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# Seminars in Genetics and Molecular Cell Biology


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## Modified Atkins diet in late-onset mitochondrial myopathy


Mitochondrial disorders are the most common group of inherited metabolic disorders. These disorders are very heterogeneous disease group that can vary from mild late-onset myopathy to fatal infantile multi-organ failure. Progressive external ophthalmoplegia (PEO) is an adult-onset mitochondrial myopathy characterized by the accumulation of multiple mitochondrial DNA deletions causing mitochondrial respiratory chain deficiency. No effective treatment exists for late-onset mitochondrial myopathies, which is the case for most mitochondrial diseases. We have previously created a late-onset mitochondrial myopathy mouse model, the Deletor mouse, replicating well the tissue manifestations of PEO-patients. We showed that in this mouse, ketogenic high-fat diet delayed the progression of mitochondrial myopathy. This diet also improved the metabolic and lipidomic changes of these mice caused by the disease. We have recently conducted a small pilot study in PEO-patient and control subjects, who followed modified Atkins diet for 1-4 weeks. We demonstrate a rapid response in PEO patients, with versatile metabolic consequences specific to mitochondrial disease. Our results indicate that the constitution of diet is crucial for patients with mitochondrial dysfunction and may have a role in disease progression.


## Thursday, May 2, 2013 at 11.00 a.m.

Institute for Genetics,
Zülpicher Str. 47 a, Lecture hall, 4th floor

Host: Thomas Langer, Institute for Genetics,
University of Cologne

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