# Mauriac Syndrome; A Rare Complication of T1 DM

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## Introduction

Mauriac syndrome (MS) is characterized by excessive intrahepatic glycogen deposition, attributed to severe hyperglycemia which diffuses by insulin-independent GLUT 2 to be trapped inside hepatocytes as G -6-phosphate.

It can rarely occur in poorly controlled T1 diabetic adolescents and is complicated by hepatomegaly, short stature, delayed puberty and hypertriglyceridemia

## Case report

17 years old T1 diabetic female patient since the age of 9 on insulin glargine and prandial regular insulin, not compliant on treatment with frequent DKA, presented by short stature and delayed puberty ( No menarche, thelarche or pubarche ), she also developed marked abdominal distension with no pain or jaundice.

## Physical examination

Height was 133 cm, <3rd percentile, proportionate. BMI was 19.7 kg/m2. Normal vital signs, (breast & pubic hair: tanner stage 1). The abdominal examination showed soft hepatomegaly. liver span of 17.5 cm Cardiac, and chest examination were normal. no evidence of diabetic retinopathy or neuropathy.

## Investigations

HbA1c was 9.6%, A/C ratio: 25 mg/g His hepatic panel revealed high liver transaminases. ALT: 325 U/l(10-50 U/l), AST: 1245 U/l (N: 0-38 U/l) cholesterol: 360 mg/dl, LDL: 249 mg/dl, Serum triglycerides: 240 mg/dl , low Transferrin sat. : 10%, low corrected Ca : 8 mg/dl, low vit D : 9 ng/ml, , low PTH : 10 pg/ml , High TSH: 6.27 uU/ml with normal FT4 , LH: 0.28 mIU/ml (1.7-8.6 mIU/ml) FSH: 1.7 mIU/ml(N: 1.5-12.1) Estradiol: <5 pg/ml (2.5 -8.4) indicative of hypogonadotropic hypogonadism. ANA, anti-TTG IgA, Anti TPO were negative, HBsAg and HCV Ab: negative Abdominal ultrasound: Enlarged liver with uniform soft fatty echo pattern, span of right lobe 17.5 cm, thyroid US was normal bone age: 14 years, MRI sella was normal.

Follow-up labs after control of last attack of DKA and strict glycemic control: ALT: 25 U/l, AST: 21U/l

## Conclusion

However, it is rare, MS should be suspected in uncontrolled T1DM with hepatomegaly, growth retardation, and delayed puberty as proper management could correct the condition.

## References

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