constant health system adequation is crucial to cope with this situation and may be so even post-pandemic.

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PARENTAL PERSPECTIVE OF FEEDING DIFFICULTIES IN CHILDREN WITH HEPATIC GLYCOGEN STORAGE DISEASES

CATEGORIA DO TRABALHO: PESQUISA

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Introduction: Hepatic glycogen storage diseases (GSD) are inborn errors of metabolism that result in a defect in glycogen synthesis or degradation, predominantly affecting the liver and muscles. Treatment is nutritional, with diet restrictions and regular use of raw cornstarch. To date, no study has evaluated the degree of feeding difficulty in hepatic GSD and/or its correlation with parental stress. Objective: This study aims to investigate the presence and severity of feeding difficulties in children with GSD and the presence of parental stress using the Parental Stress Scale. Methods: Twenty-nine individuals (parents and/or caregivers of individuals with GSD) participated in the study, responsible for children aged 6 mo. to <7yo. The presence and severity of feeding difficulties in children with GSD was evaluated through Brazilian Infant Feeding Scale (EBAI) and the presence of parental stress was evaluated using the Parental Stress Scale (EEPa), through the use of a standardized form with application of consent form (CAAE: 30895020.0.1001.5329).Results: The patients under their care were predominantly male (19/10), had a mean age of 47.75 months, with mean age at GSD diagnosis 8.39 months. Diagnosis of GSD type Ia (15) and Ib (5) were the mostly reported, followed by type III (2), VI (1), IX (2) and unknown type (4). In this sample, 22/29 (76.0%) had feeding problems, classified as mild (24.0%), moderate (24.0%) and severe (28.0%). The EBAI score was significantly higher among female patients and among patients who did not have meals with families. One parent/caregiver had high parental stress. No statistical significance was observed when comparing feeding difficulties and parental stress. Conclusions: This study corroborated the findings of feeding difficulties in groups at risk and showed the prevalence and degree of feeding problems in this population. Although the scales were not related, quantifying parental stress enabled to verify impact of the diagnosis on the quality of life the caregivers perspective. More studies in this field are warranted aiming for better treatment of GSD.

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ESTRESSE OXIDATIVO EM CÉREBRO DE CAMUNDONGOS NOCAUTE PARA ACIDEMIA GLUTÁRICA TIPO I SUBMETIDOS A UMA SOBRECARGA DE LISINA E EFEITO PROTETOR DO BEZAFIBRATO

CATEGORIA DO TRABALHO: PESQUISA

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A acidemia glutárica tipo I (AG I) é um erro inato do metabolismo de herança autossômica recessiva causado por mutações no gene que codifica a enzima glutaril-CoA desidrogenase (GCDH), levando à diminuição severa da atividade dessa enzima, e acúmulo dos ácidos glutárico e 3-hidroxiglutárico no cérebro e líquidos biológicos dos afetados. Os pacientes desenvolvem degeneração aguda do estriado durante situações de estresse catabólico, seguindo-se de distonia e discinesia. Dano cortical progressivo também ocorre nessa doença. Considerando que os mecanismos do dano estriatal e cortical não estão totalmente elucidados nessa doença, o objetivo deste trabalho foi avaliar parâmetros bioquímicos da homeostase redox, em um modelo genético de AG 1 submetido à sobrecarga de lisina que foi previamente demonstrado causar lesão estriatal nesse modelo. Um possível efeito protetor do bezafibrato (bez) também foi investigado. Camundongos selvagens (Gcdh+/+) e nocautes para GCDH (Gcdh-/-) foram submetidos ao tratamento com bez (75 mg / kg / dia) durante 7 dias. A partir do dia 5, parte dos animais Gcdh+/+e Gcdh-/-receberam uma dieta rica em lisina por 48h (4,7 %) até o dia 7, quando foram eutanasiados. O cérebro foi dissecado, e o estriado e o córtex cerebral foram isolados e