using the software Pharma. Analysis of conformities showed that 204 patients (16%) had no Pharma exit prescription but exit treatments written in the EMR and 152 patients (12%) had no data either in Pharma nor in Axigate. Among the 933 patients, 348 (37%) had a copy/pasted prescription into their EMR and 585 (63%) presented discrepancies or lack of treatment into their EMR. No patient had the exit prescription scanned into their EMR although the software allows it. Two-hundred and seventy patients (29%) had no bodyweight provided even after the pharmacist notifications. Analysis of errors’ prescriptions: 255 were incorrect (4% of 7258 total number of drugs prescribed) with 36% drug redundancies, 29% incorrect dosage forms, including 7% of excessive dose and refractory period not respected in 25% cases. These errors were formulated daily by hospital pharmacists as a pharmaceutical opinion in Pharma but not applied by physicians in exit prescriptions.

Conclusion The exit prescriptions are not always recorded with CPOE Pharma. Several nonconformities and errors in outpatients’ prescriptions, mainly absence of bodyweight and incorrect drug prescriptions are noted. Hospital pharmacists’ initiatives, such as training and communication with physicians, have been set to improve exit prescriptions which will be served by community pharmacies.

REFERENCE AND/OR ACKNOWLEDGEMENTS

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GALENIC PREPARATIONS AND RARE DISEASES: GUANIDINOACETATE METHYLTRANSFERASE DEFICIENCY: EXPERIENCE IN A LOCAL HOSPITAL

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4CPS-188

Background Guanidinoacetate methyltransferase (GAMT) deficiency is a rare disorder (prevalence <1/1,000,000), inherited as autosomal recessive traits, characterised by an inborn error of creatine synthesis. Creatine deficiency results in a combination of symptoms such as intellectual disability, autistic behaviour, seizures, speech delay and hypotonia. Magnetic resonance is used at diagnosis and follow-up. The treatment goal is an increase in creatine levels in the brain with oral supplementation. Aminoacids (aa): 96.8% of PNs met the recommended requirements (3–4 g/kg/day). Sodium (Na) and potassium – carbohydrate, lipids, micronutrients (sodium, potassium, phosphorus, calcium), volume/kg and caloric requirements.

Material and methods The best regimen was established by a multidisciplinary approach in a function of patients’ weight and laboratory data (creatine and guanidinoacetate levels). An appropriate formulation was chosen according to active substance solubility and mucous membranes irritancy. Follow-up data were recorded retrospectively through medical records.

Results Two Egyptian patients, 13 and 19 years’ old, weight 56 and 94 kg respectively, in 2012 were diagnosed with GAMT deficiency by the Paediatric Unit. We chose unitary solid formulation: ornithine maps of 5 g for the first patient (10 g/die), maps of 2 g for the second (7 g/die) (106 mg/kg/die). Creatine had been given as powder, with a specific doser, considering high daily amount: 11 gx2/die for the first patient and 12 gx3/die for the second patient (382 mg/Kg/die). Concerning sodium benzoate, an irritant for mucosa, a 20% liquid formulation was chosen, to be administered with fruit juice. Clinicians decided a posology of 59 mg/kg/die, so 9 mLx2/die were administered to the first patient, and 14 mLx2/die were administered to the second patient. Patients since 2012 have not manifested adverse drug reactions and therapy has brought a stable clinical picture: optimal creatine level, measured as peak at MR, and low levels of guanidinoacetate on spot (8.3 mcML), indicative of good metabolic control.

Conclusion GAMT deficiency is a rare cerebral disorder, with a high impact on patients’ quality of life. A palliative approach is possible only through galenic preparations. Personalised therapies allow these patients to manage intellectual and movement disability in a better way, contributing to improving and/or stabilising the clinical picture.

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ADEQUACY OF THE PRESCRIPTION OF PARENTERAL NUTRITION IN NEONATOLOGY

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4CPS-189

Background Nowadays, there is a stronger consensus on the proceedings of nutritional support with parenteral nutrition (PN) in paediatrics and nutritional requirements in order to improve the process quality and the patient’s safety.

Purpose Review the prescriptions of PN to identify the degree of adherence to the available evidence (Clinical Practice Guide SENPE/SEGHNPN/SEFH 2017) and propose areas for improvement.

Material and methods Retrospective study of newborn patients who received PN during 2017 in the area of neonatology in our hospital.

Patients divided according to the age ranges established by the guidelines: preterm newborn (RNPT) and term newborns under 1 month (RNAT).

Variables: contributions of macronutrients (aminoacids, glucose, lipids), micronutrients (sodium, potassium, phosphorus, calcium), volume/kg and caloric requirements.

Data collected from PN elaboration program, Nutriwin, treated in Excel.

Results One-hundred and seventy-nine RNPT and 2,429 PNs were prepared and validated. Aminoacids (aa): 96.8% of PNs met the recommended requirements (3–4 g/kg/day). Carbohydrates (CH): 85.4% were adjusted and 13.4% were above the recommendations (6–12 g/kg/day). The limit of CH (16–18 g/kg/day) was not exceeded. Lipids: they did not exceed the maximum limit (3–4 g/kg/day). Sodium (Na) and potassium.
Rituximab is a monoclonal antibody directed against the CD20 antigen, expressed on the surface of B-lymphocytes, promoting the lysis of the cells. It is labelled for adult different indications, non-Hodgkin’s lymphoma, chronic lymphocytic leukaemia (CLL), rheumatoid arthritis and granulomatosis with polyangiitis and microscopic polyangiitis. Nowadays it is commonly used as off-label treatment in many other diseases, including some paediatric disorders.

**Purpose**
To describe the patterns of rituximab use in a paediatric teaching hospital.

**Material and methods**
We conducted a retrospective observational study involving all patients treated in a paediatric hospital with rituximab from January 2001 to June 2018.

Clinical data were collected from electronic patients’ medical records, including: patient age, prescribing services and indication.

**Results**
The study comprised 145 patients (39% males) with a median age of 15.4 years. The principal indications according to the prescribing services were:

- Forty-seven patients of the nephrology unit: resistant or refractory nephrotic syndrome (34) and transplants—rejects (13).

- Forty patients of the oncology unit: non-Hodgkin lymphoma (23), syndrome opoclonus–myoclonus in neuroblastoma (14) and others (three).

- Twenty-five patients of the haematology unit: disease: haemolytic anaemia (11), leukaemia (four), haemophagocytic syndrome (four), thrombocytopenic purpura (two) and others (four).

- Thirteen patients of the rheumatologic diseases unit: juvenile idiopathic arthritis (four), systemic lupus erythematosus (four), vasculitis (two) and others (three).

- Twelve patients of the neurology unit: autoimmune encephalitis (nine), post–Herpes Simplex encephalitis (two) and others (one).

- Seven patients of the infectious unit: Epstein–Barr virus infection (seven).

- One dermatologic disease: Steven–Johnson disease (one).

No unexpected side effects were observed outside those reported in the summary characteristics of the product.

**Conclusion**
In paediatrics, rituximab treatment is prescribed for off-label indications. Our study shows that rituximab is used in a wide variability of disorders, where the renal disease, specifically the nephrotic syndrome, is the most common indication as a second-line treatment.

Although the utilisation of rituximab increases every year and some uses are well described, further studies for some indications are necessary to establish a correct safety and efficacy profile in children.

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