

Casos Clínicos

Reducing frequency in respiratory infections in glycogen storage disease Ib with a nutritional supplement. Case report

Rev. OFIL 2017, 27;2:210-212

Fecha de recepción: 14/06/2016 - Fecha de aceptación: 15/10/2016

CARNERO GREGORIO M¹, TORRES CRIGNA A², CARNERO GREGORIO O², CORBALÁN RIVAS A², RODRÍGUEZ CERDEIRA C³

1 Departamento de Bioquímica, Genética e Inmunología. Universidad de Vigo (España)

2 Asociación Española de Pacientes con Glicogénesis (España)

3 Departamento de Dermatología. CHUVI y Universidad de Vigo (España)

SUMMARY

Introduction and objectives: Infections are common in glycogen storage disease type Ib (GSD Ib) patients due to associated chronic severe neutropenia. Immunonutrition might help to minimize infections in these individuals. The aim is to compare the count of respiratory infection episodes in a GSD Ib patient, before and during a supplementary nutritional treatment.

Materials and methods: A retrospective case report study in a 42-year-old male GSD Ib patient. The supplementary nutritional treatment began in June 2002. A period of 20

Key Words: Glycogen storage disease, nutritional supplements, impact, immunonutrition, respiratory infections.

years was evaluated: 10 years prior to the beginning of the treatment as well as the 10 years of the actual treatment. No other dietary or therapeutic modifications were made. **Results and discussion:** Seven respiratory infections were recorded before the treatment. In all cases, antibiotic therapy as well as granulocyte-colony stimulating factor (G-CSF) were needed for the complete remission of infections. No airway infections that involved medical care or treatment were recorded in the post-treatment period.

Conclusion: Immunonutrition could help reduce the number of respiratory complications in GSD Ib patients.

Reducción de la frecuencia de infecciones respiratorias en glucogenosis Ib con un suplemento nutricional. Caso clínico

RESUMEN

Introducción y objetivos: Los procesos infecciosos son comunes en pacientes con glucogenosis tipo Ib (GSD Ib) debido a la neutropenia severa crónica asociada a su enfermedad. La inmunonutrición podría ayudar a minimizar las infecciones en estos individuos. El objetivo es comparar el número de episodios infecciosos respiratorios en un paciente afectado de GSD Ib antes y durante un tratamiento con suplementos

nutricionales.

Materiales y métodos: Estudio longitudinal retrospectivo de un paciente varón de 42 años de edad afectado por GSD tipo Ib. El tratamiento con el suplemento nutricional se inició en junio del año 2002. Se evaluaron un total de 20 años: los 10 años anteriores al tratamiento y los 10 años durante los cuales el paciente fue tratado con el suplemento nutricional. No se realizaron otras modifica-

ciones dietéticas o terapéuticas.

Resultados y discusión: Antes del tratamiento se registraron 7 infecciones respiratorias. En todos los casos se necesitaron antibióticos y factor estimulante de colonias de granulocitos (G-CSF) para la completa remisión de las infecciones. Durante el periodo del tratamiento con suplementos nutricionales no se registraron infecciones de las vías respiratorias que precisaran cuidados médicos o tratamiento antibiótico. **Conclusión:** La inmunonutrición podría ayudar a reducir el número de complicaciones respiratorias en los pacientes con GSD Ib.

Palabras clave: Glucogenosis, suplementos nutricionales, impacto, inmunonutrición, infecciones respiratorias.

Correspondencia:

Miguel Carnero Gregorio

Universidad de Vigo

(Departamento de Bioquímica, Genética e Inmunología)

Prexigueiro, 70

32418 Ribadavia - Ourense

Correo electrónico: miguel.carnero.gregorio@hotmail.com

INTRODUCTION AND OBJECTIVES

Glycogen storage disease (GSD) is a group of metabolic disorders that include those related to glycogen metabolism. At present, 16 types are known, from 0 to XV, each having different degrees of severity and impact on organs¹. In hepatocytes, there is a pathway that transforms glucose-6-phosphate (G6P) into glucose. The malfunction of this pathway leads to GSD I with a very low prevalence (about 1:100,000–1:300,000 cases). In this disorder, subtype Ib affects 20% of patients. Some of the symptoms manifested are hypoglycemia, hypertriglyceridemia, hyperuricemia and neutropenia. Due to this severe chronic neutropenia, recurrent infections are frequent in these patients, including those in the respiratory tract, which are between the most common ones².

Currently, there are multiple studies on immunomodulatory diets and their capacity to stimulate the immune system^{3,4}. At this time, no consensual conclusions have been reached on the beneficial effects in different pathologies.

Authors agree that some nutrients may help reduce the number of infections. Arginine (Arg) is one of the nutrients which may be effective in reducing infections of the upper respiratory tract^{5,6}.

The number of myeloid suppressor cells (MDSC) increase in response to certain disorders: cancer, autoimmune diseases, acute and chronic infections and stress. MDSC have a role in reducing the immune response. Moreover, MDSC regulate the availability of arginine needed for the normal function of T lymphocytes⁷. Nutritional supplements containing arginine and omega-3 fatty acids (ω -3), seem to neutralize the regulatory effects of MDSC⁸.

Arg is transformed into nitric oxid (NO) that, when in deficit, impairs blood microcirculation⁹.

Arginine supplementation improves both wound healing and immune function by ameliorating the function of T cells, often associated with traumas. Oral administration of 30 g of this amino acid in human volunteers gives rise to an increase in lymphocyte blastogenic transformation when stimulated by mitogens¹⁰.

