# Canadian Association of Neuropathologists

## Abstracts of papers and cases presented at the 40th Annual Meeting

*October 18 - 21st, 2000*

*Banff, Alberta*

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The 40th annual meeting of the Canadian Association of Neuropathologists was held from October 18 - 21st, 2000 at The Banff Centre in Banff, Alberta. Local arrangements were made by Drs. Bernadette Curry and Barry Rewcastle. The scientific session consisted of 15 platform presentations and 13 cases for diagnosis. The Royal College of Physicians and Surgeons of Canada speaker was Dr. James Rutka, Professor and Chair, Department of Neurosurgery, University of Toronto and The Hospital for Sick Children. His talk was entitled "New Insights into the Molecular Genetics and Pathology of Human Brain Tumors". The Jerzy Olszewski lecturer was Dr. Voon Wee Yong, Associate Professor, AHFMR Senior Scholar and MRC Scientist, Departments of Oncology and Clinical Neurosciences, University of Calgary. His talk was entitled "Complexities of Neuro-inflammation: Pathology and Regeneration". The invited lecturer was Dr. Catherine Bergeron, Neuropathologist, Centre for Research in Neurodegenerative Diseases, University of Toronto and Toronto Western Hospital. Her talk was entitled "The Many Faces of Human Prion Diseases".

### PLATFORM PRESENTATIONS

#### 1. Ethylmalonic encephalopathy: a first neuropathological report.

*J. MICHAUD and N. LEPAGE (Children's Hospital of Eastern Ontario and University of Ottawa, Ottawa, ON)*

Ethylmalonic encephalopathy (EE) is biochemically characterized by persistent lactic acidemia and ethylmalonic and methylsuccinic aciduria of unknown cause. We report the neuropathological findings in a nine-year-old girl who had a younger brother with the same clinical presentation. She died in a status of metabolic crisis. The brain weighed 1270g. The striatum showed the most severe changes. There were several small multicystic areas surrounded by a firm white rim. Histologically, the focal cystic degeneration was confirmed and associated with a dense fibrillary gliosis with numerous Rosenthal fibers. The density of small blood vessels was increased in other areas of the striatum, in the basal forebrain, along the third ventricle, in the substantia nigra and in the periventricular gray matter of the upper brain stem. This was associated with endothelial hyperplasia, tissular spongiosis, gliosis, neuronal loss or recent neuronal damage. Recent infarct-like foci were found in the basal ganglia, the posterior hypothalamus and the white matter of the cerebellum. Dominant changes in the striatum corroborates the diagnostic imaging data available from CT, MRI and PET studies. Recent changes in a period of metabolic crisis corroborates published reports of rapidly emerging striatal lesions. Also the vascular abnormalities, although of nonspecific nature, tend to support the clinical impression of a vasculopathy as part of the physiopathological events underlying the clinical and biochemical picture in this rare metabolic disorder.

#### 2. The neuroanatomy of Klippel-Feil Syndrome: Two more cases.

*D.H. GEORGE[1], J. DONAT[2] ([1]Foothills Hospital, Calgary; [2]Royal University Hospital, Saskatoon,SK)*

The Klippel-Feil Syndrome (KFS) is a congenital fusion of cervical vertebrae, with the triad of short neck, low hairline, and restricted neck movement. KFS may have neurological and neuroanatomical abnormalities. The most striking functional abnormality is "mirror movements". Radiology has demonstrated cervical cord dysraphism in many cases of KFS, but pathological examination is limited to two autopsy reports, one of spinal cord only. Features include: hydrocephalus, thin corpus callosum, downward displacement of the lower brain stem and cerebellum, posterior dysraphism of the medulla and cervical cord, agenesis of the pyramidal decussation, brain stem and spinal cysts, central canal duplication, and lumbar diastematomyelia. We have examined two more cases of KFS, one a 26-year-old male with mirror movements and the other a 61-year-old woman with no neurological history. The first case included brain and medulla to C1; the second included the whole

spinal cord. All the previously described neuropathological features of KFS were found in one or both of these new cases. The most consistent and severe malformations were posterior cervical cord dysraphism, and failure of decussation of the pyramidal tracts. In addition, one case had dysplasia of the medial temporal lobes; the other case had asymmetrical medullary pyramids, and hypoplasia of the left hypoglossal nucleus. These are newly described malformations for KFS. The absence of pyramidal decussation in KFS may form the anatomical substrate for mirror movements.

