come across with altered blood-glucose concentration in patients on TPN feeding who require closer monitoring with complex and dynamic treatment such as insulin. Despite such potential benefits, insulin added to TPN is still controversial due mainly to the potential risk of hypoglycaemia related to its biodisponibility.

Purpose Analyse and evaluate the efficacy and safety of fast-acting insulin added to TPN admixtures, in patients with altered glycaemia, followed up by nutrition support pharmacists (NSP).

Material and methods Observational and retrospective study carried out in a General Hospital for 19 months (January 2017 to July 2018). Data was collected from electronic clinical records and the electronic prescribing system. Data collected: total patients on TPN with altered blood-sugar levels followed up by the pharmacy team, patients treated with fast-acting insulin (TPN bag additive), daily (three times) blood-sugar levels (BMs), patient's demographics, hypoglycaemias (blood-sugar levels less than 70 mg/dL) and hyperglycaemias (BMs>180 mg/dL). Patients admitted to the critical care unit (CCU) or not followed up by the pharmacy team were excluded. We considered target BMs between 140–180 mg/dL. All insulin adjustments were done by NSP.

Results The total number of patients on NPT with altered BMs was 148, and 36 (24.3%) patients required fast-acting insulin therapy. Thirty patients were included in this study due to six being admitted to the CCU. Patients included: 20 were males (66.6%), average age 67 years (range 45–91). Twenty-five (83.3%) patients had hyperglycaemia (≥1BMs>180 mg/dL) of whom 17 (56.6%) required fast-acting insulin therapy on the TPN bag. Average NPT duration on fast-acting insulin-treated patients was 10 days (range 3–36). Average days BMs>180 mg/dL:4.5 (range 1–11). Average BMs>180 mg/dL: 242 mg/dL (range 181–427 mg/dL; mode: 220 mg/dL). One patient had hypoglycaemia non-insulin-related. None treated with fast-acting insulin had hypoglycaemia.

Conclusion Despite more than half of the patients treated with fast-acting insulin therapy having hyperglycaemia, none of them had hypoglycaemia. On the other hand, a cautious use of the fast-acting insulin TPN bag added could boost hyperglycaemias in our patients. Administering insulin along with TPN continuously appeared to be a safe method, providing a smoother glycaemic profile.

REFERENCES AND/OR ACKNOWLEDGEMENTS

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No conflict of interest.

5PSQ-006 USE OF SUBSTITUTE ENZYMATIC TREATMENT AND SUBSTRATE REDUCTION IN GAUCHER DISEASE

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Background Gaucher disease is included within lipidosis that occur due to mutations in the gene encoding the enzyme ß-glucosidase. As a result of this, a fatty substance accumulates, the glucocerebroside that is the cause of disease manifestations such as anaemia, thrombocytopaenia, hepatosplenomegaly and bone injuries. Available therapeutic options include enzyme replacement therapy (ERT) or substrate reduction (SRT) to prevent glucocerebroside accumulation.

Purpose To describe the use of ERT and SRT in patients with Gaucher disease.

Material and methods Retrospective observational study of all patients diagnosed with Gaucher disease in our area, followed up in our hospital and in treatment with ERT or SRT. Respective electronic medical records and analytics were reviewed to collect the following data: sex, age, symptomatology of Gaucher disease at the time of diagnosis, value of chitotriosidase before and after starting treatment, and adverse reactions to it.

Results A total of four patients (two males and two females) with an average age of 50 years were included. All patients had type1 Gaucher disease (not neuropathic). The initial treatment was miglustat (SRT) in three patients and velaglucerase (ERT) in one of them. The value of chitotriosidase before the start of treatment had a mean value of 11034 nmol/h/mL (7184–12777) and after treatment it was 1536 nmol/h/mL (239–3973).

All the patients presented at the beginning with typical manifestations of type 1 Gaucher disease as bone affectation (three), hepatosplenomegaly (three), anaemia (two) and thrombopaenia (two). Regarding the safety of the SRT, treatment with miglustat was started in three patients. It was finished in two cases due to bone progression and in one case due to poor tolerance (paresthaesia, diarrhoea, tremor and weight loss) and the TES was switched to imiglucerase or velaglucerase, which were well tolerated in all patients. All the patients presented improvement in the symptoms of Gaucher disease when starting ERT.

Conclusion TRS with miglustat is a convenient option due to its oral administration, although in three patients who were initially administered, it had to be suspended due to poor tolerance or progression. ERT has been shown to be effective and safe, and, despite not being curative, an improvement and even remission of certain symptoms of the disease has been proven.

REFERENCES AND/OR ACKNOWLEDGEMENTS

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No conflict of interest.

5PSQ-007 SEBELIPASE ALFA AS ENZYME REPLACEMENT THERAPY IN THREE PAEDIATRIC PATIENTS

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Background Lysosomal acid lipase deficiency (LALD) is a rare lysosomal disorder characterised by clinical with dyslipidaemia and steatohepatitis. Sebelipase-alfa is a recombinant human LAL, recently approved for clinical use in LALD.

Purpose Evaluation of the effectiveness and safety of sebelipase-alfa as enzyme replacement therapy in paediatric patients with LALD.

Material and methods Observational, retrospective study included three patients treated with sebelipase-alfa. From clinical history the following data were obtained: age, sex, diagnosis, values of total cholesterol, low-density lipoprotein

(LDL), high density lipoprotein (HDL), triglycerides (TG), alanine- aminotransferase (ALT) and aspartate aminotransferase (AST), liver size and fibrosis before and during treatment with sebelipase alfa, and adverse events. From the outpatient programme were obtained dose, administrations and weeks of treatment. The efficacy was evaluated by normalising the analytical values of the lipidic and liver profiles in three patients who participated in the clinical trial until April 2018.

