

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

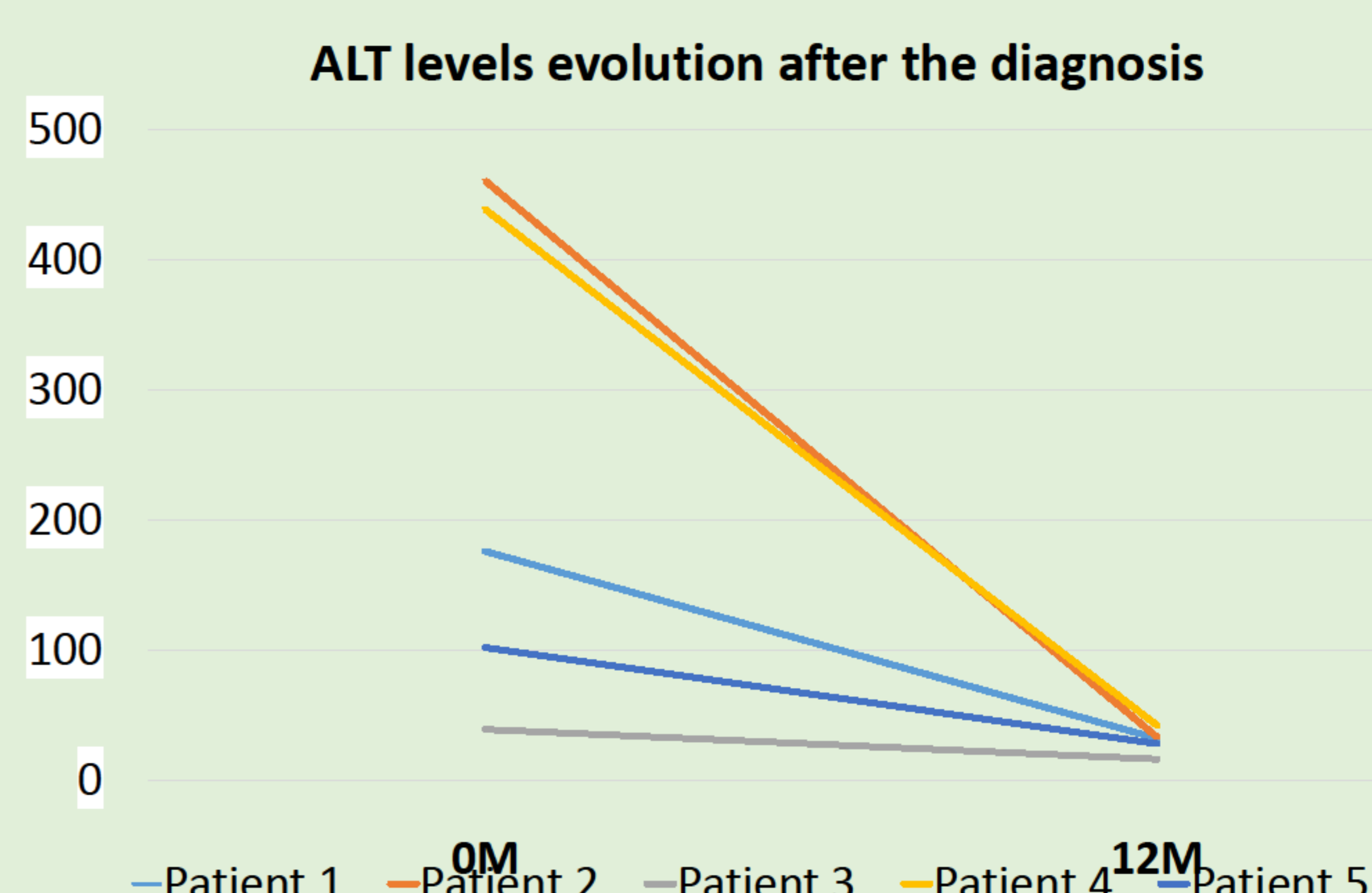
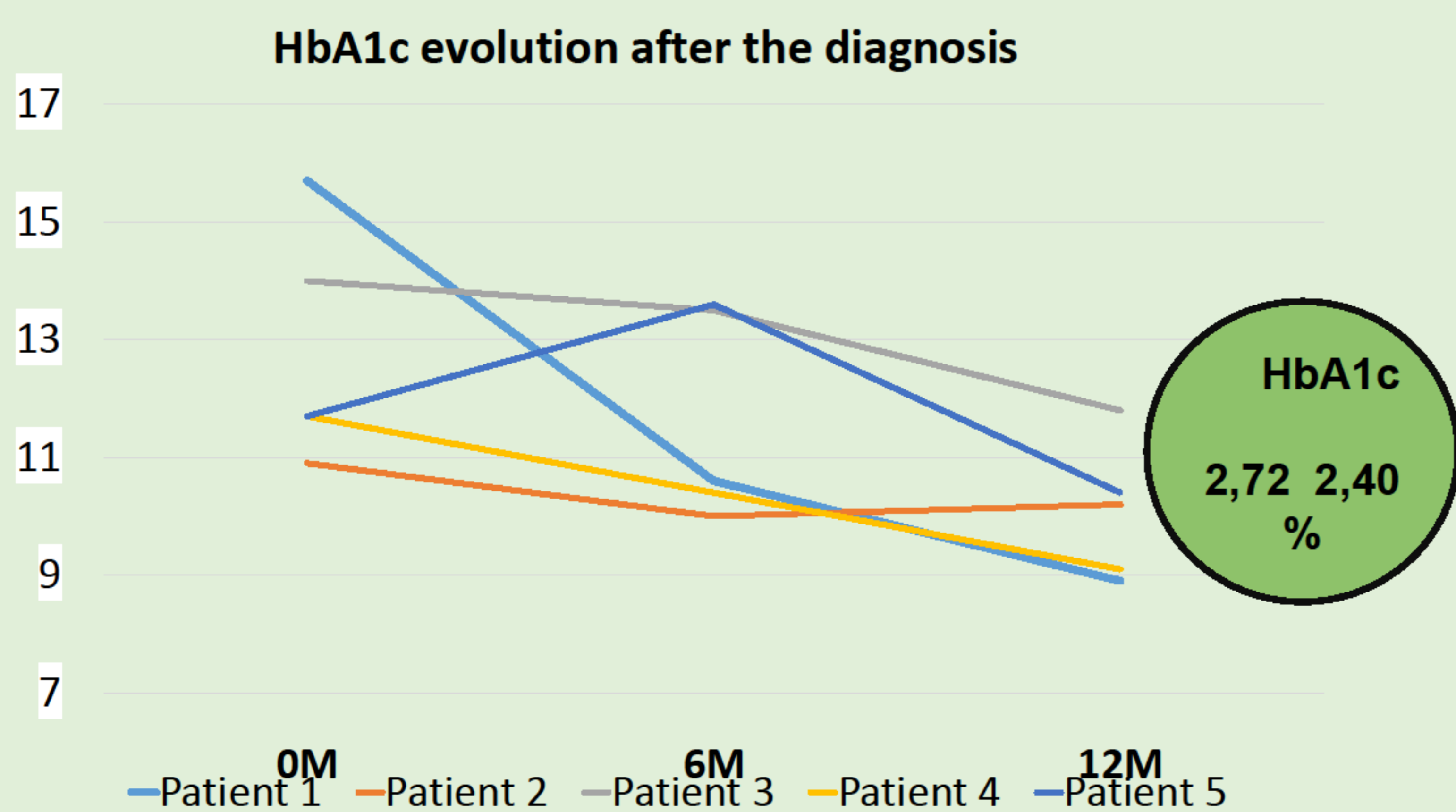
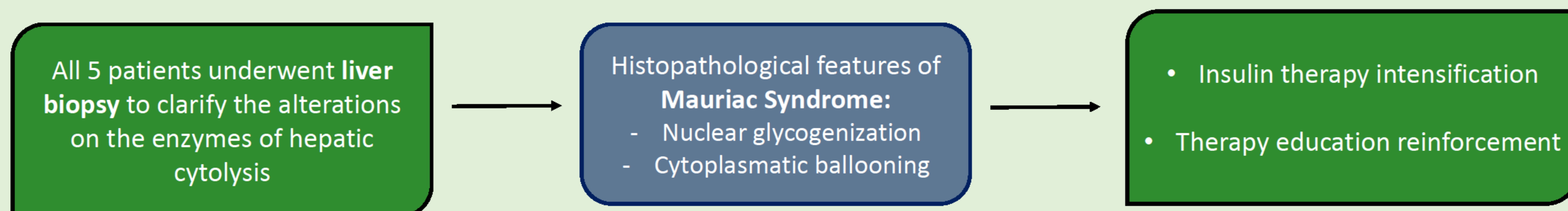
Retrospective study of **five patients** with T1DM with hepatic histopathology indicative of **Mauriac Syndrome**.

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	M	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.

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



CONCLUSIONS

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