**3. Ubiquitin immunohistochemistry suggests that classical MND, MND with dementia and FTD-MND type represent a clinicopathological spectrum.**

*I.R.A. MACKENZIE and H. FELDMAN (Department of Pathology and Division of Neurology, Vancouver Hospital & Health Sciences Centre and University of British Columbia, Vancouver, BC)*

One of the neuropathological features of motor neuron disease (MND) is the presence of ubiquitin-immunoreactive inclusions in brain stem and spinal cord motor neurons. Some patients with otherwise typical MND develop dementia and in these cases ubiquitin-positive inclusions are also present in hippocampal dentate granule cells and layer II neurons of neocortex. Recently, it has been recognized that an identical pattern of ubiquitin-immunoreactivity may be found in the cerebrum of some patients with frontotemporal dementia (FTD), in the absence of motor symptoms. Such cases have been referred to as FTD-MND type or MND inclusion dementia. To clarify the relationship between these conditions, we examined the pattern of ubiquitin-immunoreactivity in patients with typical MND (n=6), MND with dementia (n=6) and FTD-MND type (n=6). All cases of MND with dementia had characteristic degenerative changes in the motor system as well as ubiquitin-positive inclusions in the hippocampus and cerebral cortex. By definition, patients with FTD-MND type showed similar cerebral pathology, however 3/6 also had ubiquitin-positive inclusions in brain stem motor neurons. In MND without dementia, 3/6 had inclusions in the hippocampus and cerebral cortex which were as numerous as in the FTD patients. These findings suggest that classical MND, MND with dementia and FTD-MND type represent a clinical spectrum of neurodegenerative diseases which share a common pathological substrate.

**4. Iatrogenic Creutzfeldt-Jakob disease from human cadaveric dural transplants: review of two recent cases in adolescents.**

*Y. ROBITAILLE, J. RICHARDSON, R. LABRECQUE, L. CARMANT and S.J. De ARMOND (Dept. of Pathology & Cell Biology and Neuropathology, University of Montreal, McGill University & UCSF, Montreal, PQ and San Francisco, USA)*

The incidence/prevalence of iatrogenic Creutzfeldt-Jakob Disease (ICJD) in Canada was limited to rare occasional cases within the past few years. We wish to report the recent occurrence of ICJD in two adolescents who had surgery followed by transplants of human cadaveric dura.

This 18-year-old male was admitted for recent onset of ataxia in late December 1997. At age 2½, a pilocytic cerebellar astrocytoma had been excised. The surgical cavity was covered with a LYODURA cadaveric dural graft. He had yearly CAT scans to monitor any possible neoplastic recurrence. None was found up to a year before admission. The past medical and familial histories were non contributory for neurological disease including CJD, drug abuse, toxic exposures or seropositivity for HIV. On admission, neurological examination revealed disorientation, poor concentration and memory. Impaired judgment, mental slowing and decline in problem solving skills, visuo-constructional apraxia, dysgraphia and dyslexia were also already apparent. There were no localizing signs. DTR's were generally brisk. A few days after admission, generalized myoclonus developed, which became drug-resistant. He died one month after admission, deeply comatose with diffuse rigidity.

The second case was that of a 13-year-old girl at death. In June 1988, a left occipital lobe hematoma was removed along with remnants of an AV malformation. The surgical field was covered with a LYODURA human cadaveric graft. In April 1999, she was admitted for progressive ataxia and dysmetria. Generalized myoclonic seizures soon followed, subsequently complicated by a generalized decline of all cognitive functions. Death occurred five months after admission.

In both cases, widespread evidence of $PrP^{SC}$ peptides in post-mortem brain tissues was obtained by Western blots and/or immunocytochemistry.

