in vitro and by RT-PCR. MSC's migration potential was measured in the scracth assay. MSCs' interaction with Schwann cells and their effect on tumorigenesis was examined in co-culture by apoptosis markers on Flow Cytometry.

Results: NF1-MSCs' adipogenic and osteogenic differentiation potential was lower than healthy controls as assessed by staining Aizerin Red S and Oil Red O and RT-PCR for osteopontin and collagen1. MSCs cultured from dermal neurofibroma showed faster closing of the scratch compared to the same patient's normal and café au lait skin. On the other hand, MSCs obtained from plexiform neurofibroma healed late, while MSCs derived from the same patient's café au lait skin showed the fastest healing. Schwann cell-MSC co-cultures showed no specific effect of MSC on Schwann cells' Annexin V and Propidium iodide expression, neither any effect of Schwann cells on the MSC surface markers was observed.

Conclusion: These results support particular behavior of MSC form NF1 patients in terms of differentiation and cell motility in vitro that might have implicaitons on clinical behavior of their tissues.

P-09.02.2-024
Assay of the key enzymes of glutamine-methionine bicycle activity of the tissues in the animals with induced hepatic encephalopathy

B. Krasnikov, N. Gessler, **O. Klein**, A. Antipov, E. Isakova, Y. Deryabina
Bach Institute of Biochemistry RAS, Moscow, Russia

Hepatic encephalopathy with ammonium ions accumulation is accompanied by some disorder in the brain due to toxic material concentration being usually detoxified in the liver. One of the reasons for hyperammoniemia could be some imbalance in brain glutamine metabolisation induced by the key enzymes glutamine transferases (GTs), which catalyze the reaction of glutamine transamination resulting in neurotoxic product of α-ketoglutaramate (αKGM). αKGM is hydrolyzed to α-ketoglutarate and ammonia by ω-amidases.

In the study, the dynamics of the enzymes activity in the tissues and biological liquids of experimental animals with hepatic dysfunction induced by thioacetamide (TAA) was under investigation. White laboratory rats of Wistar line (female, weight of 140 g) chronically intoxicated with hepatotrophic toxine of TAA. Every 2 weeks, some biological samples were collected to assess GT-K and ω-amidase activities. ω-Amidase activity was the highest in the kidney tissue in the control and decreased by 70% in the experimental group. In the experimental hepatic ω-amidase activity decreased by 240% compared to those in the control. The average ω-amidase activity in the blood serum (0.015 nmols/mg/min) and in the brain (0.005 nmols/mg/min) differed faintly. Maximal GT-K activities were revealed in the kidneys; in the controls, it was about 250% higher than those in the experimental animals. The difference between average enzyme activities in the liver of the control and experimental animals reached 350%. The average GT-K activities in the blood serum and brain of the control and experimental animals were rather similar.

The decrease in ω-amidase and GT-K activities obtained in the study during hepatic pathology development could testify to imbalance of glutamine metabolism, possibly aimed at declining the level of αKGM neurotoxicant under the hepatic dysfunction.

Acknowledgments: Supported by the Russian Federation Ministry of Science and Education (Grant No RFMEFI60414X0116).

P-09.02.2-025
Functional characterization of clinically relevant novel mutations in ATP7B gene using the *Saccharomyces cerevisiae* model

O. Simsek Papur[1], O. Terzioglu[1], A. Koc[2]
[1]*Dokuz Eylul University, Izmir, Turkey,* [2]*Izmir Institute of Technology, Izmir, Turkey*

Wilson disease is an autosomal recessive disorder of copper metabolism characterized as neurodegeneration and liver abnormalities. It is caused by defects in the ATP7B gene.

ATP7B is responsible for the sequestration of Cu into secretory vesicles, and this function is exhibited by the orthologous Ccc2p in the yeast. We aimed to characterize clinically-relevant novel mutations of p.T788I, p.V1036I and p.R1038G-fsX8 in yeast lacking the CCC2 gene.

The patients with these mutations have copper storage abnormalities in different parts of their bodies; p.T788I mutation mainly affects the liver and the nervous system, p.V1036I mutation affects the nervous system, and p.R1038G-fsX83 mutation causes damages to the liver. To better understand the effects of these mutations on normal functions of ATP7B, we cloned human ATP7B gene onto a yeast expression vector and created the same mutations by site directed mutagenesis. Then, wild type and mutated forms of ATP7B genes were transformed into yeast cells lacking the homologous CCC2 gene for functional comparison.

First, we analyzed the expression of ATP7B and its variants in yeast cells by a real time PCR approach and Western Blot to make sure that transformed cells express the plasmids.

Expression of human wild type ATP7B gene in ccc2Δ mutant yeast restored the growth deficiency and copper transport activity, however, expression of the mutant forms did not restore the copper transport functions and only partially supported the cell growth.

Our data support that p.T788I, p.V1036I and p.R1038G-fsX83 mutations cause functional deficiency in ATP7B functions and suggest that these residues are important for normal ATP7B function.

Monday 5 September 12:30–14:30

Education, training, and career planning in molecular life sciences

P-EDU-001
An all solid-state urea biosensor based on ammonium-selective PVC membrane electrode

G. Vardar[1], M. Altikatoglu[1], I. Isildak[2]
[1]*Department of Chemistry, Arts and Science Faculty, Yildiz Technical University, Istanbul, Turkey,* [2]*Bioengineering Department, Chemistry and Metallurgy Faculty, Yildiz Technical University, Istanbul, Turkey*

In recent years, attempts were made to develop miniaturized potentiometric biosensors which is particularly important to reduce the amount of enzyme and reagents needed. The miniaturization of a biosensor is possible by using an all solid-state polymer membrane ion selective electrode which is cheap, easy to prepare and allow micro-sized construction. The use of all solid-state polymer membrane ion selective electrode as the basic sensing element also has the advantage of providing biosensors that are easy to fabricate, exhibit rapid response and have long life-

The FEBS Journal **283** (Suppl. 1) (2016) 129–417 DOI: 10.1111/febs.13808