Curriculum vitae Laurence Albert BINDOFF 24.05.2023

Current Positions:
Professor Emeritus, Dept. Clinical Medicine (K1), University of Bergen, Norway.

Education:

1991	**MD/PhD (Newcastle)** (With commendation)
1985	**M.R.C.P. (UK)**
1980	**MB, BS** Degree in Medicine
1975	**M.Sc. 1974 B.Sc. Biochemistry/Genetics (2/I)**

Main research focus
I was an academic neurologist and basic scientist primarily interested in mitochondrial disease and the mechanisms involved in tissue specific manifestations. I identified a novel disease mechanism involving abnormal RNA processing related to mtDNA mutation m.3302A>G and several new mtDNA mutations (m.3460G>A, one of the 3 commonest LHON mutations; m.3302A>G; m.12320A>G; m.13271T>C). Studies of the tissue restricted manifestations of the m.12320A>G including a novel treatment using focussed muscle degeneration (*Nat Genet* 1997 16:222-4). I have described 2 new diseases, one involving endo-cannabinoid metabolism (ABHD12) and a new defect linking a mitochondrial protease with amyloid metabolism (PITRM1). My major focus is now polymerase gamma related disease. We are using iPSC with POLG mutations differentiated into neurones, and astrocytes to investigate disease mechanisms.

Fellowships and awards

2015	Monrad-Krohn Prize for outstanding research in neurology
2011	Joint Award of Helse Vest prize for best research milieu
2011	Top graded – Excellent by external reviewer in National Research Council review
2011	Visiting Professor - Institute of Human Genetics, Newcastle University, UK.

Five relevant peer-reviewed publications of last 5 years
1. Liang KX, Vatne GH, Kristiansen CK, Levglevskyi O, Kondratskaya E, Glover JC, Chen A, Sullivan GJ, **Bindoff LA**. N-acetylcysteine amide ameliorates mitochondrial dysfunction and reduces oxidative stress in hiPSC-derived dopaminergic neurons with POLG mutation. *Exp Neurol*. 2020 Nov 29;337:113536.
2. Liang KX, Kristiansen CK, Mostafavi S, Vatne GH, Zantingh GA, Kianian A, Tzoulis C, Høyland LE, Ziegler M, Perez RM, Furriol J, Zhang Z, Balafkan N, Hong Y, Siller R, Sullivan GJ, **Bindoff LA**. Disease-specific phenotypes in iPSC-derived neural stem cells with POLG mutations. *EMBO Mol Med* 2020 Oct 7;12(10):e12146.
3. Brunetti D, Torsvik J, Dallabona C, Teixeira P, Sztromwasser P, Fernandez-Vizarra E, Cerutti R, Preziuso C, D'Amati G, Baruffini E, Goffrini P, Viscomi C, Ferrero I, Boman H, Telstad W, Johansson S, Glaser E, Knappskog PM, Zeviani M, **Bindoff LA**. Defective mitochondrial PITRM1 causes progressive neurodegeneration associated with APP and Aβ accumulation. *Embo Mol Med,* 2015 Dec 23;8(3):176-90.
4. Balafkan N, Mostafavi S, Schubert M, Siller R, Liang KX, Sullivan G, **Bindoff LA**. A method for differentiating human induced pluripotent stem cells toward functional cardiomyocytes in 96-well microplates. *Sci Reports*, 2020 28;10(1):18498.
5. Tzoulis C, Tran GT, Coxhead J, Bertelsen B, Lilleng P, Balafkan N, Payne B, Miletic H, Chinnery PF, **Bindoff LA**. The molecular pathogenesis of POLG-related neurodegeneration. *Ann Neurol*, 2014, Jul;76(1):66-81

Signed: Bergen, 24.05.2023

Laurence Bindoff