflections of a few degrees, maximal response amplitudes were obtained within the physiological range of angular deflections and angular velocities for the units tested. Most cells responded to stretch of more than one muscle. Three types of convergence were found: 1) neurons responding according to a certain direction of conjugated movements of both eyes, 2) neurons responding to movements in either direction of one plane, 3) more complicated response patterns.

A Latent Defect in Neuromuscular Transmission in Multiple Sclerosis Detected by Undue Sensitivity to Curare

Andrew Eisen and Robert Yufe, Montreal

The association of multiple sclerosis (MS) and myasthenia gravis is rare, only twelve cases have been reported to date. The possibility of a more frequent subclinical defect of transmission in MS, has been studied by measuring curare sensitivity in nineteen patients, and comparing the results with twenty-three control subjects.

Methods: 0.3 mg of curare were injected regionally (Brown et al., *J. Neurol. Neurosurg. Psychiat.* 38: 18, 1975). 3Hz trains were delivered at fixed time intervals after the injection. The amplitudes of the first and fifth responses (recorded from the first dorsal interosseus) of each post-injection train, were expressed as a percentage of the amplitude of the pre-injection first response.

Results: Twenty-five minutes after the curare injection, amplitudes of the first and fifth responses of the control group were $102.9 \pm 8.2\%$ and $97.1 \pm 12.0\%$ of the pre-injection value. The corresponding values of the MS group were only $8.12 \pm 23.4\%$ and $71.8 \pm 27.1\%$ ($p < 0.001$). Significant differences in the degree of recovery were also noted at 10, 15, and 20 minutes after the injection. The overall times to recovery of the first and fifth responses of the control group were 20.0 ± 8.2 min. and 27.6 ± 7.8 min. There was a significantly delayed recovery of the first response in 58% of the patients, and of the fifth response in 42%.

It is concluded that the increased sensitivity to curare found in MS is indicative of a latent defect in neuromuscular transmission. This finding occurs sufficiently frequently in MS as to make the association more than just fortuitous.

Radiopharmaceutical Bone Scanning in Pediatric Neurosurgery

R. P. Humphreys, D. L. Gilday, J. M. Ash, E. Bruce Hendrick and H. J. Hoffman

The diagnostic evaluation of inflammatory and metastatic neoplastic lesions of the skeleton has been facilitated by radiopharmaceutical bone scanning. This technique has been expanded by its application to a number of bony entities occurring in children — craniosynostosis, suspect cranio-spinal tumor or infection, undiagnosed back pain, and child abuse.

In a 3½ year period beginning January, 1973, 178 children were assessed with ^{99m}Tc -diphosphonate scintigraphy and production of positive scans of the appropriate skeletal part, according to their clinical diagnosis: —

Craniosynostosis	107
Skull Tumor	32
Undiagnosed Back Pain	24
Cranio-spinal infection	10
Child Abuse	5

The radiotracer evaluation of premature synostosis provides a dynamic assessment of suture closure, may indicate involvement of sutures other than those under clinical suspicion and correlates well with histologic evaluation of the excised suture. Many more primary (eg. histiocytosis, osteoma, fibrous dysplasia) than secondary tumors were uncovered by this technique, where often the scan was the only confirmatory procedure. For the child with nonradicular, atypical back pain, the yield of positive scanning was, as expected, low. On the other hand, bone scanning aided considerably in the child with suspected skull or spine infection.

In many instances, and including those of suspected child abuse, abnormal tracer patterns may be obtained at a stage when orthodox radiography appears normal, or is equivocal.

The Treatment of Stuttering with Haloperidol

T. J. Murray, P. Kelly and E. Rosenberg.
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Because of the success with haloperidol in various disorders, particularly in Gilles de la Tourette Syndrome, it was felt a long-term trial of this drug was warranted in stuttering. A double-blind cross-over study with haloperidol was carried out in 26 adult volunteers with longstanding stuttering. The components and characteristics of their speech and stuttering patterns were repeatedly evaluated and enumerated on videotape while reading a standard passage and during spontaneous speech. Detailed videotape evaluations were made at regular intervals for the following year.

Of the 18 patients who completed the trial, 11 were significantly improved on haloperidol over placebo. 3 patients benefited equally from haloperidol and placebo and 4 patients were unchanged. Improvement in stuttering occurred because of a reduction in the number of dysfluencies, increased speed of speaking and reduced secondary "struggle" aspects of stuttering.

Poor concentration, akathisia and dystonic movements caused 8 patients to discontinue the trial even though 5 of these patients had shown significant improvement on haloperidol. 12 other patients required reduction in dosage because of side effects.

Despite these promising results, we would add a cautionary note about the practicality of such therapy considering the moderate improvement expected, the seriousness of the side effects of haloperidol, and particularly the personality and attitudes of the adult stutterer. In certain stuttering patients, however, this therapy may be a useful adjunct, particularly in the initial period of a speech therapy program.

Aperiodic Intervals Between Complexes:

A Common Finding in Early Stages of SSPE?

Svetlana Ninkovic, André Merminod, Jacques Lamarche and Jean Reiher, C.H.U., Sherbrooke, Quebec, Canada

Periodic stereotyped complexes, separated by short and regular intervals are electroencephalographic hallmarks of subacute sclerosing panencephalitis (SSPE). Intervals between complexes are known to shorten during the course of the disease, from 25-30 seconds in early stages to 5-10 seconds in more advanced stages.

To date, exceptionally prolonged and irregular intervals (20-80 seconds, 36-96 seconds) have been reported only twice. The purpose of this presentation is to point out that, in the early stages of SSPE, such aperiodic complexes may be more common than heretofore reported.

Over the past 6 years, five patients with pathologically confirmed SSPE were observed. In four patients, intervals between complexes in initial waking records were irregular and prolonged, ranging from 26 to 368 seconds, 9 to 183 seconds, 19 to 107 seconds, and 16 to 107 seconds respectively. During sleep, intervals remained aperiodic, or were difficult to assess because of the changing morphology of complexes with shifting stages of sleep.

Evolution toward shorter and more regular intervals took place in three out of four patients over a two-week period. In one child, nine months have elapsed before intervals of 25"-30" were first observed; clinical deterioration was slower and more gradual than in the other three patients.

Should these findings be corroborated in larger series of patients, and should effective treatment of SSPE ever depend on early detection, the electroencephalogram could be relied upon to provide useful clues for earlier diagnosis. Aperiodic intervals between complexes represent the earliest distinctive electroencephalographic manifestations of SSPE; the longer the intervals remain aperiodic, the slower the course of the disease may be.

Horizontal Dipoles and Seizures

Spiridon Kantarjiev and Jean Reiher,
C.H.U., Sherbrooke, Quebec, Canada

The majority of spikes and sharp waves encountered in clinical practice originate from radially oriented dipoles. Discharges originating from horizontal dipoles must be exceptional, judging from the paucity of

observations on the subject. This gap in the literature prompted us to look into our own material.

Horizontal dipoles were identified in 47 out of 12,763 patients — almost exclusively in children — and were characterized by: optimal display in combined monopolar and bipolar montages; single phase reversal in exclusively monopolar recordings; double phase reversals, and steeper gradients of amplitude in straight longitudinal arrays of sequentially paired scalp electrodes; higher amplitude in bipolar than in monopolar derivations; predominant distribution along sylvio-frontal or centro-frontal axis.

An isolated finding in 36 patients, horizontal dipoles occurred in association with radial dipoles in 11; mental retardation was observed five times more often in the latter.

Seizures reported in 40 patients, were recorded as chief complaint in 30. They were isolated, infrequent, or well controlled by anticonvulsants in 34.

Ictal manifestations, insufficiently described in 2 charts, were recorded as nocturnal in 19 patients; seemingly generalized in 6; focal in 13. In 8 patients with partial seizures, ictal symptomatology correlated well with the location of the horizontal dipole; in 3, more appropriately with that of coexistent vertical dipoles; in 2, no such correlation was apparent.

In the only two patients with surgically verified lesions, the location of the lesion correlated with the positively charged pole. Generalization as to which of the two poles represents a more reliable indicator for localization of lesions must await further studies on larger groups of patients.

Les Mini-Pointes: Données Nouvelles

J. Reiher, Michel Lebel, C.H.U. Sherbrooke, Qué.,
and D. W. Klass, Mayo Clinic, Rochester, Minnesota

Les mini-pointes, ou "Small Sharp Spikes", se rencontrent uniquement pendant sommeil, presque exclusivement chez l'adulte.

A partir d'enregistrements conventionnels, nous avons, à l'encontre des Gibbs, assigné aux mini-pointes une distribution spatiale nettement plus postérieure. Nous avons également souligné la discordance entre la disparité des symptômes relevés chez les malades porteurs de mini-pointes et le caractère épileptique qu'on leur conférait néanmoins. (Reiher et Klass, 1968).

A la faveur d'enregistrements récents plus appropriés, et du calcul d'incidence des mini-pointes dans deux groupes comparables de 100 malades, épileptiques et non-épileptiques, rectifications et précisions suivantes s'imposent.

Les coordonnées topographiques consignées à date dans la littérature, incluant celles de Small (1970) et de Koshino-Niedermeier (1975), sont incorrectes. Notion inédite, les mini-pointes se distinguent de la majorité des pointes par une distribution singulière traduisant des origines indiscutables à partir d'un dipole horizontal. Des enregistrements référentiels dérivés du nasion, mettent en relief deux pôles: un pôle antérieur, négatif, limité à la région temporale antérieure, avec diffusion restreinte aux régions frontale et sylvienne ipsilatérales; un pôle postérieur, diphasique, englobant les deux régions temporales postéro-inférieures, pariétales et occipitales débordant même sur les régions temporales antérieure et moyenne contralatérales.

L'incidence des mini-pointes s'est avérée identique dans les deux groupes témoins: 25% et 26% respectivement; argument additionnel militant en faveur de la non spécificité des mini-pointes.

Il importe dès lors d'exclure ces grapho-éléments du cadre des anomalies épileptiques intercritiques, malgré des caractères morphologiques très voisins, sous peine d'interprétations fautive pouvant déboucher sur des gestes thérapeutiques inappropriés.

The Roles of the Thalamus and of the Cerebral Cortex in Feline Generalized Penicillin Epilepsy: The Relationship of Generalized Bilaterally Synchronous Spike and Wave Activity to Spindle Waves

P. Gloor, L. F. Quesney and A. Pellegrini, Montréal

In cats intramuscular injection of large doses of penicillin (300,000 to 400,000 I.U./kg) produces a transient epileptic state with clinical and electroencephalographic manifestations resembling those of human generalized cortico-reticular ("centrencephalic") epilepsy.

Bursts of bilaterally synchronous epileptic discharges resembling

spontaneous bursts can be precipitated with a high degree of probability (75 to 100%) by a single shock or low frequency electrical stimulation of brain loci which before penicillin administration produced spindle waves or recruiting responses with such stimulations. Most of the loci are located in the midline and intralaminar system of the thalamus (the "thalamic reticular system"). Other sites may upon such stimulations at times also initiate such bursts, but with a much lower probability which often, however, is still above chance level. The same results can be obtained when the electrographic syndrome of feline generalized penicillin epilepsy is reproduced by diffuse bilateral application of a weak penicillin solution to the cerebral cortex.

These findings suggest that thalamocortical volleys normally involved in spindle generation precipitate spike and wave discharges in cortex which has been rendered moderately hyperexcitable by the action of penicillin. If this is the case, spike and wave discharges induced by penicillin should be reconverted to spindle waves if the excitability of the cortical neurons involved in their production is reduced. We were sometimes successful in achieving this transformation of epileptic bursts into spindles by depressing the cortical excitability locally by the topical application of KCl which induces spreading depression, or generally by inducing cerebral anoxia.

The Effect of Na-dipropylacetate (DPA) on Generalized Penicillin Epilepsy in the Cat

A. Pallegri, P. Gloor and A. Sherwin, Montréal

Generalized penicillin epilepsy in the cat is characterized by clinical and electroencephalographic manifestations which resemble those of human generalized cortico-reticular ("centrencephalic") epilepsy. Because of the reported beneficial effect of Na-dipropylacetate (DPA) on this type of epilepsy in man, we decided to investigate its action on this animal model. The cats were prepared for chronic EEG recording in the awake state. The plasma half-life of the drug after chronic intraperitoneal administration was about 7½ hours. In 4 animals a single intraperitoneal injection of DPA (25 to 130 mgms/kg) was given one hour before the intramuscular injection of 350,000 to 400,000 I.U./kg of penicillin. The bilaterally synchronous epileptic bursts appearing in the EEG were counted for 6 hours. The plasma level of DPA was measured in blood samples taken immediately before and after the EEG recording. The number of epileptic bursts as compared to controls was reduced in each test (except one) during the 6 hour period of EEG recording. The reduction was most evident in the first 3 hours. However, no significant correlation was found between plasma DPA level, or dose injected, and the reduction of epileptic bursts. In 2 cats DPA was administered 3 times a day for 2½ days (25 to 100 mgms/kg per dose). The incidence of epileptic bursts was reduced not only during, but for many days after the end of DPA administration, at a time when presumably all drug had been excreted or metabolized. This effect was proportional to the drug dose.

The Ocular Tilt Reaction A Paroxysmal Dyskinesia

J. A. Sharpe, H. E. Rabinovitch and T. O. Sylvester, Toronto

Paroxysmal ocular motor disorders are illustrated by oculogyric crises, superior oblique myokymia, and familial intermittent nystagmus. Paroxysms of somatic motor and sensory dysfunction and diplopia occur in the course of multiple sclerosis (Matthews, W. B., J. Neurol. Neurosurg. Psychiat. 38: 617, 1975). We describe a unique synkinesis of eye and head posture, the ocular tilt reaction, in a patient with presumed multiple sclerosis. This stereotyped movement pattern is evoked in the monkey by unilateral midbrain tegmental stimulation dorsolateral to the oculomotor and trochlear nuclei (Westheimer, G., Blair, S. M., Exp. Brain Res. 24: 89, 1975; Invest. Ophthalmol., 14: 833, 1975). Attacks of conjugate ocular torsion, skew deviation and head tilting in the patient occurred thirty to forty times daily and lasted from seven seconds to approximately five minutes. The attacks exemplify the ocular tilt reaction in man.

The head tilt and ocular torsion in this synkinesis are not compensatory; they occur in the same direction. The 12 o'clock corneal meridians are rotated toward the lower skewed eye and the head is inclined laterally in the same direction. The ocular tilt reaction may be explained by paroxysmal activation of brain stem otolithic vestibular projections.

Infrared oculographic analysis showed low amplitude conjugate vertical and horizontal pendular oscillations during attacks. A phase difference of 114° and amplitude disparity between the vertical (2-4°) and horizontal (1-2°) oscillations produced elliptical nystagmus. Midbrain stimulation in the monkey elicits conjugate vertical and horizontal eye movements that displace the eyes in curved trajectories, analogous to the episodic elliptical nystagmus which accompanied the ocular tilt reaction. This distinctive brain stem synkinesis of conjugate ocular torsion, skew deviation and head tilting was treated effectively with carbamazepine.

Deriving Postsynaptic Potentials in Single Spinal Motoneurons in Man

Peter Ashby, Toronto, Ontario

The contour of the postsynaptic potential (PSP) produced by afferent stimulation can be derived from the contour of the post stimulus time histogram (PSTH) of a rhythmically firing motoneuron. The relationship is not a simple one. For example the contour of the PSTH resembles the first derivative of the rising phase of the PSP rather than the PSP directly.

In the present study the PSTH of the firing of individual soleus motor units following stimulation of the popliteal or peroneal nerve was used to explore the effects of extensor and flexor group I afferent volleys on the excitability of single soleus motoneurons in man.

The action potentials of 30 voluntarily activated motor units of soleus were recorded with concentric needle electrodes in 7 subjects. Individual action potentials were selected using a window discriminator and a delay line. A laboratory computer was used to extract the action potential by repeated averaging and to generate interval data including the PSTH.

Extensor group I volleys resulted in an early peak of increased impulse density in the PSTH of 75% of soleus motoneurons. The latency suggests an analogy with the Ia EPSP. The mean duration of the peak of increased impulse density, equivalent to the rise time of the EPSP, was 3.6 msec. Flexor group I volleys resulted in a transient reduction in the excitability of soleus motoneurons. The latency suggests an analogy with the Ia IPSP.

It is suggested that this method could be used to derive some of the characteristics of postsynaptic potentials in single motoneurons in man.

