PET images were co-registered and SUVRs were generated for several brain regions. Using Aβ immunohistochemistry (10D5, Eli Lilly), the burden of Aβ plaques was quantified in 16 regions of interest using an area fraction fractionator probe (Stereo Investigator, MicroBrightfield, VT). There were regional variations in Aβ plaque burden with highest densities observed in the neocortical areas and the striatum. On spearman correlations, *in vivo* PiB-PET correlated with postmortem Aβ plaque burden in both LOAD and ADAD, with strongest correlations seen in neocortical areas. In summary, [C11]PiB-PET has utility as a biomarker in both ADAD and LOAD.

LEARNING OBJECTIVES

This presentation will enable the learner to:

1. Discuss how PET-PiB beta-amyloid imaging is used as a potential biomarker of Alzheimer disease (AD)
2. Correlate postmortem neuropathologic evidence of beta-amyloidosis with PET-PiB data, and learn that PET-PiB is a potentially useful tool to detect beta-amyloidosis in presymptomatic and symptomatic individuals

Abstract 4

Microvascular pathology of Friedreich cardiomyopathy

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Nikolaus Friedreich (1877) was aware of heart disease in his patients but thought it was unrelated to the neurological disorder. In 1946, Dorothy Russell considered cardiomyopathy an integral part of Friedreich ataxia (FA). In addition to sparse inflammatory infiltration, sections show fibrosis and capillary hyperplasia. We examined the left ventricular walls of 41 homozygous FA patients aged 10–87 and 21 controls aged 2–69. An antibody to CD34 enabled quantitative capillary profile counts for a comparison with cardiomyocyte counts in the same field. Mean capillary counts in normals were $1926 \pm 341/mm^2$, and the median ratio of capillaries to cardiomyocytes was 1.0 (interquartile range [IQR]: 0.9-1.2). In FA, however, the number of cardiomyocytes/mm^2 was less, and the median ratio of capillaries to heart fibers was 2.0 (IQR 1.4-2.4). There was a significant correlation of the higher guanine-adenine-adenine trinucleotide length (shorter allele, GAA1) with the younger age of onset, shorter disease duration, and lower cardiomyocyte counts. The ratio of capillaries to heart fibers was higher in patients with long GAA1 repeat expansions (e.g., 3.31 in GAA1 of 1200). Double-label immunofluorescence for CD34 and S100A4 revealed co-expression in endothelial cells, supporting endothelial-to-mesenchymal transition in the pathogenesis of cardiac fibrosis (supported by Friedreich's Ataxia Research Alliance).

LEARNING OBJECTIVES

The presentation will enable the learner to:

1. Describe endothelial-to-mesenchymal transition in the pathogenesis of cardiac fibrosis in Friedreich cardiomyopathy

Session 2: Pediatric, Epilepsy and Miscellaneous Neuropathology

Abstract 5

Parasagittal intraparenchymal hemorrhage in complicated second stage labour: a report of three cases

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The increased use and mastery of ceasarian section for deliveries and the refinement of technologies for assisted delivery in the setting of dfficult second stage of labour have made intrapartum deaths more rare and modern obstetrical pracices are rarely accompanied by the classic forceps related intracranial injuries. We document a novel pattern of intracranial injury in three cases of neonatal death following prolonged labor, of which two out of three required vacuum and forceps.

All three showed similar bilateral parasagittal intraparenchymal haemorrhages and cerebral edema, in a pattern reminiscent of "gliding contusion, as well as subgaleal haemorrhage of varying amout. Two out of the three cases showed parietal bone fractures and one demonstrated extensive craniolcuniae. We briefly discuss the significance of these findings and implications for future cases.

LEARNING OBJECTIVES

This presentation will enable the learner to:

1. Explore the current theories leading to neonatal death in prolonged labor
2. Summarize the known pathological findings associated with vacuum and forceps
3. Discuss the significance of intraparenchymal hematoma in the setting of prolonged delivery