**5. Anti-EBV antibodies cross-react with alpha-synuclein: an example of molecular mimicry relevant to Parkinson's disease?**

*J.M. WOULFE (Hamilton Health Sciences Corporation and McMaster University, Hamilton, ON)*

Commercial antibodies generated against viral proteins occasionally react with host tissues. Assuming that similar cross-reactive antibodies are generated in the human host during natural infection with the virus, such cross-reactions may initiate immune mediated disease through molecular mimicry. Using antibodies generated against the latent membrane protein 1 of Epstein-Barr virus (EBV), we demonstrate intense immunoreactivity of Lewy bodies in Parkinson's disease (PD) and dementia with Lewy bodies and glial cytoplasmic inclusions in multiple system atrophy. ELISA and Western blotting techniques confirmed that this immunolabelling was due to cross-reactivity of the antiviral antibody with α-synuclein, a neuronal protein implicated in the pathogenesis of PD. This example of cross-reactivity between EBV and α-synuclein may bear implications for further elucidating infectious/autoimmune mechanisms in PD.

**6. p27 expression in multiple system atrophy.**

*H.Y. SIU, L.C. ANG, and B. YOUNG (Sunnybrook and Women's College Health Sciences Centre, Toronto, ON)*

Multiple system atrophy (MSA) is a neurodegenerative disorder histologically characterized by the presence of glial cytoplasmic inclusions (GCI) in oligodendrocytes. Immuno-histochemistry, using ubiquitin, tau, α-synuclein, C-myc, and



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p27, was performed on the basal ganglia, brain stem, and cerebellum sections for 14 MSA cases, two spinocerebellar ataxia (SCA) cases, and two control cases. Both polyclonal (Santa Cruz, 1:200) and monoclonal (Bio-Can, 1:1000) p27 antibodies were used in this study. All 14 MSA cases showed positive staining in GCI for p27. Twelve cases out of 14 showed positive staining for ubiquitin, while 13 out of 14 MSA cases were positive for α-synuclein. For C-myc, 10 MSA cases were positive, while only nine cases were positive for tau. Ubiquitin and p27 stained up the highest number of inclusions out of all the antibodies used. In SCA, no GCI were detected. The specificity of the polyclonal p27 antibody was verified by blocking the antibody with a p27 blocking peptide. No blocking peptide was available for the monoclonal p27 antibody. In normal brains, p27 is observed in oligodendroglial nuclei, however, in MSA, nuclear and cytoplasmic localization is seen in the GCI. The sequestration of p27 in the cytoplasm should theoretically promote cell proliferation; however, no proliferative activity was seen when these cases were stained for Ki-67. The mechanism for the cytoplasmic sequestration of p27 in GCI, and the possible involvement of C-myc, is still undetermined.

### 7. An improved in-vitro model forAIDS Dementia Complex.

*R. HAMMOND and S. ISKANDER (London Health Sciences Centre and University of Western Ontario, London, ON)*

The mechanism underlying AIDS dementia complex (ADC) remains uncertain. As the AIDS epidemic continues and patient longevity increases, ADC is likely to increase in prevalence. Few models exist that approximate the special environment of the human CNS infected by HIV-1. An *in vitro* system using human CNS tissue dissociated to primary cultures has become a valuable tool in examining the effects of a variety of neurotoxins including HIV-1 gp120. The cultures are maintained serum-free and with no antibiotics or antimetabolites. The cultures exhibit mature, stable neuronal and glial subpopulations and can be maintained for months. In response to various challenges, neuroglial injuries can be assayed by LDH release, apoptosis (TUNEL), dendritic changes (MAP2 immunofluorescence) and astroglial hypertrophy (GFAPimmunofluorescence). In response to gp120 exposure, these cultures undergo a qualitative and quantitative simplification of their dendritic arbour and a mild gliosis, replicating the *in vivo* correlate of ADC (Masliah et al, Annals of Neurology 42: 963-972). Furthermore, the cultures can be treated to provide protection from other oxidative stresses (Cai et al, IJRB 76(7), 1009-1017) and explore therapeutic avenues for such diseases. The cultures offer new potential in modeling a number of neurodegenerative and neurotoxic conditions specific to mankind.

