

Mauriac Syndrome – a rare type 1 diabetes mellitus complication and an opportunity for intervention

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INTRODUCTION

Mauriac Syndrome is characterized by the presence of hepatomegaly, growth retardation, delayed puberty and cushingoid features. This entity is traditionally diagnosed in patients with type 1 diabetes mellitus (T1DM) and poor glycaemic control. However, the impact of metabolic control in the normalization of hepatic analytic profile is not clarified.

METHODS

Retrospective study of **five patients** with T1DM with hepatic histopathology indicative of **Mauriac Syndrome**.

RESULTS

Patient	Sex	Age	End organ damage	HbA1c (%)	AST (10-37 U/L)	ALT (10-37 U/L)	Hepatomegaly* (<15 cm)	Other Mauriac Syndrome features
1	М	22	Retinopathy Nephropathy	15,7	226	176	17 cm	 Cushingoid features Short stature – 160cm#
2	F	29	Retinopathy	10,9	991	461	25 cm	
3	F	20	Retinopathy Nephropathy Neuropathy	14,0	102	39	22 cm	• Short stature – 156cm ⁺
4	F	20	Retinopathy Nephropathy Neuropathy	11,7	227	439	23 cm	 Cushingoid features Short stature – 155cm⁺
5	F	15	Retinopathy	11,7	150	102	21 cm	• Short stature – 154cm

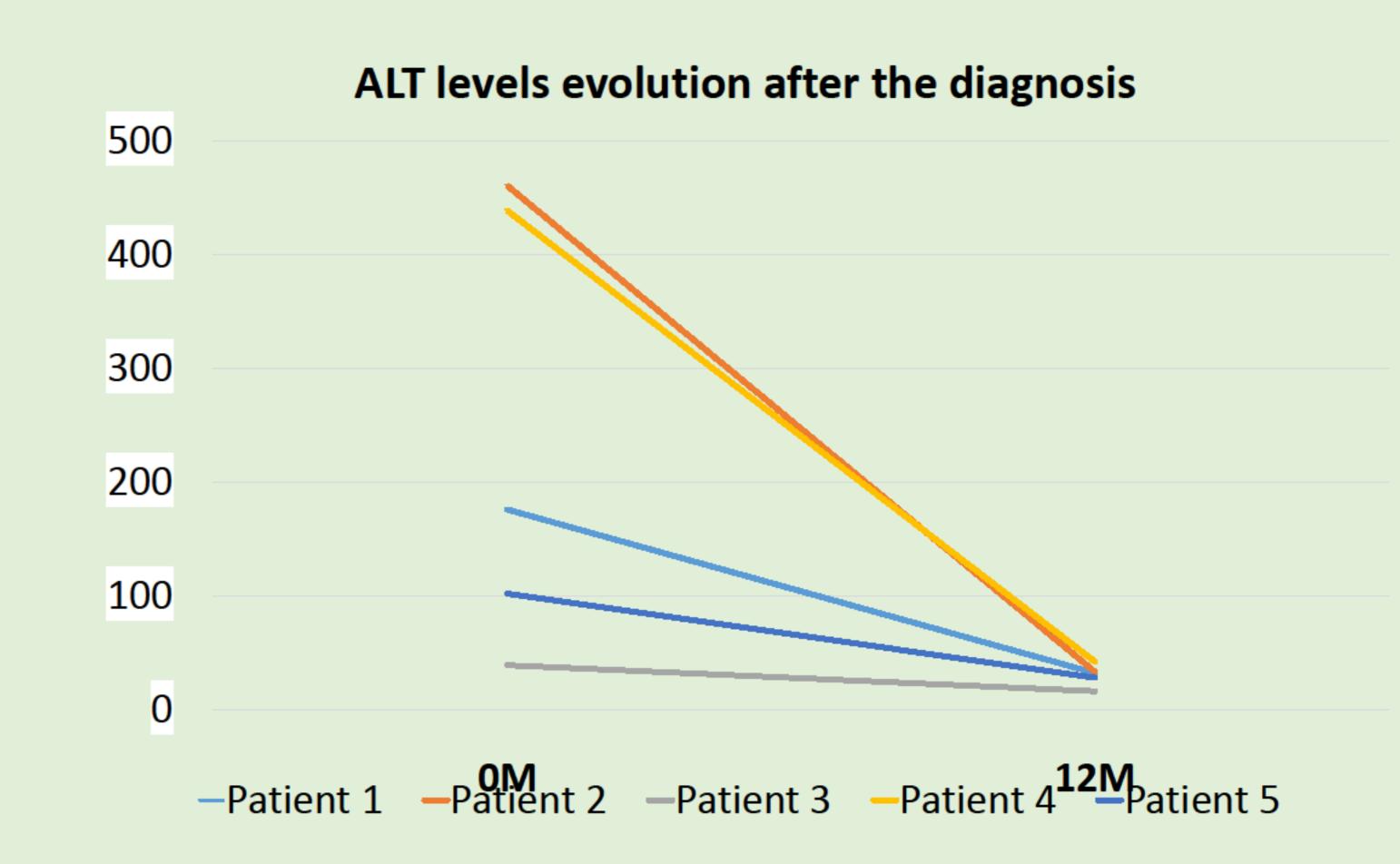
Table 1. Baseline characteristics of the study population.

All 5 patients underwent **liver biopsy** to clarify the alterations
on the enzymes of hepatic
cytolysis

Histopathological features of Mauriac Syndrome:

- Nuclear glycogenization
- Cytoplasmatic ballooning
- Insulin therapy intensification
- Therapy education reinforcement

HbA1c evolution after the diagnosis 17 15 13 HbA1c 2,72 2,40 % Patient 1 —Patient 2 —Patient 3 —Patient 4 —Patient 5



CONCUSIONS

Mauriac Syndrome is rare and not readily diagnosed by most clinicians. The <u>improvement of metabolic control seems to lead to hepatic enzymes</u> normalization in these patients, reinforcing the importance of early intervention.

References: [1] S. Memed KP, P. Larsen, H. Kronenber. Williams Textbook of Endocrinology. Elsevier Saunders. (12th edition). [2] Mauriac P. Gros ventre, hépatomégalie, troubles de la croissance chez les enfants diabétiques, traités depuis plusieurs années par l'insuline. Gas Hebd de Soc Med de Bordeaux. 1930(26):page 402.



Poster presented at:







^{*}medioclavicular measures on ecography; # under the 3rd percentile on WHO growth charts; + under the 10th percentile on WHO growth charts