Abstract 6

Non-Perfused Brain and Retino-Dural Hemorrhage

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10 cases of pneumonia causing cardiac arrest and non-perfused brain occurred at ages 40 days-30 months, in a medico-legal setting. In each deceased child, both the pneumonia and non-

perfused brain were verified histologically. Upper respiratory infection and mouth-breathing accompanied the pneumonia, with ongoing choking on formula or food in three cases, and vomiting in an additional five cases. In eight of the 10 cases, the pre-terminal event was a quiet respiratory arrest while sleeping, or being carried in the arms. Adrenaline was given up to 7 times during CPR lasting 44±32 minutes, with up to 2 hours CPR and fall in body temp to <32°C. Mean survival was 1.9±1.5 days and heparin was given for organ donation in 3 cases. The lungs showed chronic interstitial pneumonia as described by Katzenstein, with superadded acute bronchiolo-alveolar infiltrates in two cases of aspiration. The court permitted recuts and cellular characterization of the interstitial cells in one case, revealing the infiltrate was ~40% histiocytes, 5% T or B cells, and ~50% vimentin+ mesenchymal cells. All brains showed features of non-perfused brain and retino-dural hemorrhage. The observed features of non-perfused brain were blurring of the gray-white junction, edema, gross friability, histologic pallor, closure of the microcirculation, patchy acidophilic neurons and recent demarcated pan-necrosis, and pituitary infarction in one patient where hypophysis was sampled. Normally, from birth to 30 months, cerebral blood flow increases to 55% of cardiac output, accompanying physical brain growth. Restoration of high cardiac output using adrenaline-CPR means that on resuscitation, re-routing of blood that can no longer go through the non-perfused brain detours through dura, face, scalp, eyes and optic nerve sheaths. The diversion of blood around non-perfused brain results in facial bruising and retino-dural hemorrhage that can be misinterpreted as head trauma, and a common inference of child abuse in the courts. In the present series from Australia, Canada and the USA, outcomes ranged from acquittal to life imprisonment.

References

Katzenstein AL, et al. (1995) Chronic pneumonitis of infancy. A unique form of interstitial lung disease occurring in early childhood. Am *J Surg Pathol* **19**:439–447

LEARNING OBJECTIVES

This presentation will enable the learner to:

1. Investigate infant deaths including workup for interstitial pneumonia.
2. Know cerebral blood flow changes in development, and cranial blood flow dynamics in non-perfused brain.

Abstract 7

Epilepsy Related Death: the London Health Sciences Center Experience

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Premature mortality among epilepsy patients is well recognized. Except a few identifiable causes of unnatural death, more than half of the epilepsy related death remains unexplained after extensive workup. These cases are classified as sudden unexpected death in epilepsy (SUDEP). SUDEP incidence varies significantly depending on the population, the methods documenting cause of death and the availability of Neuropathological examination. An accurate diagnosis of the cause of death is needed for epilepsy related death. The goal of this study is to present the relevant clinical data, the general autopsy and Neuropathology findings of epilepsy related death investigated in London Health Sciences Center during the period of 2000 to 2011. We identified 71 cases with known history of chronic epilepsy. In the 29 cases of epilepsy associated death, the causes of death have been classified as cardiac, pulmonary, accidental (e.g. drowning), toxic (e.g. drug overdose) and non-related causes. Forty two cases are considered to be SUDEP, and were categorized according to the recently proposed SUDEP Definition and Classification. Half of the SUDEP cases have no specific Neuropathological findings. The most common identifiable lesions in SUDEP cases are perinatal/neonatal destructive lesions (29%), hippocampal sclerosis (24%), and focal cortical dysplasia (20%). These are followed by neuronal heterotopia (9%), previous head trauma (9%), and cavernoma (5%).

LEARNING OBJECTIVES

This presentation will enable the learner to:

1. Review cause of death in epilepsy related deaths
2. Discuss the practice guideline in neuropathology autopsy of epilepsy related deaths

Abstract 8

Medial Temporal Lobe Dysgenesis and More in a Man with Hypochondroplasia and Epilepsy

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Hypochondroplasia, achondroplasia, and thanatophoric dysplasia are related at the molecular level, all caused by fibroblast growth factor receptor 3 (FGFR3) gene mutations. They differ in severity. FGFR3 has critical roles in fibroblast growth factor (FGF) signalling pathways during bone growth and cerebral cortical development. Mutations of the FGFR3 gene lead to constitutive activation of FGFR3. The well-described brain malformation in thanatophoric dysplasia is characterized by gross abnormalities of temporal lobe patterning and severe dysplasia of the hippocampus. Experimental models suggest that increased proliferation, abnormal migration, and decreased apoptosis are involved. However, reports of the brain findings in hypochondroplasia are based solely on radiologic imaging.

We present the neuropathology of a 44 year-old man with hypochondroplasia, epilepsy, and significant intellectual disability. The temporal lobes are enlarged, prominent fissures traverse the inferior temporal surface, and the hippocampus is abnormally folded. Microscopically, the dentate gyrus is variably small or