(Supported by a grant from the Ontario HIV Treatment Foundation (RH).)

### 8. Cloning and characterization of *Human Suppressor of Fused (hSu(fu))*, a candidate tumour suppressor gene for medulloblastoma on chromosome 10q24.3

*M.D. TAYLOR, D. HOGG, L. LIU, T.G. MAINPRIZE, S. SCHERER, J. SKAUG and J.T. RUTKA (Sonia and Arthur Labatt Brain Tumour Research Centre, Hospital for Sick Children, Toronto, ON)*

A subset of sporadic medulloblastomas show mutations in members of the *Sonic Hedgehog (Shh)* signaling pathway, and most show overactivity of *Shh* signaling. Humans and mice with germline mutations of Patched, the *Shh* receptor, develop medulloblastomas. We set out to identify human homologues of *Drosphilia melanogaster* genes that are known to inhibit *Shh* signaling.

Using the NCBI Blast server we identified a human expressed sequence tag highly homologous to drosophila *Suppressor of Fused (Su(fu))*. With this partial sequence, we used the Rapid Amplification of cDNA Ends technique to clone *hSu(fu)*. Using cDNA clones to screen the Roswell Park Genomic (Bacterial Artificial Chromosome) Library we identified two BACs containing *hSu(fu)*. Both BACs map to chromosome 10q24.3, a region often deleted in medulloblastoma. By BAC subcloning and sequencing we determined that the genomic structure of *hSu(fu)* has 12 exons. We are in the process of doing a mutational analysis of *hSu(fu)* in a series of sporadic human medulloblastomas. After sequencing half the exons in 30 tumours, we have found one tumour with a truncating mutation that is accompanied by loss of the wild type allele. This suggests that *hSu(fu)* functions as a tumour suppressor gene in a subset of medulloblastomas.

### 9. Molecular immunohistochemical diagnosis of a glioblastoma multiforme (GBM) focus within a pilocytic astrocytoma

*T.G. MAINPRIZE, J.M. BILBAO, L. RONCARI and A. GUHA (University of Toronto, Toronto, ON)*

Astrocytomas are heterogenous tumors, both histologically and molecularly. Molecular markers of GBM were examined immunohistochemically in a specimen where there was progression from a diffuse pilocytic astrocytoma.

The molecular characteristics of a GBM were clearly noted. These included: 1) Higher MIB-1 immunopositivity (10% vs 0%); 2) Increased vascularity, with increased number of Factor VIII positive vessels and increased expression of vascular endothelial growth factor (VEGF) by the tumor cells; 3) Overexpression and activation of Epidermal Growth Factor Receptors (EGFR); 4) Expression of the mutant EGFRVIII by the GBM focus. The two signal transduction pathways implicated in the growth of human GBMs and utilized by aberrant EGFRs, mediated by PI3-Kinase and p21-ras, were also examined. Activation of the PI3-Kinase and p21-ras pathways were inferred by increased activation of their main downstream effectors Akt/PKB and MAPK, respectively. Furthermore, there was loss of PTEN/MMAC1 expression. Expression of p53 and neurofibromin were not altered.

In summary, this is a rare molecular immunohistochemical study undertaken on a single specimen harboring both a GBM

and a pilocytic astrocytoma. It demonstrates our ability to identify molecular changes in tumors using standard immunohistochemical techniques. In the future, such use of receptor and signal transduction specific antibodies should complement histological diagnosis and guide treatments aimed at specific altered pathways in these heterogenous tumors.