This report focused on the observation of respiratory tract infections due to it was the only patient's disease condition which was influenced by the introduction of nutritional supplement discussed here.

The aim was to compare the count of respiratory tract infections in a 42-year-old patient with GSD Ib, both before and during the use of an oral nutritional supplement containing Arg.

MATERIALS AND METHODS

Retrospective case report study of a 42-year-old patient affected by GSD type Ib. Related with his main pathology, the following chronic items are active in the patient: neutropenia, gastroesophageal reflux disease, like-Cronh inflammatory bowel disease, periodic furunculosis, nephrolithiasis, osteopenia and rarely iron deficiency anemia. Other not GSD related active disease: bilateral sensorineural deafness. Medical records of all respiratory tract infection episodes were collected. The number of other infection episodes was lower in comparison with the respiratory tract infections, so we only determine those ones.

Impact® enteral (Nestlé Health Science®, Switzerland) was used as the nutritional supplement treatment, with

a 250 mL dose in the morning and at night. The total arginine amount per day was 6.25 g. Impact® enteral also contains other immunonutrients as omega-3 fatty acids (1.7 g/L) and dietary nucleotides (1.2 g/L).

The supplementation started in June 2002. A period of 20 years was evaluated: 10 years prior to the beginning of the supplementation (1993-2002) as well as the 10 years of the actual supplementation (2003-2012). No other dietary or therapeutic modifications were made.

RESULTS AND DISCUSSION

The count of respiratory complications in the patient's medical records in the 10-year period prior to treatment was five: tuberculosis in RLL (right lower lobe), pneumonia in LLL (Left lower lobe), sinusitis, an undetermined count of respiratory tract and tonsil infections and cold symptoms with fever and yellowish expectoration lasting 5 days. Dates can be seen in table 1.

Oral antibiotics and treatment with granulocyte colony-stimulating factor (G-CSF) were administered for the remission of the infections in all of the above cases.

In the period of supplementation with Impact® enteral no respiratory tract infections requiring medical assistance or treatment were recorded. The patient experienced only two common colds during the 10-year treatment.

As shown in figures 1 and 2, once on treatment with nutritional supplement, the variation of neutrophil count showed a noticeable increase.

The median and variance of both periods can be seen in table 2.

This study covers the adult stage of the patient's life between the age of 21 and 40. Throughout the full 20-year period, the patient has been living in the same city, has maintained the same eating habits and has been on the same prolonged pharmacological therapy. Therefore, no other known factors have intervened in the individual.

Table 1
Medical records of respiratory complications in the pre-treatment period

Pathology	Date
RLL Tuberculosis	October 1995
Virus infection	February 1997
LLL Pneumonia	August 1997
Several infections	Summer 1998
Sinusitis	Not specified
Catarrhal inflammation	Not specified

Table 2
Median and variance

Period	Median (units/ μ L)	Variance
1993-2002	$0.3 \cdot 10^3$	0.43
2003-2012	$0.41 \cdot 10^3$	0.04

Figure 1
Blood neutrophil count in pre-treatment period

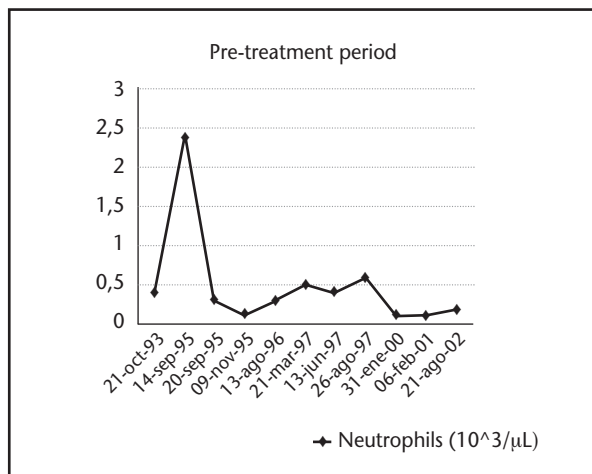
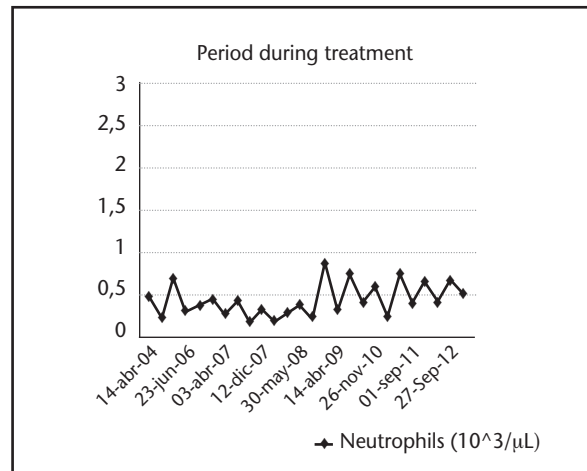


Figure 2
Blood neutrophil count during treatment period



CONCLUSION

Data show an improvement in the patient's immune system once on the nutritional supplement. The use of immunonutrition in the diet of GSD Ib patients might help to reduce the count of respiratory infections due to chronic neutropenia. Keeping a close watch on the total count of neutrophils is a good marker in assessing the effectiveness of this type of supplements.

More studies with a cohort of patients is needed to validate arginine as a supplementary immunonutritional therapy.

Conflict of interest: The authors declare no conflicts of interest.

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