P-090 - SLEEP DISORDERS CHARACTERIZATION IN KRABBE DISEASE PATIENTS’ FROM THE REFERENCE CENTER OF BAHIA, BRAZIL

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**INTRODUCTION:** Krabbe disease (KD) is an autosomal recessive lysosomal disorder affecting the white matter of central and peripheral nervous systems. It comprises a spectrum ranging from infantile to later onset disease. Sleep disorders can increase morbidity and health care costs. In controlled studies, volunteers who were restricted to four to five hours of sleep for a few days experienced worsening neurocognitive, behavioral, metabolic, and autonomic parameters. **OBJECTIVE:** To describe a serial cases of 2 individuals diagnosed with Krabbe disease followed in an reference center in Salvador, Bahia, Brazil and their sleep disorders.

**METHOD:** Observational, descriptive study, from data review of patients with Krabbe disease diagnosis followed in a reference center. Three types of questionnaires were applied to identify sleep disorders, such as the Sleep Disturbance Scale for Children (SDSC), range from 26 to 130 with a cut-off point of 39 that characterizes sleep disturbance; the Epworth Sleepiness Scale (ESS) to measure the daytime sleepiness, score >10 points indicates sleepiness; and the Conners' Scale short Form to suspect of hyperactivity disorders with a score ≤ 12 to girls and score ≤ 17 to boys as a positive finding. **RESULTS:** Two twin girls with neuropsychomotor development regression onset one 1 year and 2 months old and the other 2 years and 6 months old, associated seizures, truncal hypotonia, spasticity of extremities an biochemical diagnosis of Krabbe's disease 2 years and 10 months old and both presents loss of milestones. They are 4 years and 3 months old and both cases are later onset type of KD. The SDSC questionnaire observed that the score of Patient 1 and Patient 2 were the following values 62. Both patients pointed above the cutoff point, characterizing the sleep disorder. The analysis of Conners scale and Epworth short form scale showed that Patient 1 e 2 presented the following values 3 and 1, indicated normality. **CONCLUSION:** The sleep disorders should be performed in Krabbe disease, which are very frequent despite the lack of sufficient data in the literature. Sleep disturbances can interfere with the quality of life of the patients and caregivers.