

BO118

### PATIENTS WITH NON-V30MET TRANSTHYRETIN MUTATIONS: DATA FROM THE FAP WORLD TRANSPLANT REGISTRY (FAPWTR) SHOW LARGE VARIATION IN OUTCOME FOLLOWING LIVER OR LIVER/HEART TRANSPLANTATION

Ole Suhr<sup>1</sup>, Marie Larsson<sup>2</sup>, Bo-Göran Ericzon<sup>2</sup>, Henryk Wilczek<sup>2</sup>

<sup>1</sup>Department of Public Health and Clinical Medicine, Umeå University Hospital;

<sup>2</sup>Division of Transplantation Surgery, Karolinska Institutet, CLINTEC, Karolinska University Hospital Huddinge

**Background:** Hereditary transthyretin amyloidosis (ATTR) has been treated by liver transplantation (LTx) for over two decades. Different centres have reported the outcome of fairly large series of LTx ATTR Val30M patients, but for other mutations only few reports comprising small number of patients have been presented. We present the outcome after transplantation of non-ATTR Val30Met amyloid patients based on data reported to the FAP world transplant registry (FAPWTR).

**Methods:** Outcome data for all non-ATTR Val30Met patients registered in the FAPWTR was studied. The Kaplan-Meier method and log-rank test was used to analyse survival rates.

**Results:** The registry holds a total of 264 patients (males 174, females 90) with non-ATTR Val30Met mutations, representing 57 different mutations. The 9 most common mutations showed a considerable 10-year survival variation, ranging from 23% for Ser50Arg to 85% for Val71Ala. All mutations, except the Tyr114Cys mutation, with leptomeningeal complications revealed poor survival.

**Conclusions:** The FAPWTR data revealed large survival variations not only between different mutations, but also between mutations with similar phenotypes. Some mutations, such as Leu111Met, Val71Ala and Leu58His showed excellent survival. We want to stress that patients with other mutations than Val30Met are not a homogenous group, and we propose the term "non-Val30Met" to be used with caution and best be avoided. For several mutations data is still too limited to evaluate the efficacy of LTx, and it is therefore vitally important with continuous international collaboration to obtain more knowledge and further improve treatment guidance.

BO119

### RESULTS OF LIVER TRANSPLANTATION IN THE TREATMENT OF HEPATIC ALVEOLAR ECHINOCOCCOSIS

Marcin Kotulski<sup>1</sup>, Waldemar Patkowski<sup>2</sup>, Piotr Remiszewski<sup>2</sup>, Krzysztof Zieniewicz<sup>2</sup>, Marek Krawczyk<sup>2</sup>

<sup>1</sup>Department of General Transplant and Liver Surgery; <sup>2</sup>Department of General, Transplant and Liver Surgery, Medical University of Warsaw

**Background:** Alveolar echinococcosis (AE) is a rare disease caused by the *Echinococcus multilocularis* larvae growing in the liver. This observation suggests that liver transplantation (LTx) may be indicated when other therapies become ineffective and no extrahepatic lesions are founded. The purpose of this study was to assess the value and timing of LTx in the treatment of AE of the liver.

**Material and Methods:** A retrospective study was carried out, including all cases of LTx for AE performed in our Department between 2000 and 2014. There were 23 cases AE (17M, 6F) in middle age of  $43 \pm 13$ . In 18 cases (78%) LTx was a priori decided to be the method of management due to the advancement of the disease preventing radical surgery. In 5 cases (22%) prior surgery led to the LTx (one extensive liver resection, one unresectable alveococcosis recurrence within the liver and two cases of diagnostic laparoscopy/laparotomy). 11 classical and 12 piggy-back LTx from cadaveric donor were performed. All of the patients received additionally albendazol, prior to and after LTx – mean period 2 years.

**Results:** Complications were observed in 6 cases (26%) – wound infection in 4 cases, pneumonia in 1 and in 1 transient renal failure requiring dialysotherapy. Two patients (10%) died within the 1st post-LTx year – 1 due to sepsis leading to multiorgan failure. 2nd patient died 7 months after LTx due to sepsis after small bowel resection in the course of mechanical occlusion. In group of 6 patients appeared immunological exponents of infection recurrence in ELISA test, without changes in imaging examinations, after average  $24 \pm 12$  months. Actuarial survival rate after LTx was 91% at 1 year, 85% at 5, and 75% at 10 year.

**Conclusion:** Echinococcosis multilocularis of the liver in late stage can be considered as one of the indications of LTx, especially when other therapies are scarce and ineffective. In those cases LTx may be an appropriate option of radical treatment with excellent long term survival.

BO120

### LIVER TRANSPLANTATION FOR NONALCOHOLIC STEATOHEPATITIS: ORGAN WASTE OR SUCCESSFUL TREATMENT OF THE NEW EPIDEMIC? A SINGLE CENTER EXPERIENCE

Barbara Kern<sup>1</sup>, Benedikt Feurstein<sup>1</sup>, Josef Fritz<sup>1</sup>, Robert Sucher<sup>2</sup>, Cornelia Fabritius<sup>1</sup>, Manuel Maglione<sup>1</sup>, Claudia Boesmüller<sup>1</sup>, Dietmar Oefner<sup>1</sup>, Stefan Schneeberger<sup>1</sup>

<sup>1</sup>Innsbruck Medical University; <sup>2</sup>Charite Berlin

**Background:** Nonalcoholic steatohepatitis (NASH) may become one of the leading indications for liver transplantation (LT). The aim of this study was to describe the clinical outcome.

**Methods:** This is a retrospective analysis of 515 patients who underwent LT between 2002 and 2012.

**Results:** The incidence of NASH as primary indication for LT was 14.4% (74/515). The study population included 116 (22.5%) women and 399 (77.5%) men. NASH cohort compared to the nonNASH cohort showed no significance on patient survival ( $p = 0.109$ ). Patients with a malignancy displayed a shorter overall survival ( $p = 0.009$ ). Average MELD score was 21.0, average BMI 25.3. Patients with a lower MELD score at time of LT were associated with a significantly better overall survival ( $p = 0.043$ ). BMI  $>30 \text{ kg/m}^2$  had no impact on survival, neither in the NASH nor in the nonNASH cohort. Diabetes was diagnosed in 124 patients, compared to the patient cohort with no evidence of diabetes, overall survival was significantly shorter ( $p = 0.006$ ). NASH patients with diabetes had similar overall survival and complications when compared to NASH patients without diabetes ( $p = 0.242$ ;  $p = 0.112$  respectively). Donor data such as donor BMI  $>30 \text{ kg/m}^2$ , severe steatosis, age  $>55$  years, gender mismatch and cold ischemic time  $>14 \text{ h}$  had no impact on patient survival. Infection rate was significantly higher in the NASH cohort compared to other indications ( $p = 0.04$ ). NASH patients with HCC were associated with a significantly shorter overall survival compared to HCC patients with no evidence of NASH ( $p = 0.02$ ) while graft survival was comparable in both cohorts (Fig. 1).

**Conclusion:** Metabolic comorbidities significantly impact on patient survival. NASH predicts an inferior outcome in patients with HCC compared to other liver diseases. Accurate preoperative treatment of metabolic disorders and intensified infection prophylaxis should be considered in patients undergoing LT. Criteria for patients with HCC and NASH should be revisited.

Figure 1:

