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## Mass spectrometry-based proteomic profiling of muscular dystrophy

**Kay Ohlendieck**

Maynooth University, Ireland

Mass spectrometry-based proteomics is a key bio-analytical technique for the comparative analysis of pathological specimens. In the field of neuromuscular disorders, both two-dimensional gel electrophoresis and liquid chromatography have been employed for the large-scale separation of distinct protein populations prior to mass spectrometric analysis. Our laboratory has focused on the systematic profiling of animal models of Duchenne muscular dystrophy, a devastating muscle wasting disease of early childhood. X-linked muscular dystrophy is due to primary abnormalities in the *Dmd* gene that encodes the membrane cytoskeletal protein dystrophin. For the mass spectrometric identification of new biomarker candidates of dystrophinopathy, we used two complementary methods, fluorescence two-dimensional difference in-gel electrophoresis and liquid chromatography in combination with label-free mass spectrometry. Novel skeletal muscle-associated disease markers of fiber degeneration, myofibrosis and sterile inflammation are involved in the excitation-contraction-relaxation cycle, the extracellular matrix, the cytoskeleton, energy metabolism and cellular stress. In addition, tissue samples from the dystrophic heart and the central nervous system, as well as serum, were analysed by proteomics. Independent verification studies were carried out by immuno-blotting and immunofluorescence microscopy. In the future, the newly established proteomic biomarker candidates of X-linked muscular dystrophy may be useful for improving diagnostic, prognostic and therapy-monitoring approaches, as well as the identification of new therapeutic targets down-stream of the primary abnormalities in the cytoskeletal network.

### Biography

Kay Ohlendieck completed his Under-graduate Degree in Biology at University of Konstanz, Germany (1985), a PhD in Biochemistry at University College Cork, Ireland (1989) and a DSc in Muscle Biology at University College Dublin, Ireland (2011). He has worked as a Post-doctoral Associate at University of Iowa, Iowa City and at State University of New York, Stony Brook, as well as a Lecturer in Department of Pharmacology at University College Dublin (1995-2001). Since 2002, he is a Professor and Chair of Biology at Maynooth University-National University of Ireland. His research focuses on Skeletal Muscle Proteomics.

kay.ohlendieck@nuim.ie

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