Decorticate Spasticity: Effects of Dentate and Ventrolateral Thalamic Lesions on Stretch Reflexes in the Cat and Squirrel Monkey

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Eight cats and three squirrel monkeys (*Saimiri sciureus*) were made spastic by bilateral ablation of the primary and supplementary motor areas. The activity of their stretch reflexes in both flexors and extensors was evaluated clinically, as well as by the integrated electromyographic (E.M.G.) method. The effects of stereotaxic lesions in the ventrolateral (V.L.) nucleus of the thalamus and the dentate nucleus of the cerebellum were followed for four to sixteen weeks.

Clinical and E.M.G. evidence suggested that VL thalamotomy had no appreciable effect on the spasticity of the bilaterally decorticated animal, whereas dentatectomy had a significant ipsilateral effect in reducing the degree of spasticity.

These findings suggest that cerebro-cerebellar mechanisms are involved in the development of spasticity, and that the dentate nucleus may affect muscle tone via pathways that do not pass through the thalamus.

Cardiac Biopsy in Kearns-Sayre Syndrome

Yadollah Harati, Bernard M. Patten, Mark Sheehan, David Judge and Jeanie M. Wood

From the Department of Neurology, Baylor College of Medicine, Texas Medical Center, Houston, Texas

We report the histochemical and electron microscopic findings from a cardiac muscle biopsy obtained during insertion of a pacemaker in a

14-year-old boy with typical clinical and laboratory features of Kearns-Sayre syndrome (progressive external ophthalmoplegia, atypical retinitis pigmentosa, heart block, ataxia, deafness, mental retardation). In cardiac muscle, we noted on modified Trichrome, abnormal subsarcolemmal and intrafibrillary red stained material ("ragged-red" fibers) with vacuolization and disruption of myofibrils and excessive succinic dehydrogenase activity in the same regions. Abnormal collections of large mitochondria similar to those seen in the patient's skeletal muscle biopsy were present in the cardiac muscle on electron microscopy. Studies of mitochondria, isolated from skeletal muscle, showed mitochondrial respiratory rate reduced to about 30% of normal. Oxidative phosphorylation was partially uncoupled with respiratory control index of 2, but ADP/O ratio was normal at 3.08. Lactate infusion test (Patten, Neurology 24:986, 1974) gave normal results. Since similar mitochondrial abnormalities have been produced in thiamine deficient rats, we treated the patient with 300 mg thiamine hydrochloride daily. Hand writing, tremor and ataxia improved.

We conclude that in Kearns-Sayre syndrome cardiac muscle demonstrates morphologically and histochemically abnormal mitochondria similar to those found in skeletal muscle.

Cerebral Metabolism and Blood Flow During Surgical Anesthesia with Thiopentone

D. Simard, J. Cote, M. Rouillard & R. Langelier, Québec

Thiopentone anesthesia has been known for some time to induce a diminution of cerebral metabolism and of cerebral blood flow (CBF)^{1,2}. It was advocated more recently to be useful in protecting the brain against ischemia and increased intracranial pressure^{2,3}. However, it is difficult to conclude from these studies if thiopentone effect of CBF is really safe for the brain in surgical anesthesia, because either a vasodilating anesthetic agent was concomitantly used or because the patients were artificially ventilated. Moreover, in all these studies, except in the recent work of Carlson & al.⁴ in the rats, it is impossible to determine the rate of intravenous infusion of thiopentone.

As we had recently demonstrated that in man an infusion of 0.4% thiopentone given intravenously at a rate of 0.6 ml/kg/min. could achieve a stage of surgical anesthesia, ten patients without significant brain lesion were anesthetized using this method. Regional cerebral blood flow (rCBF) and cerebral metabolic rate of oxygen (CMRO₂) were determined before the onset of anesthesia and at 20, 40 and 60 minutes after induction; carotid and jugular blood levels of thiopentone were also assessed.

The CMRO₂ decrease was of 58% after a steady state of anesthesia with thiopentone, whereas the observed reduction in CBF was only 30%; hence with this method of anesthesia the CO₂ retention did increase the PACO₂ of 10 mm Hg., thus still allowing a reduction of CBF not too severe and still beneficial for the patients with increased intracranial pressure. No focal change of rCBF were found in these patients during this thiopentone anesthesia.

It is concluded that in the anesthesia for neurosurgical procedures thiopentone proves to be an extremely desirable anesthetic agent as during spontaneous ventilation the increase of PACO₂ tends to maintain the CBF at higher values than expected with the marked fall of CMRO₂ thus providing good brain function protection and still permitting a reduction of intracranial pressure.

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Chronic Occult Adult Hydrocephalus Presenting as Chiasmal Syndrome

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Dementia, ataxia and urinary incontinence, the classical "triad of Hakim", are now recognized as the clinical expression of occult adult hydrocephalus. The rate at which the symptom complex develops indicates that the communicating hydrocephalus associated with it develops subacutely. The authors have observed and treated two

patients in whom we have shown the same disorder in CSF circulation to exist but, because the resorptive mechanisms failed over a more prolonged period of time, i.e. chronically instead of subacutely, the clinical symptom complex produced differed from the "Hakim triad". In both cases, adults presented with headache and progressive bitemporal upper quadrantic visual field defects. Both were referred as possible cases of pituitary tumor and because of progressive deterioration in visual fields, one was explored surgically for chiasmal lesion. Conventional investigations were used to establish the diagnosis, and in one patient "plateau" waves were recorded when intracranial pressure was monitored. Both patients responded to treatment by ventriculoatrial shunt with arrest or resolution of visual field defects and relief of headache. A model for the pathophysiological substratum of this disorder will be proposed.

Evaluation de 9 malades avec sclérose en plaques (SEP) traités par ACTH et sérum antilymphocytaire (SAI) avec follow up de 4 ans

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Neuf malades dont le diagnostic de sclérose en plaques était établi de façon certaine sur le plan clinique, ont été soumis, lors d'une poussée, à un traitement à base d'ACTH et de sérum antilymphocytaire. Evaluation clinique mensuelle au cours de l'année qui suivit le traitement, faite selon l'échelle de Kurtzke. Les malades ont été revus par la suite périodiquement. Une ponction lombaire a été faite, avec dosage quantitatif des globulines, à tous les mois également durant la première année à la suite du traitement. Commentaires sur les fluctuations quantitatives des globulines par rapport à l'évolution de la maladie.

Les auteurs ont analysé les effets secondaires du traitement et l'évolution clinique de la maladie au cours des 4 années subséquentes. Comparaison des résultats obtenus chez ces neuf malades avec les données rapportées par les autres auteurs ayant étudié l'effet du sérum antilymphocytaire dans la sclérose en plaques.

Pallidofugal Projections to Thalamus and Midbrain in the Cat: An Electrophysiological Study

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Classically, pallidothalamic fibers, to ventral lateral-ventral anterior (VL-VA) and centromedian (CM) nuclei, constitute the major outflow of the striopallidal system. Pallidofugal fibers to extrathalamic structures, such as the tegmental pedunculopontine (TPP) and lateral habenular (HbL) nuclei, were also described, but their relative importance remains a matter of speculation. The present study was undertaken to determine the proportion of pallidofugal fibers distributed to each of the aforementioned projection sites and the extent of axonal branching within this fiber system. In cats, under pentobarbital anesthesia, extracellular unit activity was recorded in the entopeduncular (Ent) nucleus, homologue of the internal division of the primate globus pallidus. Stimulation electrodes were placed along pallidofugal fibers, at the level of VL-VA, CM, TPP and HbL nuclei, and antidromic responses of Ent units were sought for. VL-VA, CM and TPP were equally effective stimulation sites; each giving rise to antidromic responses of more than 50% of Ent neurons. Moreover, stimulation of the three sites produced antidromic responses of the same neuron in at least 30% of the cases. In contrast, only 25% of Ent neurons responded to HbL. This suggests that the striopallidal system, involved in the control of motor activity, makes use of the midbrain nucleus TPP, as well as of thalamic nuclei VL-VA and CM, to modulate motor neurons activity. Nucleus TPP receives a significant projection from the motor cortex. Moreover, it is located in the area of the mesencephalic locomotor center, the stimulation of which gives rise to locomotion in decerebrated cats. In other respects, nucleus HbL, involved in the limbic system, receives projections from few Ent neurons.

Pathological Findings in Disulfiram (Antabuse) Neuropathy

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Peripheral neuropathy as a complication of therapeutic doses of disulfiram may be overlooked in alcoholic patients.

We studied a 35-year-old man who developed peripheral neuropathy after taking disulfiram 500 mg. daily for four months. He complained of unsteadiness and of distal pain and numbness. He had weakness of distal muscles in the upper and lower extremities. The knee jerks were reduced and the ankle jerks absent. Pin prick and joint position were reduced distally. Touch threshold was 0.2 gm. (normal) and vibration threshold 12 μ m (raised) on the toes. The amplitudes of the evoked sensory action potentials and muscle compound action potentials were reduced. Motor nerve conduction velocities were at the lower limits of normal and the distal latencies were only slightly prolonged. Denervation potentials were detected in distal muscles. A right sural nerve biopsy revealed a large number of degenerating myelinated fibers of all diameters. There were numerous examples of early axonal degeneration with disappearance of microtubules and neurofilaments from the axoplasm. A greater number of degenerating fibers were seen on electron microscopy than could be suspected on high magnification light microscopy. There was no evidence of segmental demyelination. There was very little evidence of old axonal degeneration.

Two weeks after disulfiram was discontinued his symptoms began to improve and 4 weeks later distal weakness was less marked, sensory thresholds, conduction velocities and distal latencies had returned to normal.

The Reduction of CCNU Toxicity by Phenobarbital Pretreatment in a Murine Brain tumor model

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Nitrosoureas such as CCNU have been highly effective in the chemotherapy of experimental brain tumors. However, only minimal benefit has been demonstrated in human cerebral gliomas. The nitrosoureas produce hematopoietic toxicity which limits the amount of the drug which can be administered. It is well known that phenobarbital (PB) causes induction of microsomal enzymes and it has been recently shown that nitrosoureas are degraded by murine liver microsomes. Thus, to enable larger doses of CCNU to be administered we have studied the effects of PB pretreatment in an attempt to reduce CCNU toxicity.

In non-tumor bearing mice the calculated LD₅₀ of intraperitoneal (IP) CCNU was 51 mg/kg and 90% of the toxic deaths occurred within 10 days of CCNU administration. CCNU 40 mg/kg IP resulted in a 50% toxic death rate; CCNU 60 mg/kg IP resulted in a 75% toxic death rate. When PB was given at 125 mg/kg IP daily for 4 days prior to CCNU at 60, 90 or 120 mg/kg IP the toxic death rate was reduced to 0, 5 or 9% respectively ($p < .01$).

In brain tumor bearing mice 80 mg/kg CCNU IP caused a 92% toxic death rate, but pretreatment with PB at 25, 50 or 75 mg/kg IP daily for 4 days prior to the CCNU reduced the toxic death rate to 28, 36 or 8% respectively ($p < .01$). Furthermore, the groups receiving the CCNU-PB combination had a 150% increase length of survival over control mice ($p < .01$).

These findings show that PB pretreatment will reduce the toxicity of CCNU without eliminating its tumoricidal properties.

Intracavernous Aneurysms Originating from the Intradural Carotid Artery

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Five cases with a unique, but previously unclassified type of aneurysm are presented. These aneurysms originate from the inferior aspect of the internal carotid artery at the same level as the ophthalmic artery, but on the opposite surface. Three of the cases were associated with other intracranial aneurysms, including one with the same anomaly bilaterally. This finding and the angiographic appearance suggest that

these are congenital berry aneurysms, although atherosclerotic dilations may occur at this site. Four of the patients presented with a subarachnoid hemorrhage, two, or possibly three, probably originating from the aneurysm under discussion. In all cases, the pre-operative angiogram left doubt concerning the relationship of the aneurysm to the cavernous sinus. This doubt, and angiographic suggestion of a clippable neck, lead to surgical exploration in all cases. At operation, it was difficult to fully define the necks of the aneurysms because they were at least partially incorporated in the dural roof of the cavernous sinus. The dissection resulted in operative aneurysm rupture and early post-operative death in two of the patients. In the two other cases where a clip was applied to the aneurysm neck, post-operative angiography showed only partial obliteration of the lesion. Nevertheless, because the situation is difficult to define angiographically and direct repair may be possible in some cases, exploration is justifiable, but, with a planned alternative procedure should the primary objective be uncertain or impossible.

Smooth Ocular Pursuit: The Effect of Age on Horizontal Tracking

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The smooth pursuit system provides a sensitive parameter of brain function. Defective smooth pursuit, evidenced clinically by the presence of catch-up saccades while tracking slowly moving objects, is the most common ocular motor sign of cerebral, brain stem or cerebellar dysfunction. In order to establish criteria for the diagnosis of pursuit paresis, smooth pursuit system gain was measured in normal adults, young and elderly.

Constant velocity targets, provided by a low intensity rear projected laser, were driven horizontally in predictable triangular waveforms through a visual angle of 20 degrees at nine frequencies, from 0.125 to 2.5 hertz. Eye movements were recorded by infrared reflection technique with the subjects' heads fixed. Smooth eye movements (SEM) velocities were derived by electronic differentiation of eye position signals with sensitivity sufficient to detect saccades smaller than 10 minutes of arc. Fifteen young (age 15 to 45) and ten elderly (age 65 to 80) adults served as subjects. For each subject, fifteen optimum SEM velocities were utilized to obtain mean smooth pursuit velocity/target velocity gain at each frequency.

In the group of young subjects, mean smooth pursuit velocity gain approach unity (>0.96) at target velocities up to 30°/sec and a maximum mean SEM velocity at $39.5 \pm 9.0^\circ/\text{sec}$ occurred at a target velocity of 50°/sec. In elderly subjects, gains declined progressively below an optimum mean value of 0.89 for 5°/sec targets and a maximum mean SEM velocity of $17.6 \pm 5.2^\circ/\text{sec}$ occurred at 40°/sec target velocity. There were large intersubject variations within each age group. Smooth pursuit gain in the elderly was significantly lower than in young subjects for target velocities over 20°/sec ($p < 0.001$). The results indicate that the diagnosis of pursuit paresis in neurologic disease must be qualified by the age of the patient. Smooth pursuit is an age dependent motor system.

A New Computerized Spike Recognition Programme: Its Use at Electroencephalography

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We have designed a computerized spike recognition programme whose method differs significantly from others thus far reported. It identifies a spike when the first derivative of the ongoing EEG signal exceeds a user-defined critical value twice within 80 milliseconds, one derivative value being opposite in sign (i.e. polarity) to the other. As this method does not rely on computation of running averages, the computer is free for other tasks. Thus, it can detect spikes in 16 referentially recorded channels simultaneously. Moreover, it determines where a widely synchronous spike occurs first, counting it only in that channel. The quantity of spikes in each channel is displayed on-line by proportionately sized circles on a 4 by 4 grid array for electroencephalography and on a graphic representation of the ten-twenty system for scalp recording.

At electrocorticography, the electroencephalographer compares his rank order of visually assessed spike incidence among the 16 EEG channels with that of the computer. When results differ, the E.Co.G. is immediately reviewed, paying particular attention to each incidence of discrepancy between computer recognition (as depicted by a marker on the EEG) and visual assessment.

Our system, using only a mini computer, achieves an electroencephalographer-programme correlation of 80-90%.

Intravenous Phenytoin and Phenobarbital: Brain Content and Anticonvulsant Activity

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This study was performed to clarify the nature of the previously reported 30 minute delay to peak anticonvulsant activity of intravenous phenytoin in animals, and compare its effective brain and plasma free concentrations with those of phenobarbital. ^{14}C -phenytoin or ^3H -phenobarbital was given through indwelling jugular catheters to 65 rats and anticonvulsant activity tested by the maximal electroshock seizure test. Drug concentrations were measured by standard isotopic techniques. Free and total plasma drug levels were determined by equilibrium dialysis.

The median effective cerebral phenytoin concentration (EC_{50}) was $10.5 \mu\text{M}/\text{kg}$ (95% fiducial limits, 8.2 to 12.4) three minutes after infusion compared with $10.2 \mu\text{M}/\text{kg}$ (7.1 to 13.0) 30 minutes after infusion. The EC_{50} of phenobarbital was $8.2 \mu\text{M}/\text{kg}$ (6.7 to $9.3 \mu\text{M}/\text{kg}$) three minutes after infusion. Cerebellar concentrations were equivalent to cerebral concentrations for all rats ($r = 0.98$). Three minutes after infusion, cerebral:plasma free ratio of phenytoin was 3.73 ± 0.71 ($\pm \text{S.D.}$); the plasma protein bound:free ratio 3.70 ± 0.98 . For phenobarbital, the cerebral:plasma free ratio was 0.72 ± 0.10 ; the plasma protein bound:free ratio 0.63 ± 0.12 .

Since the EC_{50} values of phenytoin three or 30 minutes after infusion did not differ, onset of anticonvulsant effect clearly occurred as soon as adequate brain concentrations were attained; delay to peak activity may represent time required for maximum uptake. Phenobarbital was effective three minutes after infusion, and effective brain concentrations were similar to those of phenytoin. Brain content paralleled plasma protein binding, both being high for phenytoin and low for phenobarbital.

Stereoencephalographic (SEEG) Study of Four (4) Cases of Primary Amygdalo-Hippocampal Epilepsy

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The temporal limbic system is thought to be frequently involved at the origin of complex partial seizures, although some authors think that this structure is rarely the site of an independent epileptic focus. By using a very precise system to record epileptic seizures on the scalp and with depth-electrodes, it was possible to make exact anatomical, electrical and clinical correlations (Union Méd., Canada — 105, oct. '76, p. 1538-1541).

We were able to study four (4) cases of primary amygdalo-hippocampal seizures: three from the hippocampus and one from the amygdala. To differentiate the primary involvement of the amygdala from the hippocampus, we have studied various parameters: the slow and epileptic activity from the scalp and from the depth-electrodes, the ictal activity during numerous spontaneous seizures from the scalp and from the depth-electrodes, the results of electrical stimulation, the detailed clinical analysis of spontaneous seizures and of symptoms induced by electrical stimulation.

Some clinical, EEG and SEEG characteristics were detected which could help to distinguish between the primary amygdala-epilepsy and the primary hippocampal epilepsy.

In the primary hippocampal epilepsy, the EEG slow activity tends to appear more diffusely over the involved hemisphere and the epileptic activity takes the bilaterally-synchronous form although predominating over the involved hemisphere. In the primary epilepsy of the amygdala,

the epileptic and slow activity is more localized on the corresponding temporal lobe. In depth, the hippocampal epilepsy gives rise to ictal activity which spreads more diffusely, particularly to the cingular and frontal cortex.

Clinically, the study of spontaneous seizures and electrical stimulation showed that the "fear" sensation and the "chewing" movements originate probably from the amygdala while the "nausea" sensation comes from the hippocampus.

Visual Contrast Sensitivity in Multiple Sclerosis

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Despite normal or near normal visual acuity on conventional testing, a substantial number of M.S. patients complain of blurred or "washed out" vision. Using a principle for producing better lenses for military aerial photography, we tested 38 M.S. patients for low, medium and high contrast sensitivity.

A sinewave grating of dark and bright bars was projected on a cathode ray tube and the threshold for contrast sensitivity assessed at various spatial frequencies. Similar tests were done on 29 age and sex matched controls. Ten other patients were tested for the effect of optical blurring using a 1+ diopter lens to assess the effect of refractive error.

Six patients showed loss of visual contrast over all spatial frequencies. This could not be corrected by trial lenses. Eleven patients had preferential loss of contrast sensitivity at high and medium spatial frequencies, similar to the pattern with refractive error, but this also could not be corrected with lenses.

The most significant group was 9 patients with sensitivity loss restricted to medium frequency contrast. Most of these patients had normal vision by conventional examination and testing. The finding of a specific loss for medium frequency contrast has important physiological implications and provides an answer for why M.S. patients with "normal" vision may complain about visual difficulty.

Photosensitivity and Generalized Convulsive Epilepsy

B. Sternberg and G. Patry

A systematic and comparative study of generalized convulsive photosensitive and non-photosensitive was undertaken.

All patients presenting generalized convulsive seizures and potentially epileptogenic activity induced by photic stimulation and seen at Hôtel-Dieu du Sacré-Coeur between 1970 and 1975 were studied and compared with a group of epileptics presenting with generalized non photosensitive epilepsy. The two groups were matched for sex and age. Clinical and EEG data were analyzed by computer in order to try to differentiate both types of epilepsy.

1. Age on onset of seizures is the same for both groups.
2. Early in the course of their epilepsy seizure frequency is identical for both groups while it increases later on for photosensitive epilepsy which is evidence for the possibility of a certain degree of refractiveness to anticonvulsant therapy in this group.
3. Normal I.Q. is found less frequently in photosensitive patients than in those who are not photosensitive.
4. Birth trauma and febrile convulsions is more frequently seen in photosensitive epilepsy than in our control group. On the other hand, genetic transmission seems to be identical for both photosensitive and non photosensitive generalized epilepsy.

Post-Discharges Epileptiques du Type Pointe-Onde des Interneurones Corticaux et Thalamiques

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Des enregistrements de décharges unitaires et d'ondes lentes focales ont été effectués chez des singes et chats à l'aide de microélectrodes au niveau des aires corticales motrice et somatosensorielle, ainsi que dans le complexe thalamique latéral postérieur. Les neurones de projection ont été identifiés par invasion antidromique; les interneurones ont été

reconnus par leur décharges spontanées en bouffées de spikes et par leurs réponses synaptiques à haute fréquence. Des phénomènes paroxystiques auto-soutenus, du type pointe-onde, ont été sélectivement induits dans les interneurones après les réponses augmentantes évoquées par stimulation répétitive (10/sec) des voies afférentes spécifiques thalamo-corticales et cortico-thalamiques. Des longs trains de spikes à haute fréquence, avec inactivation progressive, accompagnaient la "pointe" du complexe, négative en profondeur du cortex et dans le thalamus; les décharges interneuronales récupéraient partiellement leur amplitude en relation avec le commencement de l'"onde" du complexe, composante de longue durée et de polarité positive; enfin, vers le sommet de l'"onde" les décharges interneuronales cessaient complètement. Ces données attirent l'attention sur la susceptibilité préférentielle des interneurones dans la genèse de l'épilepsie avec complexes pointe-onde et sur l'aspect très similaire de ces complexes (avec des composantes dépolarisantes rapides et une onde hyperpolarisante de longue durée) dans le thalamus et différentes aires corticales.

Improvement from Alcoholic Organic Brain Syndrome — Early EEG Changes Predict Degree of Psychologically and Clinically Assessed Recovery

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Forty chronic alcoholics, hospitalized for investigation of an organic brain syndrome, were followed by serial EEG recordings. Psychometric assessments were done at least two weeks after the last drink to avoid withdrawal effects and three months later in those alcoholics who had abstained and were available for retesting. Scored neurological assessments were done weekly. Computerized Transaxial Tomography (CTT) showed cortical atrophy and ventricular dilatation in most cases. Clinically some recovery from the chronic alcoholic organic brain syndrome with abstinence was noted in 29 of the 40 patients and marked improvement was seen in 8 patients.

The EEGs were read by an investigator who had no knowledge of the patients' clinical state or degree of recovery. Over three to six weeks there was a statistically significant increase in mean alpha frequency of patients who showed mild or marked clinically assessed recovery. Conversely, no significant change in mean alpha frequency was observed in the eleven patients who showed no clinical evidence of improvement. The most sensitive psychometric indicator of improvement was the change in the Memory Quotient score, comparing the 2-week post-admission score with that done 3 months later. This change correlated at the $P < .02$, $r = .54$, level with the change in the mean alpha frequency noted within the first month. Therefore, the degree of increase of EEG alpha frequency noted in the first month with abstinence can help to predict the degree of recovery from the chronic alcoholic organic brain syndrome. Except for two paradoxical results, greater measured cerebral atrophy on the CTT scan correlated with less clinical and psychological improvement.

Compound Narcolepsy Treated with Nocturnal Gamma-Hydroxy-Butyrate

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Eight narcoleptics aged 22-53 years with irresistible sleep attacks plus cataplexy, with or without other symptoms of the "tetrad" were treated with nocturnal gamma-hydroxy-butyrate (GHB). Three were very serious narcoleptics poorly controlled by various combinations of stimulants and REM-sleep suppressant "anti-depressant" medication. All were withdrawn from previous medication for 2 weeks or more before GHB treatment began. Twenty-four hour polygraphic recordings were performed before and at various intervals after onset of GHB therapy. GHB was administered orally at nighttime to consolidate the fragmented nocturnal sleep of narcoleptics. Usually 2.25 gm was taken hs and repeated doses of 1.5 gm with patient awakenings, if there was 3 or more hours since the previous dose. A total nocturnal dosage of 3.75-5.25 gms was usual. No daytime treatment was given.

On GHB, patients over the first 3-7 days essentially lost their main symptoms of sleep attacks, cataplexy and sleep paralysis or vivid

hypnagogic hallucinations, if present. Nighttime sleep was more refreshing and patients felt improved mood and ability to cope in the daytime. Only mild to moderate improvement of daytime drowsiness was noted on self-assessed Stanford Sleepiness Scores. Polygraphic recordings showed a marked reduction of daytime sleep, which was abolished in some patients. In the overnight period there was reduced wakefulness, fewer movements, fewer stage shifts, some increase in REM sleep and a substantial reduction towards normal densities of the rapid-eye-movements themselves. The sleep onset REM period was preserved. Clinical improvement has been maintained up to 9 months without any serious side effects.

The Electromyographic Usefulness of the Tibialis Posterior Muscle

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Differentiation between peroneal palsy and L5 radiculopathy represents a significant neurological problem. Clinical experience underlies the value of testing the function of this muscle in providing a solution i.e. involvement indicating the presence of radicular dysfunction. As such one would assume that extensive use would be made of this muscle electromyographically. However review of the literature reveals that this has been a relatively ignored muscle electrically possibly for reasons of "presumed inaccessibility".

A retrospective study was undertaken in an attempt to delineate the potential discriminatory usefulness of this muscle. Patients selected were those who had been referred for electromyographic examination of suspected neurological dysfunction of one or both lower extremities and in whom needle electrode assessment of the tibialis posterior as well as other anterior group (tibialis anterior, peroneal group and extensor digitorum brevis) and posterior compartment (medial and/or lateral gastrocnemius) muscles had been performed. The electrical parameters assessed were the presence of spontaneous activity (fibrillation and/or positive waves — indicating denervation) plus neurogenic alterations of the voluntary motor unit potentials.

Results demonstrated that three groups were easily separated. Group A consisted of 41 examinations (37 patients plus 4 bilateral). Abnormalities were demonstrated solely in the anterior group indicating peripheral i.e. peroneal nerve dysfunction. Group B consisted of 31 examinations (31 patients).

Abnormalities were detected equally in all of the muscles indicating diffuse nondiscriminative involvement i.e. radicular or peripheral nerve. Group C consisted of 34 examinations (32 patients plus 2 bilateral). Abnormalities were detected in the posterior compartment or the anterior group as well as the tibialis posterior. The majority of patients demonstrating marked alterations in this latter muscle showed corresponding alterations anteriorly, thus indicating a shared i.e. L5 radicular innervation.

Thus this muscle represents a useful source of electrical information when attempting to delineate between peripheral (peroneal nerve) and radicular (L5) dysfunction.

Uptake of Dopamine and 5-Hydroxytryptamine (5HT) by Platelets from Patients with Huntington's Chorea (HC)

D. R. McLean and T. Nihei

Platelets from Huntington's chorea patients have been reported by Aminoff et al. (*The Lancet*, 2, 1115, 1974) to show abnormally high capacity in 5HT and dopamine uptake. Because of its potential use in detecting pre-clinical HC, we attempted to confirm this observation.

The normal ranges of uptake were determined using 20 blood samples from 17 normal people. Age (between 20 and 55 years) or sex did not affect the uptake levels. The blood samples from 6 clinically confirmed patients (2 males and 4 females) were compared with normal. The age of the patients ranged between 42 and 74 years. Except the 74-year-old patient who was treated with Thioridazine, the individuals were not receiving any medication.

At dopamine concentrations from 10 nM to 200 nM, dopamine uptake by patients platelets were statistically higher although there was no difference at 1 and 5 nM concentrations. The 5HT uptake was the same for normal and patient's platelets at all 5HT concentrations tested

(0.5-50 nM). Dopamine uptake by platelets shows promise as a diagnostic tool in the detection of pre-clinical HC.

High Prevalence of Epileptic "Risk Factors" in Children with an Initial Brief Febrile Seizure

Peter R. Camfield, Carol S. Camfield and Katherine Metrakos

Four percent of all children have a febrile seizure but only 3% of these children develop epilepsy. Several clinical and electroencephalographic features may indicate which children with an initial febrile seizure are at increased risk for future epilepsy. In order to assess the prevalence of these "risk factors" we have evaluated 154 consecutive children presenting to an emergency room with their first brief febrile seizure. An EEG, detailed history and physical examination were performed in all cases 2 to 6 weeks following the seizure. A lumbar puncture was performed in 92%. Demographic features suggested that this sample was characteristic of children with febrile seizures in general.

Our analysis shows that 52.6% of these children had at least one of the supposed "risk factors" for epilepsy. These included age less than one year 19.0%, cluster of seizures 13.0%, focal seizures 11.7%, family history of epilepsy 8.5%, epileptic EEG abnormality 5.8%, encephalitis 5.3%, perinatal problems 3.0%, and neurological abnormalities 3.0%.

These "risk factors" lack specificity (many "false positives") and the literature suggests they also lack sensitivity (many "false negatives"). An individual child's prognosis cannot be accurately assessed by these factors, however they may be used in each patient to indicate areas for more intensive evaluation.

Chirurgie Stereotaxique Unilaterale et Peripherique du Torticolis Spasmodique

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Le torticolis spasmodique est une dystonie tardive étroitement liée au tremblement d'attitude familial héréditaire. Dans une série de 12 cas rapportés précédemment, la chirurgie stéréotaxique unilatérale, au niveau de VOI avec ou sans lésion pallidale surajoutée, est efficace dans plus de 70% de ces cas. La lésion pallidale est ajoutée lorsqu'on obtient pas la diminution ou l'abolition de la décharge électromyographique peropératoire lors du regard latéral du patient. Par contre, les lésions bilatérales doivent être évitées puisqu'elles peuvent résulter en manifestations pseudobulbaires dans plus de 10% de ces cas étant donné que VOI est situé dans le même plan coronal que les fibres cortico-bulbaires.

En plus d'étudier attentivement les décharges électromyographiques des groupes sterno-cleido-mastoidiens et splénius-complexus pour déterminer le côté et l'importance réciproque des groupes musculaires impliqués, ces points sont vérifiés par un blocage temporaire, soit du spinal ou de C1 et des rameaux postérieurs de C2, C3, C4.

Après la chirurgie stéréotaxique, s'il persiste certains mouvements, ces blocages sont répétés et les branches du nerf spinal ou des rameaux postérieurs en cause sont sectionnées chirurgicalement. Chez un jeune patient la simple section du spinal d'un côté de C1 et des rameaux postérieurs de C2, C3, C4 de l'autre a donné un résultat très satisfaisant. Pour un autre patient, chez qui le torticolis spasmodique est apparu quelques années après une chirurgie stéréotaxique qui avait supprimé le tremblement d'attitude sur un côté, une lésion de VOI n'avait donné qu'une amélioration temporaire. Une section ultérieure du spinal a supprimé le torticolis spasmodique.

Cette technique combinée qui a été appliquée comporte des risques minimes pour le patient. En évitant la laminectomie et l'ouverture de la dure-mère, la section sélective de C1 et des rameaux postérieurs de C2, C3, C4 restreint l'atrophie aux muscles intéressés et diminue considérablement la convalescence. D'autre part, avec l'abandon des lésions thalamiques bilatérales, les troubles pseudobulbaires sont virtuellement éliminés.

Five Year Followup of Multiple Sclerosis Patients Treated with Anti-Thymocyte Globulin

L. F. Kastrukoff, D. R. McLean and T. A. McPherson

Anti-lymphocyte (ALG) and anti-thymocyte (ATG) globulin have

been used in the treatment of acute relapses of multiple sclerosis as well as in the treatment of chronic progressive multiple sclerosis. In 1974, Seland et al., using a double blind controlled trial, reported favourable results with ATG in the treatment of acute relapses of multiple sclerosis. Of the ATG treated group, a number of patients showed significant and continued benefit at the termination of the study, one year after treatment had been completed. Those patients in the ATG treated group who had markedly elevated CSF gamma globulin levels (> 20%) appeared to do particularly well.

The present study evaluated those patients treated by Seland et al. five years ago to ascertain any long term benefit from the use of ATG. A standard neurological examination was performed on each patient and the results obtained were used to score the patient in eight functional categories. A single disability status score was then derived from these scores. Statistical evaluation of the results was performed using t-tests and analysis of covariance.

Of the twenty patients in the Seland study, seventeen have been evaluated, two will be evaluated in the near future, and one has been lost to follow-up. The patient lost to follow-up was in the control group.

Preliminary statistical analysis indicates no significant difference between the pre-treatment score and the score at five year follow-up in the control or ATG treated group. There is also no significant difference between these two groups at five year follow-up.

Of those patients in the ATG treated group who showed significant benefit at one year follow-up, five of the six patients have been evaluated. Our results indicate no significant difference between the pre-treatment and five year score in this group. In addition, there was no significant difference in pre-treatment and five year scores in the ATG treated group with markedly elevated CSF gamma globulin.

Based on the five year follow-up of patients treated for acute relapses of multiple sclerosis, there appears to be no long term benefit from the use of anti-thymocyte globulin. Notably, those patients with elevated gamma globulin in CSF, who appeared to be significantly improved following ATG therapy, failed to demonstrate any lasting benefit.

CBF Studies Before and After Cerebral Revascularization

H. Schutz, F. Taylor, M. Chiu and K. Ter Brugge

Nine patients have been selected for extracranial to intracranial arterial anastomosis on the basis of their clinical presentation, their radiological findings and their regional cerebral blood flow (rCBF) determinations. rCBF was measured using the intracarotid injection of Xe^{133} and calculating the washout of the isotope from 8 regions of the cerebral hemisphere.

In all patients selected for revascularization there was a focal or global reduction of rCBF (below 30 ml/100 gm/min) prior to surgery. Three patients have been studied three months after the operation and there has been an improvement in rCBF (over 45 ml/100 gm/min) in two of the three patients. The indications for these procedures are still controversial. However, a rational approach would be to take into account the clinical picture, the radiological examination and rCBF determinations.

Batten Disease: Progress and Prospect

Leonhard S. Wolfe, N. M. K. Ng Ying Kin, Eva Andermann, J. C. Jacob, Stirling Carpenter and Frederick Andermann, McGill University, Montreal, Quebec and Memorial University, St. John's, Newfoundland

The non-gangliosidotic forms of cerebromacular degeneration (CMD) or Batten disease are rare autosomal recessive disorders characterized by the storage of autofluorescent material in the cytosomes of various cells. In the late infantile form, these appear as curvilinear profiles (CLBs) on electromicroscopy, whereas in the juvenile and adult forms, fingerprint profiles predominate. The chemical identity of the stored material or materials is unknown. We were unable to confirm the myeloperoxidase deficiency reported by Armstrong and others.

We have recently isolated by differential centrifugation, curvilinear profiles in a highly purified form as verified by EM, from the cerebral cortex of a 7-year-old Italian-Canadian girl who died with previously diagnosed late infantile CMD. Treatment of the CLB's with lipid-

extracting solvent removed 43% of the dry weight, leaving a water-soluble amorphous fluorescent residue which was subjected to various physico-chemical studies. The lipid-free autofluorescent component was identified as complexes of retinoic acid and related compounds with peptides. Similar complexes appear to be present in the urine of other patients with late infantile CMD.

We have now recognized 51 cases of CMD in 33 families: 30 late infantile, 17 juvenile, and 4 adolescent or adult. 31 cases (61%) were identified in Newfoundlanders, and 8 (16%) in Italian-Canadians. The incidence of CMD in Newfoundland approaches that of Tay Sachs disease in Ashkenazi Jews, indicating an urgent need for a heterozygote screening method.

Our findings on the nature of the stored material in late infantile CMD may lead us to a method of carrier detection and prenatal diagnosis, as well as a possible therapeutic approach in these disorders.

Hemi²: Hypertrophy, Hypaesthesia and Areflexia

Kenneth Nudleman, Frederick Andermann, Gilles Bertrand, Eugene Rogala and Eva Andermann, Montreal

Three girls presented with a developmental syndrome of hypertrophy involving half or a quadrant of the body and not involving the face. The appearance was one of inappropriately large size of the affected side rather than atrophy of the contralateral side, although this was initially questioned.

On the affected side, there was hypertrophy of muscle and increase in diameter but not in length of long bones. Power was increased. There was areflexia and decreased pain and temperature sensation in the affected territory. The patients also had scoliosis and some foot deformity on the large side.

One patient had a lumbar myelomeningocele and the hypertrophy involved only the upper quadrant. A second had a family history of spina bifida.

EMGs, conduction studies, EEGs, skull X-rays, PEGs and CAT scans were normal. The cord was not enlarged. Chromosome studies revealed normal karyotypes. Sex chromatin was female on both sides in one patient.

A defect in the dorsal lip of the neural tube or the neural crest is postulated to explain the abnormality. The association with a closure defect in one patient and family history of a similar defect in another suggest that the developmental abnormality occurs at an early embryonic stage.

Awareness of this syndrome may facilitate differentiation from hemiatrophy of the opposite side due to cerebral or other causes.

KUFS' DISEASE:

A Study of an Italian Family with Onset of Progressive Myoclonus Epilepsy at Age 30

Frederick Andermann, Stirling Carpenter, Pierre Gloor, Eva Andermann, Leonhard S. Wolfe, S. Lal and J. Clifford Richardson, Montreal and Toronto

The proband, a 33-year-old Italian woman, was normal until the onset of generalized seizures at the age of 30. Myoclonus developed the following year rapidly increasing in severity. The patient showed mild mental deterioration. When examined during diazepam infusion, which temporarily abolished the myoclonus, she had no cerebellar or other focal deficit.

The patient was extremely photosensitive. The EEG showed paroxysmal bilaterally synchronous irregular slow multiple spike and wave discharges. The EEG spikes were associated with myoclonic EMG spikes. Coincident with the slow wave of the EEG spike and wave discharge, there was flattening of the EMG, but only in muscles under tonic voluntary innervation. The slow wave components thus seemed to inhibit voluntary tonic motor outflow from the cortex only. Apomorphine, a dopamine agonist, arrested the spontaneous and light-evoked myoclonus for several minutes, but the paroxysmal EEG response to IPS was less affected.

Kufs' disease was diagnosed from brain biopsy which revealed autofluorescent insoluble material filling large neurons and their proximal axons. Electron microscopy showed fingerprinting profiles. Similar deposits were subsequently identified in eccrine sweat glands.

Four younger siblings and her four children are clinically normal. However, a 31-year-old sister showed photosensitive EEG abnormalities and had a generalized tonic-clonic seizure during IPS. An autosomal recessive pattern of inheritance is probable.

The diagnosis of Kufs' disease must be considered in adults developing progressive myoclonus epilepsy. However, the age of onset, clinical manifestations and ultrastructural features of Kufs' disease are variable, suggesting genetic heterogeneity.

Indications and Results of Stereotaxic Seizure-Monitoring in Patients with "Bi-Temporal Epilepsy"

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In our experience, the main indications for stereotaxic seizure-monitoring in patients with psycho-motor epilepsy have been the presence of *bitemporal interictal spike activity on scalp and sphenoidal recordings*, the occurrence of ictal discharges contralateral to the side of maximal interictal epileptiform activity and the occurrence of apparently simultaneous bitemporal ictal discharges.

The surgical technique consists in inserting stereotaxically and symmetrically in both temporal lobes three strands of stainless-steel electrodes, each containing usually nine (9) recording sites with a 5mm inter-electrode distance which permits recording of both the limbic and neocortical structures. The electrodes are chronically fixed to the skull with hollowed screws to prevent artefact recording and electrode movements. The actual recording of seizures is based on the computer push-button seizure-monitoring system developed by Ives, Thompson and Gloor.

The results in the first thirteen (13) consecutive patients presenting with an initially ambiguous bi-temporal pattern are presented. A total of 854 ictal seizure discharges were recorded in these 13 patients during a recording period lasting between two and three weeks. Of those, 485 had clear clinical manifestations; 369 were "electrographic" seizures without clear clinical signs. This represents an average per patient of 37.3 recorded clinical seizures and 28.4 "electrographic" seizures.

The main finding in this series is that these seizures appear to originate in one temporal lobe rather than in both, in an overwhelming proportion in each patient (87.4% lateralization to one side). In 5.3%, there was clear evidence for contralateral seizure onset as well. In the remaining 7.3%, the side of onset could not be determined.

Twelve out of thirteen of these patients have later been submitted to craniotomy and temporal lobectomy, and the early follow-up shows that they have a decrease in their seizure tendency comparable to the so-called "unilateral" temporal cases.

Les Tumeurs Hypophysaires par Voie Transphénoïdale

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Nous présentons notre expérience chirurgicale dans le traitement des tumeurs de la selle turcique, depuis l'année 1970, soit 39 patients avec lésion intrasellaire qui ont été opérés par voie transphénoïdale.

Nous faisons la classification selon le tableau clinique présenté par les malades, dans laquelle nous trouvons quatre entités cliniques:

1. Hypopituitarisme:
 - 24 cas
 - 20 patients avec adénome hypophysaire
 - 2 patients avec crâniopharyngiome
 - 1 patient avec méningiome
 - 1 patient, tumeur indéterminée
2. Acromégalie: 5 cas
3. Syndrome de Cushing: 3 cas
4. Microadénome à la Prolactine: 7 cas

Nous faisons une analyse clinique et radiologique de nos patients et aussi une discussion sur la technique chirurgicale et l'utilité de la radiothérapie en post-opératoire.

Le mortalité chirurgicale est nulle et dans la morbidité, on note à part un déficit hormonal, un diabète insipide chez trois (3) patients, des crises convulsives chez deux (2) patients et chez un patient une récurrence de la tumeur (méningiome).

Du point de vue visuel, il y a eu atteinte chez 32 patients avec amélioration post-opératoire pour 30 d'entre eux et aucun changement chez les deux autres. Il y a eu détérioration visuelle tardive chez deux autres patients qui ont montré une amélioration immédiatement après la chirurgie.

Brachial Plexopathy after Treatment of Cancer of Breast

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Wellesley Hospital, Toronto

The effect of radiation on the peripheral nervous system may be latent and progressive. The dilemma following mastectomy is whether the brachial neuropathy represents neoplastic disease or radiation plexopathy.

Fourteen patients developed variable symptoms referable to an upper limb. Screening for metastatic disease, serial neurologic, and electromyographic evaluations were reviewed. Absence of histologically identifiable neoplastic disease and/or lack of regression of brachial plexopathy to radiation or chemotherapy was used to identify post irradiation progressive plexopathy.

The majority of patients with metastatic disease had infiltrating duct carcinoma and developed symptoms within the first 3 years after mastectomy. Horner's syndrome, vocal cord or diaphragm paresis occurred only in the metastatic group. Pain when present was referable to the scapula or the neck. Patients with progressive nonmetastatic plexopathy were more likely to have had scirrhous carcinoma. Their symptoms developed after a long latent period of 9 years in 5/6 patients. A consistent, but rather atypical EMG pattern in this group is probably indicative of chronic denervation. Steroid therapy did not alter the progressive course.

Two distinct clinical syndromes emerge that may be useful in identifying the metastatic from nonmetastatic progressive brachial plexopathy. The radiation injury to peripheral nerves is probably compounded by entrapment and postsurgical instability of the shoulder.

Further Biomechanical and Cinematographical Analysis of the Role of Sensory Afferents in Motor Control in the Cat

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The functional role of somatic afferents travelling through the dorsal columns was studied in a serially organized motor act. Cats were trained to jump up in order to release a piece of liver attached to a vertically oriented revolving wheel. Although previous behavioral studies showed a significant impairment in the cat's ability to release the liver, the observational method used above could not provide any information concerning the mechanisms underlying the deficit. We therefore studied the mechanical effort of the animal while on the ground (force transducer platform) and complemented this by high speed cinematography (64 frames/sec.) in order to get insight into these mechanisms. After dorsal column section the following parameters, directly related to the developed force, were significantly decreased: 1 — height of jump, 2 — total maximum force, 3 — power, 4 — peak force. Film analysis showed that intact cats consistently extend their limbs in a smooth and progressive way towards the liver in order to release it. The distance between the tips of the forelimbs remained more or less constant during the flight. Postoperatively, instead of the regular pattern described, the extension of the limbs was interrupted by fast flexion movements and the distance between their tips increased significantly during the flight. We think the impairment observed in the animal's ability to develop peak force after dorsal column section may relate in part to the suppression of low threshold cutaneous signals from the hindlimbs travelling through fibers in the dorsal column involved in supra-segmental loops. The impairment observed in the forelimb extension may be associated with the fact that the dorsal columns are the exclusive afferent path to brain centers for muscle spindles and for low threshold joint afferents. Signals from these proprioceptors may be of importance in supra-segmental loops involved in the control of the precision and accuracy of forelimbs.

Effect of Dorsal Neck Muscle Afferents on Structures Involved in Oculomotor Control

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It was previously reported that both dorsal neck and extraocular muscle afferents project and converge at the single cell level onto neurons in the frontal cortex in regions corresponding with the frontal eye fields in the cat (Exp. Neurol., 1977, In Press). Present analysis shows that in addition to these excitatory inputs to frontal brain regions, stimulation of extraocular muscle nerves also inhibited neuronal activity as was shown against a background of excitation created through the iontophoretic release of glutamate. Whereas units responded to stimulation of the nerve of the superior rectus contralateral to the recording site, the same units were inhibited with stimulation of the superior rectus nerve ipsilateral to the recording site. The inverse relationship also held. The latencies of excitation by one input and those of inhibition by the other overlapped, and were 15-20 msec. Available data indicate that these excitatory and inhibitory interactions within extraocular muscle inputs in frontal brain areas parallel the agonist-antagonist interactions known to exist between pairs of extraocular muscles during eye movements.

In order to test whether dorsal neck muscles have an effect on extraocular muscles at the peripheral level, experiments were conducted in which the nerves of dorsal neck muscles were stimulated while evoked activity was recorded directly from extraocular muscle nerves. Results indicate that evoked activity was elicited in the nerves of the superior rectus and the lateral rectus muscles of the eye by electrical stimulation of dorsal neck muscle afferents.

Dietary Exclusions in the Therapy of Vascular Headaches

William Pryse-Phillips, St. John's, Newfoundland

The traditional therapy of vascular headaches has stressed that following careful history, examination and, where indicated, investigation, various schemes of drug therapy be employed.

Because of conflicting evidence as to the efficacy even of Ergot and because of the unwanted effects so commonly produced by the agents of general use, a scheme of management was devised whereby the role of a diet and of exogenous chemical substances could be evaluated.

Patients clinically diagnosed as having vascular headaches of migraine type (any variety) were instructed to keep a diary of all substances consumed over a six week period. Their headache frequency was assessed during this time. The possible association between certain foods and the occurrence of headaches was noted in each case and the patient instructed to abstain completely from foods which contained monosodium glutamate, nitrates or high concentrations of tyramine, betaphenylethylamine and certain other vasoactive amines. They were reassessed following this at regular intervals.

Results are reported from the first 200 patients managed in this way. Over 60% of patients claimed such a relief from headaches that further medical attention was not, they claimed, necessary any more.

A higher proportion of patients were aware of dietary precipitants to migraine headaches although they found that exclusion of the offending substances did not relieve all the headaches. In less than 25% of cases however, no dietary precipitants could be identified.

The relevance of these findings to certain models of production of migraine headaches now under survey will be discussed.

Transplantation of Human Schwann Cells from Metachromatic Leukodystrophy and Control Nerves to Immune-Suppressed Mice

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Schwann cells can be transplanted into nerves of animals of the same or different strains (J. Neurocytol. 5: 565, 1976). Combinations of regenerated axons and transplanted Schwann cells from normal and abnormal nerves have made possible the demonstration of a primary Schwann cell deficit in Trembler (Nature 265: 73, 1977) and Quaking mouse neuropathy. The present communication reports the successful transplantation to immunologically deprived mice of human Schwann cells and fibroblasts from a patient with metachromatic leukodystrophy (MLD) and from control nerves.

Control human sural nerve fascicles, 5 mm in length, were obtained from amputations and routine biopsies and grafted between the stumps of transected sciatic nerves in 26 mice. Nerve segments from a biopsy of a 10-year-old patient with proven MLD (material was kindly provided by Dr. P. Humphreys, Montreal Children's Hospital) were similarly grafted into 12 mice. To prevent rejection all host animals received antilymphocytic serum. Serial phase and electron microscopic examinations of grafted nerves 10 days to 3 months after grafting showed: (1) no rejection of the human grafts in the ALS-treated mice; (2) increasing numbers of myelinated and unmyelinated fibers within the graft and distal stumps of grafted nerves; (3) a connective tissue arrangement within the graft that resembled that in the donor human nerve. From 2 to 3 months after transplantation of MLD nerve segments into mice there were metachromatic granules in Schwann cells of regenerated fibers within the graft. To confirm the presence of human cells within grafts of regenerated mouse nerves, the immunosuppression was discontinued in a group of animals 1 to 2 months after grafting; under these conditions fibroblasts and Schwann cells in the graft were rejected but cells within proximal and distal stumps were tolerated by the host.

