Journal of Pediatric Advance Research

ATHENAEUM SCIENTIFIC PUBLISHERS

Research Article

Exocrine Pancreatic Insufficiency: A Case Report at the Mother and Child Center of the Chantal Biya Foundation

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Citation: Makowa LK, et al. Exocrine Pancreatic Insufficiency: A Case Report at the Mother and Child Center of the Chantal Biya Foundation. J Pediatric Adv Res. 2023;2(1):1-6.

http://dx.doi.org/10.46889/JPAR.2023.2101

Received Date: 25-01-2023

Accepted Date: 13-02-2023

Published Date: 20-02-2023

Abstract

Introduction: Exocrine pancreatic pathologies are rare conditions in children. Their diagnosis should be evoked and systematically sought in any child presenting with severe acute or chronic abdominal pain, malnutrition and steatorrhea. The management is complex and may be multidisciplinary over the long term.

Case Presentation: We report the case of a 7-month-old infant referred for unexplained malnutrition. Born macrosomic, since the age of 6 weeks he had been vomiting and not gaining weight. Then the progressive appearance of steatorrhea and a malabsorption syndrome led to acute malnutrition. Exocrine pancreatic insufficiency was demonstrated with low lipase and the patient was taken care of. Treatment consisted of oral administration of pancreatic enzymes and nutritional rehabilitation. Significant and favorable evolution was noted as early as within two weeks from treatment initiation.

Conclusion: Exocrine Pancreatic Insufficiency (EPI) is a rare but severe condition in pediatrics. The etiological diagnosis is often linked with genetic diseases, and so not always obvious in our context. However, clinical evaluation and complementary examinations may serve as diagnostic guidelines. Management is palliative and allows rapid improvement of the patient as early as within two weeks from start.

Keywords: Pancreatic Insufficiency; Malabsorption; Malnutrition

Introduction

Exocrine Pancreatic Insufficiency (EPI) is the inability of the pancreas to produce enough enzymes to digest and absorb food. It is a rare condition in children worldwide, and even less in Africa [1-9]. A loss of approximately 97% of pancreatic exocrine function is required to develop significant symptoms. According to Scheers, et al., these characteristic signs and symptoms include chronic diarrhea, steatorrhea, underweight and growth retardation [1-12]. We present a case of EPI, an exceptional pathology in pediatrics, which occurred in an infant born macrosome and who had a normal diet with breastfeeding before the onset of the disease [13-20]. This case allows us to highlight the diagnostic challenges due to metabolic repercussions and limited resource settings, as well as the therapeutic difficulties with a need for regular multidisciplinary follow-up [21-39]. Our discussion will be supported by a review of the literature on the topic.

Case Presentation

We report the case of a 7-month-old male infant referred for unexplained malnutrition. From the history of the disease, it was noted that since the age of 6 weeks the child presented with postprandial vomiting with a stagnation of the weight curve. Initial investigations including abdominal ultrasound, urinalysis and culture had respectively ruled out hypertrophic pyloric stenosis,

digestive malformation and urinary tract infection. Other multiple consultations had followed in different hospitals, during which the child had been transfused for severe anemia, antibiotics had been prescribed and nutritional support with therapeutic milk F100 instituted. The evolution was marked with the persistence of weight loss, prompting a pediatric endocrinology consultation. As past history, the child was born at 41 weeks of Gestational Age (GA), with a birth weight of 4100 g, without delay in the expulsion of meconium. His vaccines were up to date for his age, there was no known allergy or atopy. His diet had consisted of exclusive breastfeeding for up to 2 months, then an Anti-Reflux breast milk substitute. Weaning and food diversification was done at the age of 6 months, but the child ate very little because of vomiting. He presented a psychomotor delay with a developmental age of 5.75 months. He was the 5th in a family of 5 children, from a non-consanguineous couple with an advanced parental age. Fig. 1 shows the family genetic tree.

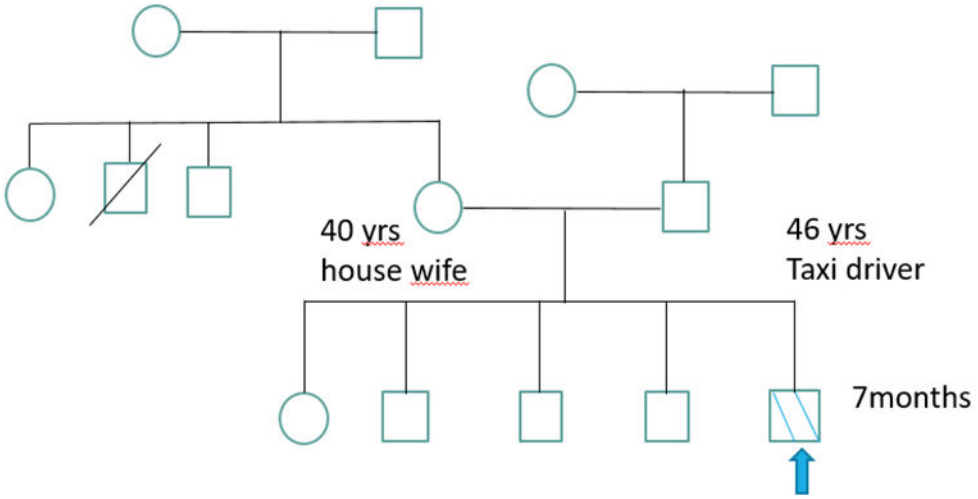

Figure 1: Family genetic tree.

During systems enquiry, pseudo constipation was reported with hard stools at first, then soft, shiny, frequent, abundant, and malodorous. With this, was associated non-bilious vomiting. There was no cough or other respiratory symptoms.

On physical examination, the general condition of the patient was altered by severe acute to chronic malnutrition, with a weight of 4200 g, a height of 62 cm, and the various indices: height/age, weight/age and weight/height were < - 3 z-score. The child presented with sparse hair, a very weak adipose panicle, no notable abnormalities on the pulmonary examination, no finger clubbing, and no edema of the limbs. This clinical picture suggested a malabsorption syndrome whose etiology remained to be investigated. The complementary examinations requested included the lipid profile, which found a reduction in blood lipids, in particular: total cholesterol at 0.71 g/l, HDL cholesterol at 0.29 g/l, and LDL cholesterol at 0.15 g/l. But, on the other hand, triglycerides were normal at 1.35 g/l. The complete blood count showed hypochromic macrocytic anemia at 7.9 g/dl, while the exploration of the lymphocyte and thrombocyte lines was normal. We then asked for lipasemia which was reduced to 18 IU, blood amylase was within the limits of the norm. The blood electrolytes showed hyponatremia at 130 meq, hyperkalemia at 5 meq, and hypercalcemia at 104 mg/dl. Serum chloremia, phosphate, magnesium, protein and iron were normal, as were uraemia, creatinine, TSH and T4. Faced with this clinical and biological malabsorption syndrome, the diagnosis retained was exocrine pancreatic insufficiency. However, the absence of molecular biology prevented us from deepening etiological research.

The treatment consisted of nutritional support, considering caloric needs of 110 kcal/kg/d, with a food sheet that was explained and given to the parents. We also prescribed the administration of pancreatic extract including digestive enzymes (Creon) at initial doses of 2000 U/kg/d, during each non-dairy meal of the day. Iron supplementation at 3 mg/kg/d was equally prescribed, as well as fat-soluble vitamin (ADEK) at 1 ml/d. A first step follow-up appointment was scheduled and the child was reassessed after two weeks of treatment. Within this period, the child was seen with effective weight gain and a general condition improvement. Natremie, kalemia and calcemia had also returned to normal values. As a matter of fact, the weight had increased to 5 kg, making a net gain of 800 g within 2 weeks. We were thus able to gradually increase the doses of pancreatic extracts being administered to the patient, towards a maximal possible dose of 10000 U/kg/d. This was done by taking into account the child's

meal quantities and his appetite, for greater effect. Fig. 2 shows the evolution of the child's weight within the first two weeks of treatment.

