P-001 - TRANSITION FROM CHILD TO ADULT IN PATIENTS WITH INBORN ERRORS OF METABOLISM (IEM): OUR EXPERIENCE WITH PATIENTS DERIVED FROM PEDIATRIC HOSPITAL

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INTRODUCTION: Advances in diagnosis and treatment of IEMs have improved their prognosis and there are more patients who reach adulthood and need to be transferred to an appropriate center for their age. OBJECTIVE: To describe our experience in the care of adolescents and adults with IEM derived from a pediatric hospital: population characteristics, strengths and difficulties encountered in monitoring, challenges to be addressed. MATERIALS AND METHODS: Review of medical records (MR). We describe filial and anthropometric data, statistics of consultations and interventions carried out. RESULTS: Our team consists of a specialist doctor and a nutritionist. From 2011 to 2018, 20 patients have been referred: 65% women, 35% men. Age first consultation: 18.9 ± 2.55 years BMI: 24.7 ± 4.86 kg/m2 Pathologies: - 20%: alteration of urea cycle - 15%: glycogenosis III and phenylketonuria - 10%: propionic acidemia, fructose intolerance/fructosemia, glycogenosis IA - 5%: deficit of HMGCoALiasa, deficit of betaketothiolase, isovaleric acidemia and homocysteinuria Progression of referrals 2011-2018: from 1 to 20 patients. 20% did not attend to consultation during 2018. Consultations/patient/year: between 1 and 2.93. Interventions: calcium supplementation in 40% for not meeting requirements. Strengths: - 100% understand their illness and know treatment guidelines. - 100% brought copy of pediatric MR - 10% had hospitalization due to metabolic decompensation of infectious origin. - Fluid contact with derivative pediatricians. Difficulties: - 20% bring dietary records - 20% discontinuously comply dietary guidelines (according to availability of formulas and aproteic foods). - We don’t have a specialized laboratory. Patients do analysis in pediatric center. - Up to 2017 there was a neurologist specialized in IEM. Today patients must be referred. - Difficult compliance with emergency letter on guard with metabolically decompensated patients. Challenges to address: - Chronic complex disease in the context of adolescence. - Physiological or pathological situations that appear concomitantly: pregnancy, overweight (30%), obesity (10%), dyslipidemias - Contraception and incorporation of family planning (Genetic Counsel). CONCLUSIONS: The experience in the transition has been positive and opens the expectation of interaction between pediatric and adult hospitals, serving as experience for other transitions of other chronic diseases.