These experiments suggest that (1) regenerated mouse fibers can be myelinated by human Schwann cells; (2) abnormalities such as in human MLD nerves can be reproduced by human Schwann cells in the grafted animal; (3) the present technique may be helpful for the investigation of biologic mechanisms in normal and pathologic nerves of man as well as for experimental animals.

Post-Anaesthetic Augmentation of Muscle Damage as a Presenting Sign in Three Patients with Duchenne Muscular Dystrophy (DMD)

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Three children, none of whom was recognized to have Duchenne Muscular Dystrophy underwent anaesthesia with halothane and succinyl choline for surgery not involving trauma to muscle. In two patients the procedure was adenoidectomy done because the prominent tongue suggested adenoid obstruction. The third child developed fever with the induction of anaesthesia and his strabismus surgery was not performed.

All three children immediately post anaesthesia passed red-brown urine which was positive to orthotolidine but contained no red blood cells, findings compatible with myoglobinuria. The urine cleared in 24 hours in all.

Serum creatine phosphatase values immediately after anaesthesia were markedly elevated and then gradually fell over several days to the range usually seen in Duchenne Dystrophy. Duchenne Dystrophy was confirmed in all three by clinical examination, family studies, EMG and muscle biopsy. At least one of the children was weaker for a period after the anaesthesia.

Halothane and succinyl choline may cause muscle damage even in normal individuals. Children with DMD until now have not been considered especially vulnerable. Our experience suggests that they are more vulnerable than others and this should be taken into account when they require anaesthesia. We have documented the absence of such changes in patients with DMD receiving ketamine anaesthesia.

The enhancement of muscle breakdown in patients with DMD after anaesthesia with halothane and succinyl choline raises issues regarding the nature of this reaction and its relationship to the pathophysiology of Duchenne Dystrophy.

Benign Paroxysmal Vertigo of Childhood and Its Relationship to Basilar Artery Migraine

Gordon Watters, The Montreal Children's Hospital

Benign paroxysmal vertigo of childhood (BPVC) described originally in children less than 4 years of age by Basser (1964) has as its cardinal symptom brief attacks of vertigo lasting less than one minute. Disordered vestibular functioning documented by caloric testing was the only abnormal finding. No associated headaches, EEG abnormality or family history of migraine was noted.

In basilar artery migraine (BAM) there may be brief or sustained vertigo and ataxia. We have seen 22 children who have had attacks of BPVC which evolved into BAM. Family histories in all cases were positive for migraine while vestibular testing was abnormal in some (5 of 8 tested) and EEGs were abnormal in 10 of 22. Other studies were negative.

The evolution of BPVC into BAM suggests that local vasoconstriction may be the mechanism producing the attacks of BPVC. The deficits in vestibular function may be analogous to deficits accepted within classical and basilar artery migraine.

BPVC is a relatively common disorder in child neurology. Recognition of its association with migraine may be important in understanding the pathophysiology of the disorder and is helpful in reassuring migraineous parents that their child has a good prognosis.

Ten Years Experience with Various Forms of Myelotomy

L. P. Ivan

During the past ten years, from the author's own material, twenty-six patients had surgery for the relief of spasticity. Ten patients were treated with bilateral longitudinal myelotomy, ten patients with unilateral myelotomy, one patient with T-griseotomy and five patients with RF-griseotomy. All patients had good to excellent immediate relief of spasticity with all procedures. Recurrence of spasticity occurred in 20% of the cases and most frequently among those who had unilateral myelotomy. RF-griseotomy, first reported by the author in 1972 gave satisfactory results but the sample is too small for final conclusions. One patient (the only mortality in the group) who died two days after RF-griseotomy due to pulmonary embolism had histological examination of the cord. From this unique material, useful information is available to explain the possible mechanism of myelotomy. It appears that not only the disruption of monosynaptic and polysynaptic pathways is important but the loss of neurons in lamina 6 and 7 of Rexed is also responsible for the physiopathological mechanism that decreases the spastic state.

Adult Onset Nemaline Myopathy

A. K. W. Brownell (Calgary) and J. J. Gilbert (London)

Adult onset nemaline myopathy (AONM) is a rare disorder. The clinical picture is one of subacute onset of proximal muscle wasting and weakness in middle age. Creatine kinase (CK) levels are normal and the electromyogram (EMG) suggests a "neurogenic" disorder. In contrast to the congenital variety of nemaline myopathy where the prognosis is good, the adult variety is invariably associated with a bad prognosis. We have recently examined such a case.

A 47-year-old Chinese man was severely disabled because of long standing asthma and emphysema. Neurologic examination by one of us (AKWB) eight weeks prior to the patient's death disclosed a generalized decrease in muscle bulk that was most pronounced in the anterior compartment muscles of the legs. Strength was mildly decreased in all muscle groups except for the anterior compartment muscles which were moderately weak. Muscle stretch reflexes were brisk. CK levels were elevated five times above normal and EMG studies were suggestive of a "neurogenic" disorder. A muscle biopsy taken from the left vastus lateralis disclosed the typical features of nemaline myopathy by light and electronmicroscopy.

The patient died as a result of respiratory failure. At autopsy nemaline rods were identified in all muscles sampled including the diaphragm. The brain, spinal cord, ventral roots and proximal peripheral nerves were normal.

This case of AONM is the first reported with elevated CK levels. This abnormality plus the findings of normal anterior horn cells and proximal peripheral nerves at autopsy supports the concept that the disease is "myopathic."

Methohexitone Activation of Sphenoidal EEG

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Sphenoidal EEG recordings in 98 consecutive patients (age 28.6 ± 9.97 , range 11-55, M. 55, F. 43) were obtained over a two year period.

Teflon coated, flexible, stainless steel wires (35 AWG, .0055 in., 1.4 metric) were inserted by a Townsend introducer (S.L.E.) without local anaesthesia; the wires were stripped of insulation for 2 mm. at the tip. The average depth of insertion was 4 cm. No complications were encountered other than some soreness of the jaw and occasional surface haematomas. A 1% solution of Methohexitone was given intravenously to activate the recordings and an average of 4 cc (40 mg) was given. Artefacts were reduced by coiling the sphenoidal wires on Blenderm (3M) tape and the crocodile clips were taped to the cheek.

The main source of patients was by referral from psychiatrists, neurologists and internists. There was no significant difference in the age of patients referred over two years (27.36 ± 8 , 29.7 ± 11.58) and the mean age of the patients is similar to the mean age of onset of temporal lobe epilepsy (28, Currie et al, 1971). The average age of patients with abnormalities (30 ± 12) did not differ from the mean age of the total patients.

Twenty patients showed abnormalities in the activated sphenoidal recording; in ten patients an abnormality was present only in the sphenoidal record; in six patients the abnormalities were seen only during methohexitone activation.

Abnormalities were noted in three (temporal focus, 1; temporal sharp waves, 2) of nine patients who were referred because of aggressive outbursts.

We have found that such activated sphenoidal recordings are a safe, simple and sensitive investigation in patients who are thought to suffer from temporal lobe symptoms.

Existence Possible d'un Groupe de Neurones Serotoninergiques dans l'Hypothalamus du Rat

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L'examen radioautographique de l'hypothalamus de rats adultes, prétraités par un inhibiteur de la monoamine oxydase et soumis à une perfusion intraventriculaire prolongée de sérotonine tritiée (5-HT-³H) 10^{-4} ou 10^{-5} M, révèle la présence de corps cellulaires neuronaux ayant fortement accumulé le traceur, dans la pars ventralis du noyau dorso-médian. Aucune réaction de ce genre ne'est détectée au même endroit après administration de noradrénaline tritiée dans des conditions analogues. Par contre, le marquage est inchangé après administration concomitante de 5-HT-³H et de noradrénaline non radioactive en concentration 10 fois plus élevée. Les neurones réactifs forment un groupe lâche dans la pars ventralis du noyau dorso-médian. Leur nombre total peut être chiffré à 1,000 de chaque côté du troisième ventricule. En microscopie électronique, on constate que les corps cellulaires marqués sont grossièrement sphériques (diamètre moyen: 10 μ m) et que 30% de leur volume est occupé par un noyau profondément indenté. La structure fine de ces corps cellulaires n'apparaît pas différente de celle des autres neurones de la région. Cependant, tandis que des terminaisons nerveuses chargées de 5-HT-³H sont trouvées en contact synaptique avec les péricarions des neurones non marqués, une telle situation n'a jamais été observée dans le cas des neurones marqués. On en conclut à l'existence, dans le noyau dorso-médian de l'hypothalamus, d'une population de neurones possédant une affinité sélective pour la sérotonine exogène et qui pourraient contenir cette amine à l'état endogène. Il est possible que ces neurones contribuent significativement à l'innervation sérotoninergique de divers noyaux de l'hypothalamus et/ou de l'éminence médiane.

Chronic Relapsing Polyneuropathy

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20 patients with a non familial chronic progressive and often relapsing polyneuropathy are reviewed. This polyneuropathy is characterized by diffuse motor weakness, elevated CSF protein, slowing of nerve conduction velocity and pathologically by inflammatory cells with adjacent demyelination in peripheral nerves and roots. 14 patients have had a relapsing course — 8 relapses were spontaneous, 7 after corticosteroid therapy and 9 when steroids were decreased.

There was a marked male predominance with the age of onset greatest in the third and sixth decades. Weakness was diffuse, symmet-

rical and moderate to severe in the limbs. 2 patients had facial weakness; 15 had areflexia; and 5 had an action tremor. Vibration sense loss was present in 14 and position impairment in 8.

CSF protein was elevated in 14 of 19 patients. EMG showed a loss of motor unit potentials and potential amplitudes. Denervation potentials were recorded in 5 patients. Motor nerve conduction velocities were diffusely decreased, but variable with mean velocities being 50% of normal. 8 patients had sural nerve biopsies that showed variable losses of large myelinated fibres, segmental demyelination and remyelination, and axonal degeneration.

The average duration of the disorder in the 20 patients is 3.9 years (7 months to 13 years). 18 patients received corticosteroids with improvement in 15; 9 remain on maintenance steroids. Presently, 11 patients are normal, 5 have mild residual findings while 4 have moderate to severe persistent defects of motor and sensory function. This neuropathy shares common features with the acute polyradiculoneuropathy of the Guillain-Barre type and immunological mechanisms are presumably related to its pathogenesis.

Extracranial-Intracranial Arterial Bypass in the Treatment of "Giant" Intracranial Aneurysms

S. J. Peerless, C. G. Drake and G. G. Ferguson

Extracranial-intracranial (EC-IC) bypass has been used as an adjunct in the treatment of seven "giant" aneurysms which have proven to be inoperable by direct surgical ablation. The purpose of the bypass was to provide collateral flow, allowing subsequent occlusion of the immediate feeding artery or arteries.

Three patients presented with dominant middle cerebral (MCA) aneurysms. Following a single superficial temporal to cortical artery anastomosis, one has tolerated almost complete occlusion of the middle cerebral artery and another complete occlusion of the mainstem of the MCA with a double anastomosis using both the superficial temporal and occipital arteries. The third patient has tolerated complete occlusion without deficit.

Four patients with giant carotid bifurcation aneurysms have had EC-IC bypass procedures. One patient of these four, developed severe but transient ischemia on three attempts at proximal occlusion prior to the bypass. Following bypass, occlusion of the intracranial carotid and A-1 segment has been tolerated well. A second patient with a giant carotid bifurcation aneurysm underwent immediate proximal occlusion which was well-tolerated. One case died of a recurrent hemorrhage of the aneurysm before the proximal vessel could be occluded and a second had a major ischemic complication following embolization from a giant carotid ophthalmic aneurysm. These cases indicate that the extracranial-intracranial bypass procedure is capable of supporting the entire flow to the middle cerebral territory and that rapid enlargement of the shunt occurs in the circumstance of a large pressure drop across the anastomosis.

Prospective Study of Electroencephalograms in Children of Low Birth Weight

H. G. Dunn, N. L. Auckland, M. D. Low, J. U. Crichton, A. M. Robertson, E. N. Tredger and A. Karaa McBurney

In a prospective study of 501 children of low birth weight (LBW) who mostly weighed 2041 gm (4½ lb) or less, 277 had electroencephalograms (EEGs) recorded after incubator treatment at a mean conceptual age of 39 weeks ("late neonatal" EEGs), 249 had EEGs at a chronological age of about 1 year, and 259 at about 6¾ years. 203 control children of full birth weight (FBW) provided 100 EEGs after 1 year and 117 after 6¾ years. In the LBW children 26% of neonatal records, 18% at 1 year and 46% at 6¾ years were considered borderline or abnormal. Among FBW controls, only 2% of EEGs at 1 year were borderline (none definitely abnormal) and 28% at 6¾ years were borderline or abnormal. Paroxysmal discharges (spikes or spike-waves) occurred in 42% of abnormal records of LBW and in only 21% of abnormal records of FBW children at 6¾ years. Most abnormalities of the "late neonatal" EEG in LBW infants resolved, and overall normality of this EEG was not significantly related to neurological status of WISC Full-Scale I Q. at 6¾ years. In contrast, definite abnormalities of the EEG at 1 year were nearly always associated with subsequent neurological abnormalities.

and the result of that EEG was significantly associated with subsequent WISC I.Q. score ($p < 0.05$). "True prematures" had a slightly higher proportion of normal records than "small for dates" children, but the difference was not significant.

Mesure du Flux Sanguin Cérébral par Injection de Xenon-133: Le Temps d'Augmentation

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Les études du flux sanguin cérébral (FSC) à l'aide de traceurs diffusibles sont pour la plupart basées sur l'analyse du temps de disparition (washout) tandis que les études à l'aide de traceurs intravasculaires sont fondées sur la détermination du temps d'augmentation (TA) d'un niveau de base à un niveau maximum de radioactivité cérébrale. Cependant, cette dernière méthode d'analyse s'avère aussi utile lorsqu'on utilise un traceur diffusible comme le xénon.

Chez 31 chiens qui ont reçu des injections rapides d'une solution de xénon-133 données sous pression et par voie intraveineuse, le TA mesuré sur le crâne mis à nu était à peu près deux fois plus long que la période d'infusion radioactive au cerveau telle que mesurée au niveau de l'artère carotide par un détecteur. Puisque le TA est directement proportionnel à la période d'infusion radioactive au cerveau, le TA peut servir comme index d'irrigation cérébrale artérielle. TA prolongé = irrigation indirecte, TA court = irrigation directe. Par contre, le temps de disparition est lié au métabolisme.

Une patiente illustre très bien de concept. Une jeune fille qui avait un vieux ramollissement cérébral gauche et une sténose de la carotide droite à la suite d'un accident s'est présentée avec une maladresse épisodique de la main gauche. L'étude FSC intraveineuse a démontré un TA prolongé de côté droit avec disparition normale du traceur. Cela indique une irrigation indirecte et un métabolisme normal de l'hémisphère droit. Quant à l'hémisphère gauche, le TA était court mais la disparition était prolongée. On a conclu à un infarctus cérébral avec métabolisme ralenti mais irrigation directe. L'artériographie a confirmé cette conclusion.

Le TA nous renseigne sur l'irrigation artérielle alors que la disparition est liée au métabolisme. Dans les cas où un pontage (by-pass) est considéré ces deux index peuvent servir à identifier les patients qui sont les meilleurs candidats pour une chirurgie de revascularisation, à savoir ceux qui ont un métabolisme quasi-normal malgré une irrigation insuffisante.

Intradural Spinal Arachnoidal Cysts: A Cause of Obscure Pain Syndromes

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Congenital cysts or diverticulae of the arachnoid overlying the posterior aspect of the thoracic spinal cord are often asymptomatic and may appear in incidental findings on myelography. Rarely, they may cause neurologic deficits as a result of compression or traction on the spinal cord or nerve roots. They may also present with pain as the only symptom, either non-specific back pain or radicular pain referred into the chest or abdomen.

Because of the referred nature of the pain initial diagnostic procedures may be directed towards structures outside the nervous system. Even when a spinal lesion is suspected, the diagnosis may be missed as regular prone myelography often fails to demonstrate the posteriorly located cysts and supine myelography is required. The diagnostic challenge presented by these lesions is illustrated by two cases, both of whom underwent extensive investigation for cardiac or abdominal disease before the true origin of their symptoms was appreciated.

A 20-year-old woman presented with a three-year history of intermittent sharp pain in the lower posterior thoracic region radiating into the right upper quadrant of the abdomen. Radiologic investigations of the stomach, bowel, gall bladder, and kidneys failed to reveal a cause of the pain. Neurologic examination was normal except for a poorly defined band of hyperesthesia involving dermatomes T7-T10 bilaterally. A myelogram carried out in the prone position was negative, but

myelography in the supine position revealed four large cystic out-pouchings of the arachnoid over the lower thoracic region. Laminectomy confirmed the presence of typical thin walled, posteriorly located arachnoidal cysts.