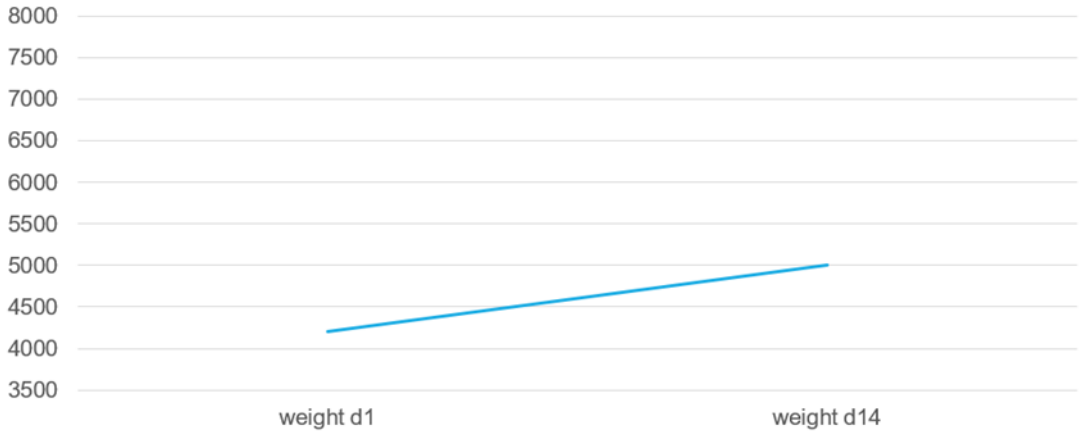

Figure 2: Weight gain curve within 14 days.

Discussion

Exocrine Pancreatic Insufficiency (EPI) is a rare condition in children. In Africa, the prevalence is unknown and few scientific articles have been published on the topic [1-12]. EPI is the inability of the exocrine pancreas to secrete digestive enzymes, including pancreatic amylase, lipase, and trypsin. This causes difficulties in digesting food from the jejuno-duodenum and as a consequence, absorption abnormalities in the ileum, although some infants may be feeding properly [1-9, 13-20]. It is estimated that a loss of approximately 97% of pancreatic exocrine function would be required for signs and symptoms to develop. Due to digestive and absorptive dysfunctions, most of the food ingested and nutrient content are wasted in the stools of patients, as described in the case reported. The signs and symptoms may occur as early as within the very few days following birth, in which case it appears an emergency with need for adequate assessment and management to prevent rapid deterioration [40-43]. This with possible coexistence of associated malformations, infections, infant wasting and with considerable parental worries as repercussion [21-30,40-49].

Clinically, patients present with a malabsorption syndrome comprising steatorrhea with generally copious, oily, pale, and malodorous stools [1-9]. Thus, will follow a deficiency in fat-soluble vitamins (A, D, E, K), weight loss or lack of weight gain, growth retardation, bone damage and coagulation disorders [1-12, 30-39]. On the paraclinical level, the biological malabsorption syndrome is manifested by a deficiency anemia in vitamin B-12 especially, a decrease in blood lipids, sometimes with the exception of triglycerides. Indeed, the absorption of iron takes place better in an acid medium and therefore is independent of the pancreatic juice which is alkaline in essence. Nevertheless, frequent vomiting with metabolic alkalosis and loss of appetite caused both forms of anemia in our patient. It should be noted that despite the poor absorption of lipids reflected by their overall low blood level, serum lipoprotein lipase works to normalize triglyceridemia to the detriment of other lipids [1-12,30-39]. On the other hand, the hypercalcemia observed, unlike hypocalcemia expected with vitamin D deficiency can be explained by parathormone effect, although phosphatemia remained normal. However, the considerably lowered lipasemia, because secreted almost exclusively by the pancreas, permitted to attribute the disorders in our patient to EPI [1-12,30-39]. This contrasted with normal amylase level, but whose secretion is both salivary and pancreatic, and therefore less specific to EPI diagnosis.

When faced with a maldigestion-malabsorption syndrome in pediatrics, cystic fibrosis is almost always evoked, due to its severity and epidemiological frequency, especially in Caucasians. Nevertheless, Lacrosniere, et al., in 2021 noted an increase in cystic fibrosis from 2 to 10% in blacks, 83% of which was expressed through exocrine pancreatic insufficiency [1-12,30-39]. However, the difficulties of access to molecular biology in our context made the exclusion of various genetic causes difficult at first sight. This including cystic fibrosis as well as the rare Shwachman-Bodian-Diamond syndrome and the Johanson-Blizzard syndrome. In effect, these pathologies are characterized by EPI, but often associated with multisystem involvement that were not found in our patient [7-9].

Conclusion

Exocrine pancreatic insufficiency is a rare condition in children, very often linked with genetic diseases. The etiological diagnosis is based on molecular biology which is not always available in some context. Therefore, there is need for detailed clinical examination of patients and judicious choice of complementary tests for orientation. Management is essentially palliative based on the oral administration of pancreatic enzyme preparations. Regular follow-up of such pediatric patients is required, considering regression of digestive disorders, weight gain improvement growth and development.

Acknowledgement

Hospitals authorities and all collaborators to this project.

Conflict of Interest

The authors have no conflict of interest to declare.

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