## LEARNING OBJECTIVES

This presentation will enable the learner to:
Recognize key anatomic divisions of the amygdala

1. Describe how different neurodegenerative diseases affect the amygdala
2. Consider how anatomic specificity of protein aggregation is important in the classification of neurodegenerative diseases

### Abstract 13

### The tissue proteome of dorsal root ganglia in Friedreich ataxia

*AH Koeppen[1], AM Travis[1], J Qian[2], JE Mazurkiewicz[2], BB Gelman[3], S Pelech[4], C Sutter[4]*

[1]VA Medical Center, Albany, NY, USA; [2]Albany Medical College; [3]University of Texas Medical Branch, Galveston, TX, USA; [4]Kinexus Bioinformatics Corp., Vancouver, BC, Canada and University of British Columbia

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Dorsal root ganglia (DRG) at all levels of the spinal cord are a prominent target of Friedreich ataxia (FA). The lesions include hypoplasia of neurons, proliferation of satellite cells, infiltration by IBA- 1-reactive monocytes, and formation of residual nodules. Paucity and smallness of DRG neurons account for the lack of large myelinated axons in dorsal roots and sensory peripheral nerves. The lack of myelin in dorsal roots can be attributed to low levels of neuregulin 1 type III (NRG1[III]). Lysates of 13 DRG of genetically confirmed FA patients were analyzed by antibody microarray with 878 different validated antibodies that target structural and signaling proteins, and by Western blots. KIT and mTOR, two proteins involved in cellular proliferation, were significantly upregulated in the DRG of FA. KIT is a transmembrane receptor that dimerizes when it binds two molecules of stem cell factor (SCF) in its extracellular domain and becomes activated as protein tyrosine kinase. Immunohistochemistry with anti-KIT generated reaction product in satellite cells of normal DRG and prominent labeling of these cells in FA that co-localized with SCF on double- label immunofluorescence; SCF was present in S100-positive satellite cells rather than monocytes. Immunohistochemical reaction product of mTOR and other mTOR complex proteins, such as hamartin (TSC1), tuberin (TSC2), raptor (mTOR complex 1) and rictor (mTOR complex 2) was also present in satellite cells of normal DRG and DRG of FA. Antibodies to two downstream proteins that are considered to be indicators of mTOR activity, P70 S6K and 4E-binding protein 1, revealed no reaction product in DRG of FA. TSC1, TSC2, and mTOR are best known from their roles in tuberous sclerosis, but expression of these proteins, and KIT, in DRG may contribute to signaling cascades underlying the proliferation of satellite cells in FA.

## LEARNING OBJECTIVES

This presentation will enable the learner to:

1. Discuss cellular proliferation in the pathogenesis of the DRG lesion in Friedreich ataxia

## CONFLICT OF INTEREST

AHK is a consultant to PTC Therapeutics of South Plainfield, NJ USA. SP and CS are majority owners of Kinexus.

### Session 5: Neuropathology practice

### Abstract 14

### Canadian Association of Neuropathologists Workforce Survey, 2019

*P Shannon*

Mount Sinai Hospital, Toronto

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To characterize the professional occupation of Canadian neuropathologists and estimate the future employment demands in neuropathologists, all the active members of the Canadian Association of Neuropathologists in Canada (n=53) were surveyed by E-mail, inquiring as to their estimated date of retirement, their current employment and practice profile, and as to any practice trends they had noticed. 49 members replied: all but one practice at medical school centers. 38 practice exclusively in neuropathology and three of these are employed at less than 75% of a full time equivalent. The remaining practices are mixed neuropathology and anatomical pathology, and one practices exclusively ophthalmic pathology. 35% reported significant neuropathology sub specialization (e.g. forensic, pediatric, neuromuscular). 42% reported greater than 10% of time dedicated to research (of these, median 30%) and 35% greater than 10% time spent in teaching, and 9% greater than 10% time in administration. Of the 49 surveyed, as of the spring of 2019, 14%(seven) of the full time neuropathologists can be expected to retire in the next 10 years, and 6% (three) with mixed AP/NP practices.

## LEARNING OBJECTIVES

This presentation will enable the learner to:

1. Understand the current spectrum of practice of Neuropathologists across Canada
2. Describe the patterns of employment and anticipated retirements of Canadian Neuropathologists