A 23-year-old man was investigated for persistent epigastric pain which occasionally radiated around the left costal margin into the back. Neurologic examination was entirely normal. Because of an unusual contour of the heart on chest x-ray and an abnormal ECG, he underwent detailed cardiac investigations including cardiac catheterization without a cause for the pain being demonstrated. He subsequently developed weakness in his legs and numbness over the lower extremities and trunk up to the T7 level. A myelogram showed a complete block at the T6-7 level, and subsequent laminectomy revealed a large arachnoidal cyst compressing the cord at this level.

Cerebral and Retinal Vascular Complications of Inflammatory Bowel Disease

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Cerebrovascular involvement in the course of ulcerative colitis and Crohn's disease is rarely recognized. We document five such cases: recurrent retinal artery branch occlusion, choroidal infarction, carotid thromboembolism, brainstem ischemia, and hemorrhagic infarction from presumed cerebral venous thrombosis. Of these five patients, age 34 to 57, three had Crohn's disease and two had ulcerative colitis.

The usual risk factors for cerebrovascular disease were absent as determined by cerebral and retinal angiography, cardiologic investigation, glucose tolerance tests and serum lipid analysis. There was no evidence of vasculitis. Coagulation studies revealed thrombocytosis, short partial thromboplastin times and elevated fibrinogen and factor VIII. The abnormal coagulation state that accompanies inflammatory bowel disease (Lam, A. et al. *Gastroenterology* 68: 245, 1975) may lead to arterial and venous thrombosis. Our experience indicates that inflammatory bowel disease can predispose to stroke.

Patterned Visual Evoked Responses: A Sensitive Detector of Occult Optic Neuritis

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We present a large-series confirmation of Halliday's discovery that latency of pattern VER is a sensitive indicator of an acute or chronic lesion in the anterior visual system, even when vision is symptomatically normal. This test thus can detect occult optic neuritis in patients suspected of having multiple sclerosis (MS).

The image of a checkboard pattern is abruptly shifted horizontally on a screen 50 cm. in front of the patient. Occipitally evoked potentials are averaged for 128 positional shifts. The evoked potential latency to the first major positive peak ranged from 70 to 95 msec. in normals and 93 to 95 msec. in neurological patients without anterior visual system lesions and who were not MS suspects.

Significant unocular or binocular delays occurred in all 20 MS patients with known optic lesions and in 10 of 14 MS patients without apparent ocular abnormalities. Delays occurred in 4 of 14 patients with chronic myelopathy in whom MS is suspected. In other patients, suspected of having early MS, delays occurred in 1 of 6.

This innocuous, technically reliable test, provides valuable diagnostic data in patients whose neurological abnormalities suggest MS. However, other anterior visual system lesions could also create abnormal responses.

Further Motor Unit Studies in Duchenne Muscular Dystrophy

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Following the publication of motor unit counting studies in 19 patients with Duchenne muscular dystrophy in Newcastle, England (McComas, A. J. et al., *J. Neurol. Neurosurg. Psychiatr.* 34, 461-468, 1971), it has been possible to examine 26 patients in the Hamilton area. This new study has extended the previous one by enabling certain additional observations to be made. In 24 patients (92%) a loss of

functioning motor units could be demonstrated in at least one of the following muscles — thenar group, extensor digitorum brevis (EDB) and soleus; the hypothenar muscles differed in being less frequently and less severely involved. Within the population of patients there was no correlation between the age of the patient and the number of functioning motor units in the thenar and EDB muscles. A similar conclusion was reached in a serial study of 4 patients, now aged 12-13 years, who had been investigated over a four-year period. In the latter group, the behaviour of the soleus muscles stood in marked contrast; not only were there significant losses of units but the maximum evoked muscle responses (M-waves) declined to approximately one-third of their previous sizes. An unexpected finding was that the EDB of one of the four patients showed substantial, though temporary, increases in the M-wave and in the number of functioning units. The interpretation of these additional findings is that (a) there is a definite neural abnormality in Duchenne muscular dystrophy, (b) in the intrinsic muscles the abnormality may be expressed in early childhood and show little change thereafter, (c) in a larger muscle, the soleus, there is a striking deterioration in neuromuscular function at about 10 years, and (d) the dystrophic process may occasionally undergo a remission, possibly through temporary restoration of function in 'sick' motoneurons.

Définition des Sillons Corticaux Chez l'Homme par l'Angiographie Cérébrale

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La définition angiographique d'un sillon correspond à l'ensemble des segments artériels et veineux profonds qu'il contient et qui moulent ses parois chacun à son niveau. La lame vasculaire profonde constituée par tous les segments profonds d'un sillon est le fait anatomique de base qui explique l'angiogramme cortical. La profondeur, l'orientation et la forme des surface invaginées sont inscrites dans les images radiographiques des trajets vasculaires. Afin de faciliter leur reconnaissance sur les radiographies, la plupart des photographies et dessins anatomiques qui sont présentés sont pris dans les mêmes projections. En réunissant les trois projections latérales, antéro-postérieures et axiales d'un sillon et de ses vaisseaux, nous pourrions progressivement mieux concevoir la réalité tri-dimensionnelle des lames vasculaires des sillons qui doivent remplacer dans notre esprit les lignes figurant les sillons sur les schémas d'édification habituels. Sur l'angiographie corticale, le critère fondamental est la profondeur d'un segment ou au contraire sa position superficielle. De ce point de vue, ni le mode de division des troncs, ni leur calibre ne joue aucun rôle: plusieurs petites branches profondes peuvent définir un sillon tout comme une artère plus volumineuse; celle-ci peut cheminer à la surface d'une circonvolution et ce ne seront que ses branches qui en plongeant dans les sillons voisins en définissent l'emplacement.

C'est pour cette raison qu'il faut abandonner certains noms comme artère "central", "pré-central", "angulaire", puisque ces termes sont "misleading" car ils suggèrent une relation spécifique d'une artère à un sillon ou à une circonvolution, ce qui ne colle pas avec la réalité anatomique.

A l'aide de ce nouveau concept donnant une vue tri-dimensionnelle, nous présenterons l'anatomie vasculaire des principaux sillons du cerveau, principalement ceux de la région centrale et de la région sylvienne.

Cette nouvelle approche que nous proposons pour étudier l'angiographie cérébrale revalorise à nos yeux la valeur de l'angiographie cérébrale et en fait un instrument diagnostique tout aussi populaire et tout aussi valable que peut l'être la tomographie axiale computerisée.

Essential Tremor and Regional Dystonia

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Essential tremor is considered by most neurologists to be a monosymptomatic disorder which reflects a minor disturbance of central motor control mechanisms. However for over one hundred years reports have periodically appeared suggesting that other abnormal motor phenomena such as torticollis, other dystonic phenomena,

choreiform limb or facial movements and nystagmus may appear in association with essential tremor.

A retrospective study of 100 patients who consulted their physicians because of essential tremor was made searching for evidence of associated motor symptoms or signs. Evidence of an associated regional dystonia was found in 9 patients. The dystonic symptoms involved the neck alone in 4 patients, the neck and upper extremity in 3 patients, and 1 upper limb alone in 2 patients. One further patient who presented because of essential head and hand tremor has a sister with gross torticollis. In all patients tremor preceded the appearance of dystonic symptoms by at least eight years. The severity of the dystonic symptoms varied considerably.

Almost all elderly patients with essential tremor felt that their symptoms had increased over the years and several had developed minor extrapyramidal features other than dystonia. None had developed gross Parkinsonian features or cerebellar ataxias.

This relatively high incidence of dystonic phenomena in patients with essential tremor has not been reported previously. Several investigators however have reported a high personal and family history of tremor in patients who present with spasmodic torticollis.

This retrospective study suggests the need for careful documentation and prospective study of patients presenting with essential tremor, spasmodic torticollis, writer's cramp or other dystonic syndromes. A meaningful relationship between essential tremor and these dystonic syndromes could have important implications regarding the site of pathophysiology of these disorders.

Transplantation of Dystrophic Spinal Roots into Sciatic Nerves

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In spinal roots of dystrophic mice, neonatal Schwann cell multiplication is impaired (Bray et al., *J. Neurol. Sci.*, in press) and many axons lack Schwann cell sheaths (Bradley and Jenkinson, *J. Neurol. Sci.* 18: 227, 1973). Because the expression of certain Schwann cell disorders can be reproduced after transplantation (Aguayo et al., *Nature* 265: 73, 1977), L3 or L4 spinal roots from dystrophic (C57 BL/6Jdy^{2J}/dy^{2J}) and control mice were grafted into sciatic nerves in the following combinations: normal → normal; dystrophic → normal; normal → dystrophic; dystrophic → dystrophic. To prevent rejection all animals received antilymphocytic serum twice weekly. After 5 weeks, the mid-graft segments and stumps were examined by electron microscopy.

In all combinations, axons regenerated from the proximal sciatic nerve stumps, through the grafted roots and into the distal sciatic segments. After regeneration the grafted segments contained many myelinated fibers of various size and there were no naked axons. Myelin lamellae/axon circumference ratios were similar for all four groups of animals.

Thus, the abnormality which characterizes dystrophic spinal roots cannot be reproduced by various combinations of axons and Schwann cells from dystrophic and normal animals. This suggests that the focal deficit of Schwann cell ensheathment in dystrophic mice is secondary to an abnormality of axonal surface membranes at the spinal root level.

Regional Cerebral Blood Flow Determinations by Common Carotid Artery Injection of Xenon 133

R. D. Rudelli, V. C. Hachinski, J. W. Norris, P. W. Cooper

Estimation of regional cerebral blood flow (rCBF) by the intracarotid Xenon 133 method is at times precluded by stenosis of the internal carotid artery (ICA). Thus rCBF measurements cannot be done in the very patients with cerebrovascular disease in whom such studies would be most helpful.

We carried out rCBF studies by common carotid artery (CCA) Xenon 133 injections in 12 patients. We also determined rCBF by the intracarotid method and blood flow in the external carotid artery (ECA) in 5 of these cases. rCBF by CCA injection was on the average 7% lower than by ICA injection, when changing PaCO₂ was taken into account. The reactivity of the rCBF to changing PaCO₂ (hyperventilation) was 6% Δ in CBF/mm of PaCO₂, which is comparable to the ICA method.

Focal rCBF changes due to hemispheric lesions were clearly demonstrable by the CCA injection method

Despite some loss in accuracy compared to the intracarotid technique, CCA injection of Xenon 133 is a reliable method of determining rCBF, even in the presence of stenotic lesion of the ICA.

Effects of Hypercapnia on Visual Evoked Responses in Acute Cerebral Infarction

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Inhaled CO₂, a potent cerebral vasodilator, has been utilized for treatment of cerebral ischemia. Studies in experimental animals suggest that vasodilatation may actually reduce blood flow to ischemic cerebral tissues. In a study using baboons, hypercapnia frequently reduced or abolished visual evoked responses (VERs) in the ischemic hemisphere. To determine the effect of hypercapnia on VERs in humans, we have compared occipital VERs at baseline and during hypercapnia in 20 patients with acute unilateral cerebral infarction (10 with and 10 without homonymous hemianopsia) and in 10 age-matched normal controls. VERs were judged on the basis of interhemispheric symmetry and were considered abnormal by the following criteria: 1) peak latency differences exceeding 5 msec, 2) amplitude differences of more than 50% of the smaller value; and 3) unilateral lack of one or more VER deflections. In controls, eight of 10 had normal baseline VERs which did not change during hypercapnia. Two controls had unilateral lack of a VER deflection; one became symmetrical during hypercapnia and one remained asymmetrical. In patients with cerebral infarction, one of 10 with hemianopsia and six of 10 without hemianopsia had normal baseline VERs; all except one remained normal during hypercapnia. In thirteen patients with abnormal baseline VERs, hypercapnia caused improvement of VERs in four, deterioration in three, and no change in six. We conclude that hypercapnia may have variable effects of cerebral function as measured by VERs in patients with cerebral infarction; these effects may be deleterious in some instances and beneficial in others.

Hydrocephalus: Compensated or Arrested?

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The advent of computerized axial tomography has made detection of changes in ventricular size very easy. We have been surprised on several occasions when C.A.T. scans have been done on children who were said to have "arrested" hydrocephalus. This has prompted us to reassess our definition of "arrested hydrocephalus."

Schick and Matson, in 1961 defined arrested hydrocephalus as cessation of head growth until the growth of the child caught up to the size of the head. Others have used the term arrested hydrocephalus without defining it in terms of head or ventricular size.

In our experience, arrested hydrocephalus as defined by Schick and Matson is very rare. Many of our hydrocephalic infants who have had a shunt inserted have had a return to normal head size. Post-operative C.A.T. scans on these infants have revealed a return of ventricular size to normal, and thus this group might be said to be "shunt arrested."

Another group of children have been seen who have large heads, but no definite symptoms or signs of raised intracranial pressure. C.A.T. scans on these children have shown mild to moderate ventricular dilatation. We have done further studies on these children, and feel that their hydrocephalus is compensated, but not arrested.

Several other children have been seen, with symptoms of occasional headache and mild memory difficulty. These children have shown progressive head enlargement, and massive ventricular enlargement has been documented on C.A.T. scans. Intraventricular pressure monitoring has been done in one case, and periods of raised intracranial pressure have been documented. This child was thought to have had "arrested hydrocephalus" because of the absence of symptoms of raised intracranial pressure, and the appearance of the skull roentgenograms.

Details of 4 cases, which illustrate these various points, will be presented.

We conclude that spontaneous arrested hydrocephalus is very rare, shunt arrested hydrocephalus occurs frequently, and a significant number of children with hydrocephalus become compensated, but need regular follow-up assessments to be sure that they are not slowly becoming active, or "decompensated."

Does Pargyline Alter Fast Axoplasmic Transport?

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The monoamine oxidase inhibitor pargyline will induce a myopathy in rats which can be prevented by prior sciatic nerve section. We reported that pargyline will increase the rate of fast axoplasmic transport of ^3H -labelled material in the sciatic nerve of these animals. Bisby however, claimed that pargyline did not alter axonal transport. We therefore examined the effect of pargyline under carefully controlled conditions and found an increase in the axonal transport rate. Rats received pargyline (75 mg/kg ip) daily for 7 days. The left and right sciatic nerves were ligated 3 cm distal to the L-4 dorsal root ganglion. The spinal roots on one side were sectioned 1 cm distal to the sciatic nerve cell bodies while those on the other side remained intact. $75 \mu\text{Ci}$ of ^3H -leucine was injected into the region of the sciatic nerve cell bodies on both sides of the spinal cord. Animals were maintained at 37°C and sacrificed between 1.5 and 5 hrs. after labelling with isotope. The accumulation of isotope in 1 cm nerve segments proximal and distal to the ligature was measured. The nerve with severed spinal roots served as a control for blood borne radioactivity. We found a time dependent accumulation of isotope at the ligature of the intact nerve. In pargyline-treated animals, a fast transport rate of 1100 mm/24 hrs of ^3H -label was obtained while in control non-drug treated animals the rate was 408 mm/24 hrs.

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Iatrogenic Femoral Nerve Injury

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(a) Hip Surgery

Femoral nerve injury complicates hip surgery less frequently than does sciatic nerve palsy. The resultant quadriceps weakness, however, is a serious disability in a patient with an abnormal hip joint. The case material presented here includes two patients in whom the nerve was divided via the anterior approach to the hip joint and five patients in whom the nerve was damaged from the lateral approach for total hip replacement. The cases are reviewed and an anatomical presentation is made to show the ease with which the Hohmann (anterior) retractor can directly compress the femoral nerve, despite the distance from the laterally placed incision. The nerve injury may be further aggravated by stretching the fascicles in overcoming pre-operative fixed flexion deformities. The results of operative exposure of the damaged nerves have been very poor because of local anatomical features at the point of injury. The major problem is the early branching of the nerve once it passes beneath the inguinal ligament so that it is difficult to find distal stumps to which nerve grafts might be sutured. The pathology is reviewed and an electronmicroscopic study of one of the nerve biopsies is presented. This reveals an almost total obliteration of myelinated fibers and the survival of a few Schwann cells and non-myelinated axons.

(b) Anticoagulant therapy

A patient suffering from bilateral femoral nerve compression resulting from bilateral iliopsoas haemorrhage is presented. These nerve injuries complicated anti-coagulant therapy. Radiological studies, essential to the formulation of the correct diagnosis, will be shown and the operative exposures described. The patient made an excellent improvement on the right side and a moderate improvement on the left.

The conclusion is that the successful management of iatrogenic femoral injuries presents a particularly difficult clinical exercise and therefore the emphasis must be on education to prevent their occurrences.

Lymphocyte Transformation Responses in Multiple Sclerosis Related to HLA Type

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We have studied 224 clinically definite Multiple Sclerosis (MS) patients for HLA A & B and 121 of these for 6 specificities of HLA-D. We found that HLA-A3 (37%), B7 (41%), B18 (12%), and Dw2 (47%) are significantly increased over control values. Dw3 antigen was diminished from 11.2% in normals to 3.3% in MS patients. Twenty-nine of these patients, all in a stable phase of disease, were studied with PHA, Con A, PWM, and herpes simplex virus as stimulants for lymphocyte transformation. Mitogens were studied against unfractionated lymphocytes, T cells (E rosetting), non-T cells (non-E rosetting), and 50-50 recombination of T and non-T cells. The herpes simplex viruses were studied against unfractionated lymphocytes. There was no clear reduction or exaggeration of mitogen responsiveness related to HLA types, though the separated T cells from Dw2 positive patients had a greater response to Con A and PHA than cells from Dw2 negative patients. Non-T cells did not respond in any significant fashion to these mitogens. T cell preparations generally had lower responses per cell than did unfractionated lymphocytes. Recombination of the T and non-T preparations generally augmented the response. Using two different herpes simplex (type 1) antigens the Dw2 positive patients (N=13) had much lower responses than did Dw2 negative patients (N=12). Almost all (9/10) of the patients who had a negative response or a very low response carried the Dw2 antigen. Four patients with Dw2 responded in what seemed to be a normal fashion. Chi square testing showed that this difference was statistically significant using each of the herpes simplex antigens separately with a P level < 0.05 using one antigen and a P value of < 0.01 with the other antigen. A relationship between an HLA linked "susceptibility factor" in Multiple Sclerosis and the postulated immune response (Ir) gene has been suggested many times. The finding of a Dw2 related hyporesponsiveness to herpes simplex antigen certainly supports the concept that the HLA-D locus is very close to the Ir locus on chromosome six.

A Photographic Method for Enhancing the Appearance of Oligoclonal Globulin in Cerebrospinal Fluid

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The study of cerebrospinal fluid (CSF) proteins in Multiple Sclerosis (MS) has shown that there is a qualitative change in the gamma globulin. This has been referred to as the oligoclonal aspect of the CSF proteins. The usual identification of this pattern is by electrophoresis of concentrated CSF in agar gel preparations. We have adapted a method electrophoresis using cellulose acetate strips and photographic enhancement for increasing contrast. CSF samples are centrifuged to remove debris. Proteins are concentrated x 100 by use of a minicon B15 concentrator. Cellulose acetate electrophoresis is performed using a tris-barbital-sodium barbital buffer at PH 8.8 for 40 minutes at 200 v. Strips are stained with ponceau S, destained, cleared, and dried. The dried strips are placed on chalk paper under matte glass and photographed at multiple exposure times (f 3.5) using Kodak high speed film No. 2575. This photographic enhancement brings out contrast in staining. One hundred-forty-five patients have been studied. Fifty-three patients satisfy the Schumacher (1965) diagnostic criteria for clinically definite M.S. In 37 of these (70%) an oligoclonal pattern was seen. In another 10 (19%) suspicious changes were seen. Six patients were completely normal, but many of these had inadequate testing because of lack of adequate concentration (< x 100) of the cerebrospinal fluid. Forty-four suspected MS patients were studied and 36% of these had the oligoclonal pattern. Two patients with SSPE had been studied and they showed markedly positive oligoclonal patterns. Forty-six neurological control patients were studied and none had the oligoclonal pattern. This adaptation of widely used methods will enable CSF protein electrophoresis to be performed as a routine procedure for the identification of an abnormality which may be specific to Multiple Sclerosis and SSPE.

**Mécanismes dopaminergiques dans le cortex cérébral.
Etudes microiontophorétiques**

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En cours de superfusion du cortex occipital avec la dopamine (DA), on observe, chez le chat, une augmentation de l'amplitude de l'onde négative lente (ONL) du potentiel évoqué visuel. Cependant, l'éjection microiontophorétique de DA dans le cortex sensori-moteur du rat entraîne habituellement une diminution prolongée (4 à 6 minutes) de la fréquence de décharge. Nous avons examiné l'effet de l'administration microiontophorétique de la DA sur les neurones du cortex occipital du chat, activés par stimulation électrique du nerf optique. L'éjection de DA réduit la fréquence de décharge de ces neurones activés par voie physiologique. La réduction de l'excitabilité neuronale est maximale pendant 2 à 4 minutes et d'une durée totale de 5 minutes environ. Par contre, une éjection d'acétylcholine sur les mêmes neurones augmente leur fréquence de décharge et dure quelques secondes seulement. Ces résultats suggèrent que la DA endogène peut agir sur le degré d'excitabilité des neurones corticaux en déprimant leur activité. L'augmentation de l'amplitude de l'ONL du potentiel évoqué visuel en cours de superfusion avec la DA exogène pourrait correspondre à un hyperpolarisation des neurones corticaux.

The Neurophysiology of Human Information Processing

T. W. Picton, E. Courchesne, K. B. Campbell and D. T. Stuss

When a stimulus is informative its evoked potential contains a late positive component with a peak latency of 300-400 ms, and with a centroparietal scalp distribution. Such a component is particularly evident when the stimulus provides feedback information about perceptual or motor performance.

The information content of a stimulus is defined as minus the logarithm of its probability, and thus the information content of confirming or disconfirming feedback can be varied by changing their relative probability. In a task involving the detection of near-threshold sounds, the probability of being correct was altered by changing the intensity of the sound; and in a task involving a one-second time estimation, the percentage of correct responses was manipulated by changing the required accuracy of the estimation. In both paradigms the amplitude of the late positive component increased with increasing information content of the feedback stimuli.

During trial-and-error learning, feedback information is used to initiate new hypotheses for testing. In a paradigm derived from clinical card-sorting tests, subjects were asked to determine by trial-and-error the correct sorting criterion (e.g. colour, shape, etc.) for visual stimuli. Prior to discovering the correct criterion, the feedback evoked potential contained in addition to the large late positive component an even later positive wave in the 500-800 ms latency range, that was quite distinct from the preceding positivity by being of greater amplitude in the parieto-occipital regions. These two positive waves therefore possibly represent different stages of information processing: the evaluation of incoming information, and its utilization in perceptual re-adjustment.

Effect of Met-enkephalin and Morphine on 1-Noradrenaline-Stimulated Cyclic AMP Formation in Rat Cortex and Hypothalamus

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Met-enkephalin (Met-enk), an endogenous pentapeptide, has morphine-like activity, but the mechanism of its action is unknown. In the present study, we investigated the effect of Met-enk and of morphine on basal and 1-noradrenaline-stimulated cyclic AMP formation in homogenates of rat cortex and hypothalamus. In both brain regions, morphine and Met-enk reduced the basal level of cyclic AMP formation slightly, while naloxone alone had no effect. Both morphine (10^{-5} M) and Met-enk (1.0μ g/ml) inhibited the 1-noradrenaline-stimulated cyclic AMP formation by approx. 50% in both brain regions. This inhibitory action was reversed by naloxone (10^{-5} M). The results suggest that Met-enk may exert central effects by interacting with noradrenergic receptor in the brain.

Fungal Intracranial Masses

J. L. Robinson, P. Forcier, J. C. Péchère and J. A. Chabot

Two cases of fungal cerebral space occupying lesions will be presented. The first was operated on elsewhere with a diagnosis of cerebral tumour — excision was macroscopically complete. The lesion recurred after 6 months, and because of doubt about the pathology, a second craniotomy was performed. A candida albicans abscess was found, and review of the microscopic slides confirmed that this had been the original diagnosis. Only subtotal resection was possible, but with systematic antifungal medication the lesion was sterilized. The second, caused by cryptococcus neoformans, presented as an occipital tumour with raised intracranial pressure. As well as a total resection of this large tumour, we immediately gave systemic antifungal treatment with a satisfactory result. Neither patient had any underlying pathology, or had had treatment, predisposing to the development of fungal infections. There were no lesions present in any of the other systems. In the differential diagnosis of an intracranial mass, large fungal granulomata should be considered even if the patient does not have a generalized debilitating condition. Systemic antifungal therapy causes many toxic side effects. Nevertheless even a macroscopically total resection of these lesions should be followed up by drug treatment if one hopes to cure the patient.

**Projection Pontocerebelleuse. Etudes Anatomiques
Experimentales Chez le Chat**

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Les projections des noyaux pontiques au cortex et aux noyaux cérébelleux ont été analysées à l'aide de méthodes expérimentales. Après des injections de L'leucine tritiée impliquant la quasi totalité des noyaux pontiques, on a d'abord recherché la distribution globale dans le cortex cérébelleux. Cette projection se fait au niveau des portions latérales du lobe antérieur et sur tout le lobe postérieur. La densité maximale de projection se trouve au niveau du vermis des lobules VI, VII et VIII. Il n'y a peu ou pas de projection dans le lobe flocculonodulaire. Il n'y a que très peu de fibres pontiques qui se distribuent dans la partie centrale du lobe antérieur correspondant au vermis et à la région paramédiane. La projection est principalement croisée mais une distribution directe se rencontre dans toutes les parties du cervelet déjà mentionnées. Une autre série d'expériences comportant, des lésions des noyaux cérébelleux et l'étude de la dégénérescence confirme cette distribution. Dans les noyaux cérébelleux, on n'a retrouvé qu'une projection peu abondante avec l'une et l'autre des deux méthodes expérimentales. La projection se fait sur les noyaux dentelés et elle est surtout croisée. Il a été possible d'établir que la source de cette projection dans la protubérance est le nucleus reticularis tegmenti pontis. Se l'on compare la projection de la protubérance avec les autres afférences à fibres moussues des lobes antérieur et postérieur, il apparaît que les zones de chevauchement sont peu nombreuses, ce qui justifie le concept de pontocerevet proposé par Brodal.

Human Spinal Cord Protein (HSCP): Partial Characterization and Localization

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MacPherson and Yo reported earlier (J. Immunol. 110, 1371, 1973) that guinea pigs pretreated with a bovine spinal cord protein (BSCP) did not develop experimental allergic encephalitis (EAE) when they were subsequently injected with disease-inducing doses of bovine spinal cord (SC) or purified bovine myelin basic protein (BP). Immunochemical studies showed that human SC and human SR contained proteins analogous to the bovine proteins.

In order to investigate whether HSCP also had the capacity to prevent immunologically induced demyelinating disease, HSCP was isolated from HSC and HSR by, 1) extraction with 0.15M sodium chloride, 2) absorption on CM-52 cellulose, 3) stepwise elution with sodium acetate buffers at pH 5.8 containing increasing concentrations of sodium chloride, and 4) gel-filtration on Sephadex G-50.

HSCP isolated from HSC and HSR have an electrophoretic mobility in agar at pH 8.5 corresponding to a β -serum globulin and occur in two antigenically distinct molecular forms which may be separated by isoelectric focusing. The HSCP proteins from SC and SR cross-react extensively with anti-BSCP serum and have amino-acid compositions and molecular weights similar to those of BSCP isolated from BSC and BSR. Immunohistological studies with anti-HSCP sera indicate that the protein is localized mainly in axons in the central and peripheral nervous systems and seems to be absent from myelin.

Efficacy of Thromboendarterectomy for Total Occlusion of the Internal Carotid Artery

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Forty consecutive patients on whom 42 thromboendarterectomies were performed for total internal carotid artery occlusion were followed for a mean period of 50 months.

Patency was successfully restored in 52% of the procedures, though only 58% of these arteries remained patent on late angiography. Eighty-six percent of arteries operated on within 72 hours of the apparent time of occlusion were successfully reopened.

Patients were divided into two groups on the basis of presence or absence of neurological, medical and angiographic risk factors. The low-risk group experienced 0 postoperative mortality and 7% postoperative morbidity, compared with 22% and 41% respectively in the high-risk group ($p < 0.05$).

On long-term follow-up, patients with persistent patency of the internal carotid artery showed 0 mortality and 0 morbidity, compared with 17% mortality, 21% cerebral morbidity, and 25% overall morbidity in patients whose arteries remained occluded. This is a significant difference in overall morbidity and mortality between patients with persistent patency and those with persistent occlusion ($p < 0.05$).

In summary, the success rate for restoration of patency in total occlusion of the internal carotid artery justifies early operation. Low-risk patients can be selected preoperatively. The long-term prognosis appears to be better in patients with persistent patency than in those with persistent occlusion.

The Use of Halo Devices for Patients with Acute Spinal Cord Injury

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During the past three years, we have used various halo devices for the treatment of patients with acute cervical spinal cord injury. The halo devices are used for the reduction of spinal deformity, immobilization of the spine, and for early mobilization of the patient. The most useful of these devices has been the low profile halo vest assembly, which has been used extensively in patients with either complete or incomplete spinal cord injury. There have been no instances of pressure sores developing in areas of cutaneous anaesthesia in patients with complete spinal cord injuries. Operations for cord decompression or spinal fusion have been performed without difficulty from either anterior or posterior approaches with patients in the halo vest assembly.

Although they have some limitations and shortcomings, we have found halo devices to be extremely helpful in the immediate and later stages in the care of patients with acute cervical cord trauma. In our Unit, they have almost completely replaced the use of skull tongs.

Effects of Short and Long Term Delayed Local Cooling in Spinal Cord Injury

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The beneficial effects of local cooling after spinal cord injury were reported last year. The current series is to determine whether delayed prolonged cord cooling is of added benefit.

Dogs with an experimental compression cord injury were divided into untreated and treated groups. In treated dogs the injured spinal cord was cooled using an extradural heat exchanger at 6°C for 1, 4 or 20 hours, beginning four hours after injury. Dogs were observed until neurologically stable or for at least two months after injury.

Untreated animals failed to attain significant motor recovery. Those undergoing four hours of delayed localized cord cooling had significantly enhanced motor recovery. Those undergoing cord cooling for one hour showed a similar but less dramatic improvement. Animals undergoing prolonged local cord cooling of 18 to 20 hours duration did not regain significant motor improvement compared with untreated dogs. This failure of improvement is not due to the prolonged presence of the extradural cooling apparatus.

CONCLUSIONS:

1. This confirms previous work in this laboratory that short-term-local cord cooling is beneficial following injury.
2. Delayed prolonged (greater than 4 hours) cooling does not provide additional clinical benefit.

Oxiperomide, Dyskinesias and Parkinsonism

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Dyskinesias are the most frequent complication of treatment in Parkinsonism. They appear to result from overstimulation of certain dopaminergic receptors in the striatum. Attempts to reduce them pharmacologically have been hitherto always accompanied by a corresponding return of the Parkinsonian symptomatology.

Recent experimental evidence (Costall and Naylor 1975) suggests that oxiperomide could have a more selective blocking effect and might influence the dyskinesias without affecting the Parkinsonian symptomatology.

We have therefore selected six patients suffering from Parkinson's disease with prominent treatment-induced dyskinesias. Oxiperomide was added to their usual drug regime, starting at a dose of 5 mg/day and increasing progressively to 15 mg/day.

All patients had a significant reduction of their involuntary movements (50 to 100%). The improvement was sustained for as long as therapy was continued 2.5 to 4 months. Best results were obtained when the dyskinesias occurred at a fixed time of the day as we found the protective effect of oxiperomide to last about six hours.

On 10 mg or more a day, there was an increase in the Parkinsonian symptomatology, but in five patients it could be controlled either by reducing the dose of oxiperomide or by increasing the dose of antiparkinsonians. These five patients had the same degree of control of their Parkinsonian symptomatology without dyskinesias. We also tried oxiperomide in thirteen patients suffering from spontaneously occurring dyskinesias. 3 patients suffered from Huntington's chorea, 1 from Tourette's syndrome, 5 from Generalized Torsion dystonia and 4 from adult-onset focal dystonia. Oxiperomide was given to these patients as a single oral dose of 10 mg. The three patients with chorea and the one with Tourette's syndrome had a 50% improvement. Of the dystonic patients only two of the adult cases one with torticollis and one with oro facial dystonia had an improvement of more than 50%.