**10. A Novel Syndrome of Familial Pediatric Posterior Fossa Tumours in a Family with a Germline Mutation of *hSNF5***

*M.D. TAYLOR, N. GOKGOZ, I. ANDRULIS, T.G. MAINPRIZE and J.T. RUTKA (Department of Laboratory Medicine and Pathobiology, Sonia and Arthur Labatt Brain Tumour Research Centre, University of Toronto, Toronto, ON)*

We present a family in which affected members develop posterior fossa brain tumours as infants, including malignant rhabdoid tumour and choroid plexus carcinoma. The *hSNF5* gene on chromosome 22q11 was recently shown to be deleted or mutated in the majority of sporadic renal and CNS rhabdoid tumours. *hSNF5* is believed to take part in nucleosome remodeling thus controlling the access of transcription factors to promotor regions of DNA. Mutational analysis of the *hSNF5* gene by PCR and sequencing in the family described, demonstrated a single base pair change in the splice site at the 3' end of exon 7. This mutation disrupts the 100% conserved splice site recognition sequence resulting in the exclusion of exon 7 from the mature cDNAas shown by RT-PCR. Exclusion of exon 7 results in a subsequent frame shift and generation of a premature stop codon. This mutation was found in all tumours from this family and in the Germline of all affected and some unaffected family members. Tumours showed loss of the wild type allele by both PER and RT-PER in keeping with a tumour suppressor gene role for *hSNF5*. We propose that Germline mutations of *hSNF5* predispose to a novel autosomal dominant syndrome with incomplete penetrance of familial posterior fossa tumours.

**11. Coincident HIV-polymyositis and AZT myopathy: now you see it, now you don't.**

*R. HAMMOND, K. KAYE and B. DEMAERSCHALK (London Health Sciences Centre and University of Western Ontario, London, ON)*

A 53-year-old male presented with proximal muscle weakness and tenderness of three months duration affecting his legs more severely than his arms. He had been HIV-1 positive for several years and was being treated with AZT, Indinivir, Ritonavir and 3TC. He had no AIDS defining illnesses and his CD4 count was stable (330). Neck flexors were mildly weak as were proximal arm and leg muscles (MRC grade 3 - 4) with loss of bulk and tone. EMG studies revealed myopathic features with increased insertional activity. His CK was normal (66). A muscle biopsy revealed an inflammatory myopathy consistent with HIV-polymyositis, but in addition showed a large number of ragged red fibres with an obvious succinate dehydrogenase/ cytochrome-C oxidase discordance. Ultrastructural examinations corroborated the latter with many fibres bearing excessive mitochondria with atypical morphologies, abnormal cristae and paracrystalline inclusions. The patient remained off AZT and was cautiously treated with corticosteroids resulting in clinical improvement. A repeat biopsy showed resolution of the ragged red fibre pathology. The case is unusual for the coincidence of HIV-polymyositis and AZT myopathy and furthermore for the opportunity to witness a biochemical and ultrastructural reversal of the AZT effect.

**12. PMP22 mutations may affect ER quality control mechanisms through abnormal interactions with calnexin.**

*G.J. SNIPES, J. COLBY, J.J. BERGERON and K. DICKSON (Montreal Neurological Institute and McGill University, Montreal, PQ)*

Mutations affecting Peripheral Myelin Protein-22 (PMP22) are responsible for hereditary human and murine neuropathies, including Charcot-Marie-Tooth disease and the Trembler-J neuropathy. We have shown that disease-causing PMP22 mutations are associated with abnormal retention of the mutant protein in the endoplasmic reticulum of myelinating Schwann cells. To do this, we compared the distribution of epitope tagged PMP22 proteins delivered by adenovirus injected into peripheral nerves using double label confocal microscopy. We then investigated whether any of several candidate ER resident chaperone proteins might be associated with wild-type or mutant PMP22. By immunoprecipitation, we found that PMP22 associates with the molecular chaperone, calnexin, via a lectin-like interaction. Furthermore, pulse-chase studies indicated that mutant PMP22 (Tr-J) has an abnormally prolonged interaction with calnexin suggesting the possibility that mutant PMP22 might sequester calnexin. Indeed, we have demonstrated by immunofluorescence and electron microscopy that mutant PMP22 and calnexin are colocalized in prelysosomal structures resembling myelin figures and, possibly, in aggresomes in cells transfected with mutant PMP22 fused to green fluorescent protein (GFP). Similar structures were identified in homozygous Tr-J sciatic nerves. These findings may explain the more severe phenotype associated with mutant PMP22 proteins as compared to haploinsufficiency of PMP22 as observed in hereditary neuropathy with liability to pressure palsies (HNPP).