Results Three male brothers 12, 15 and 17 years' old diagnosed with LALD before 5 years, heterozygous for mutation in the LIPA gene c.894G>A, c.256C>T. Before the trial, patients presented abnormal analytical values (except TG), hepatomegaly and fibrosis. Patients received continuous treatment with sebelipase-alfa at a dose of 1 mg/kg/2 weeks intravenously. Preparation of the medication was carried out by the hospital pharmacy service. In April 2018, after 225, 183 and 114 weeks of treatment respectively, all three patients maintained the values analysed in the range of normality (except HDL in two and TG in one patient). Hepatomegaly reversed in all patients. The means of the values and of the percentages of variation to the basal were: cholesterol 150.66 ±22.89 mg/dL (−34.19%), LDL 89.33±12.20 mg/dL (−46.90%), HDL 35.66±6.35 mg/dL (+1.55%), AST 30.66 ±4.04 IU/L (−64.88%), ALT 21.33±4.93 mg/dL (−65.99%) and TG 130±85.91 mg/dL (+2.12%). Concerning safety, two patients who suffered diarrhoea, and adverse effects related to the infusion were not reported.

Conclusion LALD is a rare disease, and sebelipase-alfa is the first drug authorised for its treatment. The response to treatment with sebelipase-alfa has been favourable from the beginning, with an improvement in the studied variables and a good safety profile in the reported cases.

REFERENCES AND/OR ACKNOWLEDGEMENTS

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No conflict of interest.

5PSQ-008 *CYP2C19 SNP'S* INFLUENCE ON CLOPIDOGREL RESPONSE IN PERIPHERAL ARTERY DISEASE PATIENTS

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Background Clopidogrel is a prodrug, metabolised to its active metabolite especially by the *CYP2C19* enzyme. The effect of *CYP2C19* polymorphisms on clopidogrel efficacy in coronary disease had been widely researched. The clopidogrel label recommends testing the *CYP2C19* loss of function alleles before the start of the treatment and the DPWG and CPIC pharmacogenetic dosing guidelines recommend switching clopidogrel in case of carrying the *CYP2C19*2 SNP* in coronary patients with stent. This remains unstudied in peripheral artery disease (PAD) patients.

Purpose Explore the influence of *CYP2C19* genetic polymorphisms on clopidogrel response in PAD patients.

Material and methods Peripheral artery disease patients treated with clopidogrel after percutaneous transluminal angioplasty were recruited. They were tested for carrying the *CYP2C19*2*, **3* (loss of function (LOF)) and **17* (gain of function (GOF)) allele. The primary endpoint was the occurrence of atherothrombotic ischaemic events, diagnosed by ultrasound imaging, during 12 months' follow-up. Furthermore, we collected data about clinical parameters (age, sex, ethnicity), co-medication during follow-up, vascular risk factors and surgical parameters.

We tested the association between carrying LOF or GOF alleles and the primary endpoint in a univariate analysis, and multivariate analysis including those clinical parameters previously related to clopidogrel response. OR and HR were calculated and P-values<0.05 were considered statistically significant.

Results Seventy-two patients were recruited, mean age 67.4 ±9.4 years, 22.2 females and 100% were caucasians. Carrying *CYP2C19 LOF* alleles was significantly associated with the primary endpoint in the single analysis (OR=4.49; 95% CI: 1.45 to 13.84; p=0.009), in the multivariate analysis (OR=4.89; 95% CI: 1.32 to 12.83; p=0.018). This association remains significant if we perform a survival analysis (HR=4.07; 95% CI: 1.80 to 9.20; p≤0.001). On the other hand, carrying *CYP2C19 GOF* alleles was not related to the primary endpoint.

Conclusion *CYP2C19 LOF* polymorphisms show a higher effect on clopidogrel response in PAD patients than that provided in acute coronary syndrome patients. These SNPs may be used as genetic markers of clopidogrel response in PAD patients. Clopidogrel treatment may be guided by *CYP2C19* genotyping, although further analysis should be performed.

REFERENCES AND/OR ACKNOWLEDGEMENTS

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No conflict of interest.

5PSQ-009 *CYP2C19 SNP'S* INFLUENCE ON CLOPIDOGREL RESPONSE IN CEREBROVASCULAR DISEASE PATIENTS: FINAL RESULTS

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Background Clopidogrel is a prodrug, which is metabolised to its active metabolite especially by the *CYP2C19* enzyme. Carrying some polymorphisms, contained in the DNA region encoding the *CYP2C19* expression, have shown a significant association with a lack of clopidogrel efficacy among coronary patients. This association had been widely researched and the clopidogrel label recommends testing the *CYP2C19* loss of function alleles before the start of the treatment, even DPWG and CPIC pharmacogenetic dosing guidelines, and recommend switching clopidogrel in case of carrying the *CYP2C19* loss of function alleles in coronary patients with stent. This remains unstudied in cerebrovascular-disease patients.

Purpose Explore the influence of *CYP2C19* genetic polymorphisms on clopidogrel response in cerebrovascular disease patients.

Material and methods Patients after stroke or transient ischaemic event (TIA) treated with clopidogrel after hospitalisation were recruited. These were tested for carrying the *CYP2C19*2*, **3* (loss of function (LOF)) and **17* (gain of function (GOF)) alleles. As primary endpoint we considered