Distribution of Acetylcholinesterase within the Midbrain, Pons, Medulla and Spinal Cord of DFP-Treated Monkeys

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Five monkeys pretreated with di-isopropylfluorophosphate (DFP) were used to study the distribution of acetylcholinesterase (AChE) within the neurons of the brainstem and spinal cord. Following DFP treatment AChE activity within the neuropil is considerably reduced and as a consequence the neurons are easily identified. An intense activity is present in the soma and processes of the following groups of neurons: the motor nuclei of the III, IV, V, VI, VII, X and XII nerves including nuclei Edinger-Westphal and dorsalis of X; the neurons of the ventral horn and nuclei intermedio-medialis and intermedio-lateralis; neurons of the pedunculo-pontine tegmental (subnucleus compactus) and the dorsolateral tegmental nuclei, the magnocellular division of the

red nucleus, nuclei pontis oralis, pontis caudalis and gigantocellularis and the lateral vestibular and fastigial nuclei. Different types of neurons may be observed in the locus coeruleus and more especially in the different divisions of the substantia nigra. A moderate AChE activity of the soma and processes is present in the interstitial nucleus of Cajal and the dorsal nucleus of Clarke. A light to moderate AChE activity of the soma and a weak or undetectable activity of the processes is shown by the neurons of the nucleus of the mesencephalic root of V, the nuclei dorsalis raphae, annularis, linearis and ventralis of Gudden, the pontine nuclei, the cerebellar dentate and interpositus nuclei and the principal and accessory inferior olivary nuclei. In areas such as the pontine gray and olivary nuclei where a retrograde degeneration of the neurons occurred as a consequence of cerebellectomy the AChE activity of the neuropil was not restored following DFP treatment.

Baclofen: Selective Inhibition of Excitatory Amino Acid Release

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Electrophysiological evidence suggests that the antispastic agent Baclofen (β -(p -chlorophenyl)- γ -aminobutyrate, Ciba-Geigy) may act by rendering afferent terminals less excitable [S. Fox et al., *Neurosci. Abs.*, 2, 1003 (1976); R. A. Davidoff and E. S. Sears, *Neurology*, 24, 957 (1974)] and so depress the release of some transmitters in the CNS. Its effect on release of tissue amino acid transmitters (labelled via the metabolism of [U- 14 C] D-glucose) and exogenous labelled amino acid transmitters (preloaded into the tissue) was studied using slices of guinea pig cerebral cortex, release being evoked by electrical field stimulation. Baclofen (4 μ M) had no effect on the release of tissue 14 C-GABA but depressed that of tissue 14 C-aspartate and 14 C-glutamate by 40-60%. Release of preloaded 3 H-GABA was depressed by 40% and that of pre-loaded 14 C-aspartate and 14 C-glutamate by 60-70%. It is unlikely that changes in the release of tissue amino acids were due to depleted pools as the level of tissue aspartate decreased only 25% while those of tissue glutamate and GABA increased 20% and 14% respectively. Moreover, incorporation of 14 C from [U- 14 C] D-glucose into tissue amino acids paralleled the respective changes in their levels. It was concluded that the action of Baclofen in cerebral cortex is to depress the release primarily of excitatory amino acids evoked by electrical stimulation. The mechanism of this inhibition may be similar to the reduction in afferent terminal excitability observed by others [S. Fox et al., (1976); R. A. Davidoff and E. S. Sears (1974)].

Effect of Baclofen on Synaptosomal Uptake and Release of Putative Amino Acid Transmitters

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The antispastic agent, Baclofen [β -(p -chlorophenyl) γ -amino-butyric acid, Ciba-Geigy Ltd.], at a concentration of 10 μ M, inhibited the uptake of 14 C-glycine into guinea pig cortical synaptosomes by 18% and 30% when medium glycine concentrations were 7.5 and 150 μ M, respectively. Similar but smaller inhibitions (8% and 13%) were also observed with synaptosomes isolated from spinal cord. The uptake of 14 C-glutamate, -aspartate and -GABA into either cortical or spinal synaptosomes was not affected. The release of both exogenous 14 C-glycine and -GABA from cortical synaptosomes evoked by 66 mM KCl, was unaffected by Baclofen but that of -glutamate and -aspartate was consistently inhibited by 10%. Baclofen had no effect on the release of any of these amino acids from spinal synaptosomes. These results suggest that the antispastic action of Baclofen may be explained in part by a net facilitation of inhibitory mechanisms due to an inhibition of glycine uptake. Moreover, because Baclofen inhibits excitatory amino acid release from cortical slices evoked by electrical stimulation (S. J. Potashner, these proceedings) but is without effect on amino acid release from synaptosomes evoked by very strong potassium depolarization, it seems likely that in intact tissue, Baclofen may hyperpolarize afferent terminals [cf. S. Fox, *Neurosci. Abs.*, 2, 1003 (1976); R. A. Davidoff and S. Sears, *Neurology*, 24, 957 (1975)] and depress their invasion by action potentials.

Effects of Kainic Acid Diethylester on Dorsal Horn Interneurons in Cat Spinal Cord

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Glutamate (GLU) and aspartate (ASP), two acidic amino acids (a.a.), are likely candidates for excitatory transmitters in vertebrate central nervous system (CNS). Kainic acid (KA), a natural analog of GLU, is a more potent excitant than GLU. The diethylester of KA (KDEE) strongly depressed acidic a.a. responses and some synaptic potentials in frog spinal cord (Padjen, A., *Proc. Meet. Can. Fed. Biol. Soc.* 19: 18, 1976). In order to characterize these interactions as well as to study structure-activity relationship of excitatory a.a. in CNS we have examined the effects of microiontophoretically applied KDEE, glutamate diethylester (GDEE) and acidic a.a. on extracellularly recorded firing rates of single dorsal horn interneurons in cat spinal cord. KDEE and GDEE usually depressed spontaneous neuronal activity without changing the amplitude of the action potentials; in that respect KDEE action outlasted that of GDEE. Spike activity of superficial dorsal root fibres was insensitive to both esters. KDEE reduced the neuroexcitatory effect of GLU, ASP, KA, quisqualic acid and acetylcholine as well as the neuronal firing evoked by peripheral sensory stimulation. These effects were reversible and related to the iontophoretic currents employed. Our results indicate that esterification of KA as well as of GLU converts these excitatory a.a. into depressant compounds. The mode of action of these esters remains to be clarified.

Idiopathic Facial Palsy and Sensitization to Peripheral Nerve Myelin Basic P2 Protein

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Bell's Palsy has only recently been thought of as possibly being an auto-immune disease. To evaluate this concept we have employed the Bendixen leukocyte migration inhibition (LIF) assay using a basic (P2) protein of peripheral nerve myelin as antigen. Thirty-nine controls and 12 facial palsy patients were assayed with P2 protein, myelin basic protein (BP) and acetyl-choline receptor (AChRP). The group of normals gave a % migration of 100 ± 13 , 96 ± 8 , and 103 ± 16 . Facial palsy patients gave values of 80, 87, and 96: those with miscellaneous diseases 107, 87, and 104; and multiple sclerosis patients 103, 78, and 94. Positive results were regularly found in the first week after onset of facial paralysis, and occasionally in the second or third week. Similar results were seen in some Guillain Barre, brachial neuritis, and surprisingly in some ALS patients. However, only 1/12 MS patients, and no stroke patients (2 with lower motor neuron facial nerve involvement). Positive BP results were seen in 3 facial palsy, and 12 miscellaneous, including 7 of 12 MS patients. No positive results were seen with AChRP.

Results of an earlier investigation by Abramsky et al. demonstrating cellular immunity in Bells Palsy are confirmed. Our findings, however, do not explain important differences such as unilaterality of facial involvement in "Bells Palsy" as opposed to Guillain Barre. Idiopathic facial palsy appears to be a suitable disease for study of the interrelationships of virus infection and immune aberrations which may underly the destruction of myelin.

Dendritic Abnormalities in Human Epileptic Neocortex: A Golgi-Cox Study

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We found clearly defined abnormalities in the dendrites of all six cases in whom epileptogenic neocortex was resected for alleviation of seizures and was stained by the Golgi-Cox technique. The changes included reduction or loss of dendritic spines, a beading deformity on the dendritic shafts, and concertina-like folding of the dendrites. Resection sites were in the occipital lobe in 3 cases, frontal lobe in 2 cases, and in the Rolandic region in one case. Routine stains revealed that the majority had ectopic neurones in the white matter and minor cortical architectural abnormalities. Five had gross structural abnormalities as well, such as a biopsy scar (one case).

Our findings indicate that the similar changes described by the Scheibels in human epileptic hippocampus can be extended to include epileptogenic foci in the neocortex as well. The findings resemble closely the dendritic abnormalities described by Westrum et al. in experimental epileptic neocortex in the monkey.

We are unaware of any previous description of dendritic abnormalities in epileptogenic human neocortex in the modern literature.

Clinical Experience with Rivotril in Children

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The efficacy of Rivotril in 21 young epileptics (15 boys and 6 girls) aged 4 months to 17 years has been studied. The patient sample includes 14 cases of generalized myoclonic epilepsy and 5 cases of partial epilepsy with elementary or complex symptomatology. In addition, one patient suffering from a degenerative disease of the central nervous system and one patient with a Lennox Gastaut syndrome were included in the trial. The results show that Rivotril has the greatest therapeutic benefit in patient suffering from myoclonic epilepsy (absence). No definite conclusion can be made regarding the efficacy of Rivotril in other forms of epilepsy due to the small number of patients included in the trial and also to the fact that the patient had been very resistant to other drugs. The patient with the Lennox Gastaut syndrome showed a temporary clinical amelioration. However this effect was short lived and the patient relapsed after 3-4 weeks of treatment. The patient suffering from progressive degenerative disease of the central nervous system did not respond to the treatment administered.

The most frequent side effect was somnolence which was observed in 10 out of 21 patients. Aggressiveness, periods of agitation and crying spells were noted in 4 patients. No significant biochemical or hematologic changes were noted during treatment with Rivotril.

Etude Comparative des Hématomes Epiduraux à l'Hôpital de L'Enfant-Jésus — 1971-1975

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Nous présentons une étude statistique et comparative des hématomes épiduraux traités à l'Hôpital de l'Enfant-Jésus de 1971 à 1975. Nous avons retenu 28 dossiers sur 44, lesquels représentent des hématomes épiduraux "purs", c'est-à-dire non associées à d'autres lésions hémisphériques du type de la contusion cérébrale, lacération ou hématome sous-dural.

L'incidence annuelle est de 7% et s'est rencontrée surtout chez le jeune patient de sexe masculin. C'est surtout le traumatisme à basse vélocité qui fut responsable de la lésion. Treize patients (46%) ont présenté l'intervalle lucide typique de l'hématome épidural. Onze patients (40%) se sont présentés inconscients à la chirurgie dont six (6) l'étaient devenus par la seule progression de leur hématome.

Seulement 25% des patients ont présenté les signes classiques de l'hématome épidural et plus de 25% étaient décérébrés. 50% des patients étaient bradycardes, la majorité des hématomes étaient situés à la convexité et variaient de 50 cc à 300 cc. Six (6) patients sont décédés dont trois (3) avaient présenté un coma d'emblée après le traumatisme. Trois (3) patients quittèrent avec des déficits neurologiques et 19 patients (68%) ont quitté neurologiquement intacts. Il n'y a eu aucun décès chez les patients qui se sont présentés conscients ou semi-conscients à la chirurgie, alors que la mortalité fut de 71% chez les patients inconscients.

Regional Cerebral Blood Flow, Moyamoya Syndrome and Neurofibromatosis

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The association of neurofibromatosis with multiple intracranial arterial stenosis and thrombosis is known but extremely rare (1).

We had the opportunity to study such a case with angiographic and regional cerebral blood flow studies in a 22-year-old woman who had consulted for a chronic pulsating headache.

Three years prior to admission this patient had experienced a right hemiparesis with aphasia which cleared almost completely over a year. On examination she had the classical stigmata of Von Recklinghausen disease and a very slight right hemiparesis.

The angiographic studies revealed a complete obstruction of the terminal part of the right carotid artery, an almost occluded left middle cerebral artery and a marked stenosis of the left posterior cerebral artery, left anterior cerebral artery and basilar artery.

There was a classical radiological picture of Moyamoya syndrome in the central regions brought in by retrograde filling via the posterior communicating arteries and end to end anastomosis.

In spite of such an impaired intracerebral circulation, the regional cerebral blood flow studies, with a 35 detector apparatus and using a technique previously described (2), revealed an homogeneous and normal cerebral blood flow in both cerebral hemispheres.

Moreover a selective external carotid injection of Xenon-133 revealed an increased flow which tends to prove that there is more collateral flow adjusting to her disease, thus perhaps explaining her vascular headaches.

REF.:

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Experience in Dermatomyositis of Childhood

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Since 1967, 9 children with dermatomyositis have been treated at the Izaak Walton Killam Hospital for Children. All but one have been seen by one of the authors (J. T.) The rarity and variability of this disease makes it hard to draw conclusions but from the experience gained in following these children, the following points emerge:

1. The differential diagnosis from other collagen vascular diseases or muscular dystrophy can be extremely difficult.
2. The diagnostic value of muscle biopsy is controversial but skin biopsy and nail fold microscopy may give added information.
3. The subtle prolonged and unpredictable course of the disease requires close supervision over many years.
4. Steroids remain the initial drug of choice, though their side effects were disastrous in 4 out of 9 patients. Other immunosuppressants may offer a satisfactory alternative in some situations.
5. Pencilamine was effective in the management of contractures in the 2 patients so treated.
6. The long term prognosis of our patients is in agreement with other published experience.

Cases that illustrate these points are shown; the lesson to be drawn from them is that meticulous care and anxious appreciation of the possible complications are necessary for the management of these patients.

Aluminum: A Neurotoxic Factor in Senile (Alzheimer) Dementia

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Several lines of evidence implicate the trace metal aluminum in the pathogenesis of Alzheimer's disease. Aluminum stimulates certain neurons to produce excessive amounts of 100 Å diameter filaments in experimental animals at brain concentrations similar to those found in the human disease. Furthermore, human fetal cerebral neurons grown in tissue culture and exhibiting synapse formation, exposed to concentrations of aluminum found in Alzheimer's disease, develop neurofibrillary degeneration and die. The sites of aluminum binding with chromatin are DNA and acidic proteins. The effect of aluminum upon the thermal denaturation and refolding of DNA indicate that aluminum stabilizes guanine-cytosine bonds and has a specific affinity for adenine-thymine base pairs. In addition aluminum interacts with chromatin acidic proteins and thereby may alter gene transcription. These interactions of aluminum with molecules involved in gene expression may determine the mechanism of toxicity of this element.

C.T. of the Spinal Canal Using the Limited Traverse Technique

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S. Taylor and C. Thompson

The EMI body scanner has the possibility of scanning a section of the body which can be restricted, for example, to the spinal canal and surrounding tissues. Normally, the 13-inch scan mode would be utilized to scan the abdomen of an average adult patient. This provides limited information regarding the spinal canal in its content. By using the 10-inch scanning mode, it is possible to "zoom" down on the spinal canal, so that the photon activity of the x-ray beam can be concentrated on that particular area. The usual thickness of the slice is 1.3 cms.; work is being carried out to reduce the thickness of the slice to 5 mms. in order to improve the definition of the scanned area. We hope to develop a scanning mode of 5 to 6 inches in order to cone down even more and enhance the diagnostic capabilities. Primary results will be shown and discussed.

**Hyperkinetic Behaviour and Absence Attacks with
Left Cerebral Atrophy in Children**

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This paper presents four children with a picture of hyperkinetic behavior and absence attacks. Three boys and one girl aged 6, 8, 11, and 6 years respectively were seen with the following manifestations: Absence attacks starting at 29, 36, 60 and 35 months of age, all of them associated with hyperkinetic behaviour starting about the same age and

only one of them showed aggressive behaviour. All were right handed. During the last trimester of pregnancy the mothers of these children developed a viral respiratory infection. No history of childhood viral diseases. EEG showed profound changes in all, consisting of poorly organized cerebral activity mainly in the left side. Isolated spike and wave complexes in three and multiple spikes in the other one. This spike activity was much more pronounced on the left temporal lobe. Pneumoencephalogram showed left ventricular dilatation and various degrees of cerebral atrophy on the same side. Development and language were normal until the disease started following which progressive dysphasia ensued.

Due to lack of response of seizures to medical therapy left temporal lobectomy was performed in all with diagnostic and therapeutic purposes. Better control of hyperkinetic behaviour and seizures was achieved following operation.

Light microscopy showed mild increase of cellularity predominantly astrocytic. Ballooning and vacuolated cytoplasm was seen in astrocytes surrounding the neurons at the base layer. The neurons showed karyolysis, swelling and loose nuclear chromatin, some clusters of cells showed shrinkage with loss of Nissl granulations, acidophilia and cytoplasm swelling. No inflammatory infiltration or other changes of inflammation were seen, perivascular spaces were empty. Electron microscopy in two cases showed intracytoplasmic inclusion bodies.

This picture of Hyperkinetic Behaviour with Absence Attacks and progressive Dysphasia in otherwise normal children may be due to slow virus disease acquired from the mother during the last trimester of pregnancy. More research is needed to rule out other etiologic factors as well as culture and identification of viruses to clarify this interesting condition.