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**13. Familial myopathy with tubular aggregates.**

*J.M. BILBAO and S. A. COHEN (St. Michael's Hospital, University of Toronto, ON)*

Tubular aggregates were ubiquitous in a muscle biopsy (Oct.1999) of a 34-year-old mother of a normal 9-month-old boy, with a history of familial neuromuscular disorder affecting her mother (now bedridden), her sister and a niece; two nephews are unaffected. Her muscle weakness was detected in infancy; it was noted that she walked on her toes. She had Achilles tendon release at the age of nine and eye surgery for strabismus at the age of 12 years. She had never walked nor run well. From her teens on she experienced progressive muscle weakness involving lower extremities and eventually the shoulder girdle. The CPK determination is 460. The patient had a muscle biopsy at the age of 11 which was inconclusive. Cryosections of most recent specimen show basophilic granular sarcoplasmic masses that are

bright red with Gomori-trichrome and react intensely for NADH-TR and adenylate deaminase. They are nonreactive with SDH and appear as optically empty on the ATPase preparation. Ultrastructurally, large amounts of tubular aggregates and dilatation of endoplasmic reticulum are present preferentially about myonuclei.

Tubular aggregates were first described in the periodic paralysis and their relation to sarcoplasmic reticulum is well-established. Tubular aggregates are also the salient histologic feature of a rare familial disorder characterized by isolated progressive muscle weakness. Tubular aggregates seem to be a nonspecific change and may represent an adaptive response of the SR to various insults to the muscle fiber.

**14. Inclusion body myositis: the first autopsy study.**

*W.L. STEFANEK[1], B. CURRY[1] and A.K.W. BROWNELL[2] (Departments of Pathology and Laboratory Medicine[1] and Clinical Neurosciences[2], University of Calgary, Foothills Medical Centre, Calgary, AB)*

An autopsy case of inclusion body myositis (IBM) has not yet been reported in the literature. IBM is a late-onset progressive inflammatory myopathy characterized by an endomysial inflammatory infiltrate, muscle fibre degeneration/regeneration, rimmed vacuoles, and poor treatment response. We describe the first autopsy case of IBM. The patient was a 90-year-old female with a 12 year history of progressive weakness whose terminal admission was for bilateral pneumonia and septic shock. The diagnosis of IBM was made at 81 years of age from a vastus lateralis biopsy. Until the time of death, the patient was ambulating with assistance and over the years presented to the hospital on numerous occasions with recurrent aspiration pneumonia and swallowing difficulties. At autopsy, skeletal muscle was sampled from the following groups: sternocleido-mastoid, deltoid, esophagus, throat, diaphragm, intercostal, psoas, and anterior tibialis. Histology revealed that the following muscles exhibited the typical features of IBM: quadriceps, sternocleidomastoids, esophagus, diaphragm, and throat. The presence of disease in the quadriceps, esophagus and throat muscles correlates clinically with the patient history of leg weakness, swallowing difficulties and aspiration pneumonia, and with the gross findings at autopsy of almost complete absence of these muscle groups. This is a unique case in that it is the first documented autopsy examination of a patient with IBM.

