of ameliorating EAE. In this study, we further analyzed the effect of FSD-C10 on oligodendrocyte formation.
Material and methods: Female adult C57BL/6 mice were immunized with MOG_{35-55} to induce chronic EAE. The study was approved by the Ethics Committee of Shanxi Datong University. Mice received 10 μl/ nostril (5 μg/μl) FSD-C10 at the entrance of the nostrils on day 3 until day 27 p.i. Mice that received the same volume of ddH_2O nasally served as EAE controls.
Results: FSD-C10 inhibited proliferation of CD4, CD68 and $CD11b^+$ immune cells, caused less immune cells in brain. In contrast, FSD-C10 stimulated NG2-expressing oligodendrocyte precursor cell and GalC-expressing oligodendrocyte proliferation, causing more oligodendrocyte formation. In addition, FSD-C10 also stimulated the production of neurotrophic factors NT-3 and GDNF, shifted activated microglia from M1 to M2 phenotype and inhibited inflammatory responses in the brain.
Conclusions: These results indicate that FSD-C10 promotes oligodendrogenesis and remyelination by elevating growth factors and strengthening of the microglial neuroprotective phenotype (M2) conducive for repair. (Grant: The Department of Science and Technology, Shanxi Province of China, 2013081058; Research Project Supported by Shanxi Scholarship Council of China, 2014-7).
Keywords: oligodendrogenesis, neurotrophic factors, inflammatory responses, NG2, GalC.

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Mixed Topics 2
Establishing a human neuronal derived-iPSC model to clarify the pathogenetic mechanism for PKAN

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Mutations in the *PANK2* gene, which encodes pantothenate kinase 2, underlie an autosomal recessive inborn error of coenzyme A metabolism, called pantothenate kinase-associated neurodegeneration (PKAN). PKAN is characterized by dystonia, dysarthria, rigidity, retinal degeneration and severe iron accumulation in the brain. The pathogenesis mechanism of this disorder remains largely unknown. Current models recapitulate only a subset of the pathological manifestations and lack any neurological phenotype in particular the iron deposition in the brain. Considering all of this, there is the urgency of establishing meaningful experimental models to determine the pathological events leading to the disease. Toward this aim, we have employed iPS cell technology for generating an *in vitro* disease model. We have succeeded in generating derived-iPSC neurons from patients and relative healthy donors by using the Sendai reprogramming vectors. Cells were analyzed for mitochondrial functionality, oxidative status and iron metabolism. The derived-iPSC neurons display a reduced antioxidant defense, increased level of ROS development and abnormal electrophysiological properties respect to the control. Furthermore, PKAN derived neurons present electron dense aggregates inside the mitochondria of a still unknown nature. In conclusion, we succeeded in obtaining a human neuronal model to study PKAN disorder and the preliminary results indicated that PKAN derived neurons present altered oxidative status, abnormal mitochondrial functionality and morphology. The financial support of Telethon (Grant n°: GGP11088) and AISNAF is gratefully acknowledged.

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Mixed Topics 2
Investigation of regulatory factors in Lipid Storage Myopathies (LSM) with triglyceride accumulation

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Background: Triglycerides are massively stored in LSM, i.e. carnitine deficiency, RR-MADD and NLSD-M (ATGL deficiency), conversely, in CPT-II deficiency lipid droplets are almost absent.
Objective: We aimed to analyse factors that regulate degradation and *PPAR-gamma* pathways. The Transcription Factor-EB (TFEB), a master regulator of lysosomal biogenesis and autophagy, is induced by starvation through an autoregulatory feedback loop and exerts a global transcriptional control on lipid catabolism via *PGC1α* and *PPARα*.
Patients and methods: We selected 6 patients with LSM: 2 RR-MADD, 1 carnitine deficiency, 2 NLSD-M, 1 CPT-II deficiency. Muscle immunofluorescence for TFEB and p62 (marker of protein aggregates) and fetal myosin (marker of regeneration) and immunoblot using p62, LC3 antibodies was done.
Results: While in 2 NLSD-M patients there was a co-localized overexpression of p62 and TFEB in some atrophic fibers, some of which were regenerating, in Carnitine and CPT-II deficiency their reaction appeared normal. In regenerating fibers TFEB localized in the cytoplasm (inactive form), whereas in atrophic fibers it localized in the nuclei (active form). Vacuolated and atrophic fibers did not display p62-positive protein aggregates, indicating, together with the LC3-II and p62 immunoblot analysis, that the autophagic flux is still preserved. Furthermore, we studied plasma myo-microRNA of 3 relatives of another NLSD-M family to observe their regulatory role in muscle. Myomicro-RNAs were inversely correlated to residual muscle mass in this NLSD-M family. Mitochondrial enzymes in two subsequent muscle biopsies of the index patient were decreased revealing an essential role of ATGL in mitochondriogenesis.
Conclusion: Nutrition and autophagy are important in RR-MADD and NLSD-M, when there is a progressive wasting of muscle.

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Genetic profiling of Indian patients with glutaric acidemia type I

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Background: Glutaric acidemia type I (GA-I), is an inborn error of metabolism caused due to deficiency of the enzyme Glutaryl-CoA