**15. Nimodipine reduces behavioral deficits and white matter degeneration in young rats with hydrocephalus.**

*M. R. DEL BIGIO and E. M. MASSICOTTE (Department of Pathology, University of Manitoba, Winnipeg, MB)*

Hydrocephalus causes damage to periventricular white matter at least in part through chronic ischemia. Hydrocephalus was induced in 3-week-old rats by injection of kaolin into the cisterna magna. We first demonstrated that calcium ion accumulates in the periventricular white matter, and that calpains are increased in quantity and activated. We tested the hypothesis that treatment with nimodipine, an L-type calcium channel blocking agent with demonstrated efficacy in a range of cerebral ischemic disorders, would ameliorate the effects of experimental hydrocephalus. Rats were treated by continuous administration of nimodipine for two weeks, beginning two weeks after induction of hydrocephalus. Rats underwent repeated tests of motor and cognitive behavior. At the end of the treatment period the brains were analyzed by histopathological and biochemical means. Nimodipine treatment prevented the declines in motor and cognitive behavior that were observed in untreated rats. Ventricular enlargement, determined by magnetic resonance imaging, was reduced and the corpus callosum was thicker. The sodium channel blocking agents mexiletine and riluzole were not protective. The protective effect of nimodipine is most likely on the basis of improved blood flow, although prevention of calcium influx-mediated proteolytic processes in axons cannot be excluded. Adjunctive pharmacologic therapy might be of benefit to patients with hydrocephalus.

[Funded by the Manitoba Health Research Council and Medical Research Council.]

# Titles of Diagnostic Case Presentations

**1. Acquired (nonWilsonian) Hepatocerebral Degeneration**

*W. HALLIDAY (Division of Neuropathology, Toronto Western Hospital - UHN, Toronto)*

**2. Cerebrotendinous xanthomatosis**

*J.B. LAMARCHE and B. LEMIEUX (Department of Pathology and Pediatrics, Centre Hospitalier Universitaire, Sherbrooke, Quebec)*

**3. Adult polyglucosan body disease**

*A.H. KOEPPEN and A. LOSSOS (VA Medical Center, Albany, N.Y. 12208 USA and Hadassah University Hospital, Jerusalem 91120 Israel)*

**4. White matter degeneration in galactosemia**

*D. P. AGAMANOLIS and M.S. PLATT (Children's Hospital Medical Center of Akron and Summit County Medical Examiner's Office, Akron, Ohio)*

**5. Fowler's syndrome**

*C.E. HAWKINS and L.E. BECKER (Department of Laboratory Medicine and Pathobiology, University of Toronto and The Hospital for Sick Children, Toronto)*

**6. Dementia of motor neuron disease type; with intranuclear inclusions**

*J.M. WOULFE (Hamilton Health Sciences Corporation and McMaster University, Hamilton, ON)*

**7. Oligodendroglioma (WHO Grade II)**

*B. CURRY, R. AUER, A. PINTO and W. STEFANEK. (Department of Pathology & Laboratory Medicine, University of Calgary, Foothills Medical Centre and Alberta Children's Hospital)*

**8. Desmoplastic neurotropic melanoma**

*J.M. BILBAO and A. SEIVWRIGHT (Department of Pathology, St. Michael's Hospital, Toronto, ON)*

**9. Metastatic carcinoma in a meningioma**

*C. HAWKINS, M.M. FELDKAMP*, C. TATOR*, S. NAG (Divisions of Neuropathology and *Neurosurgery, Toronto Western Hospital at The University Health Network and University of Toronto)*

**10. Chordoma with malignant spindle cell components**

*B. LACH, (Department of Pathology, King Faisal Specialist Hospital & Research Centre, Riyadh, Saudi Arabia)*

**11. Prior transsphenoidal resection of pituitary adenoma with iatrogenic plugging of surgical defect with adipose tissue and cartilage**

*R. PARKER, F. DURITY and I.R.A. MACKENZIE (Department of Pathology and Division of Neurosurgery, Vancouver General Hospital and University of British Columbia).*

**12. Churg-Strauss syndrome with necrotizing vasculitis and interstitial myositis, post-antileucotriene receptor therapy, in a patient not previously treated with steroids**

*Y. ROBITAILLE AND F. BERTHELET (Dept. of Pathology & Cell Biology, University of Montreal, Centre Hospitalier Universitaire de l'Université de Montréal and Ste-Justine Hospital Montreal)*

**13. Subacute sclerosing panencephalitis**

*J.P. ROSSITER (Department of Pathology, Queen's University and Kingston General Hospital, Kingston, ON)*



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