

# A possible case of glycogenic hepatopathy in a schoolboy with poorly controlled type 1 diabetes mellitus

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## Abstract

Glycogenic hepatopathy (GH) is a rare condition that can occur in individuals with poorly controlled diabetes mellitus, particularly in those with type 1 diabetes mellitus (T1DM). This condition is characterised by hepatomegaly and elevated liver enzymes. GH is associated with the accumulation of glycogen in the liver, which is a result of uncontrolled hyperglycemia and excessive glycogen storage in liver cells. We present a case of a 15-year-old boy who has been diagnosed with T1DM at the age of 8 and has experienced recurrent episodes of diabetic ketoacidosis (DKA). In this specific instance, he was admitted for treatment of DKA and was also diagnosed with GH.

**Key words:** glycogenic hepatopathy, diabetes mellitus, diabetic ketoacidosis, hepatomegaly and elevated liver enzymes

## Introduction

Patients with type 1 diabetes mellitus frequently present with abnormal liver function reports, indicating abnormal glycogen deposition, nonalcoholic fatty liver disease (NAFLD), fibrosis, and cirrhosis.(1) Because NAFLD is the most common aetiology for abnormal liver function, the rare presentation of Glycogenic Hepatopathy is often underdiagnosed.(2) GH is associated with the accumulation of glycogen in the liver, which is a result of uncontrolled hyperglycemia and excessive glycogen storage in liver cells.(3) The diagnosis of GH is very important because it can be reversed with good glycemic control.(4) The gold standard diagnostic investigation is confirming glycogen deposition in hepatocytes through periodic acid Schiff (PAS) staining in a liver biopsy. GH does not progress to cirrhosis and has a good prognosis if diagnosed early and treated. In this report, we discuss the diagnostic process of GH in our patient.

## Case presentation

A 15-year-old schoolboy, who has been known to have T1DM for 7 years and has a recurrent history of admission for DKA, with his last admission occurring 4 months ago due to poor adherence to insulin therapy during schooling, presented with abdominal pain, vomiting, and generalised weakness for a duration of 2 days. He did not have a history of fever, consumption of ayurvedic or herbal preparations and a history of previous blood transfusion. His usual insulin regimen included 6 units of soluble insulin with three meals and 12 units of glargine insulin at bedtime at 10 p.m. His daily insulin intake was 30 units over 24 hours, which corresponds to 0.85 units per kilogram per day. He does not have any history of previous liver disease, and there are no significant liver diseases in his family. He is not an alcoholic or smoker.

On arrival at the hospital, he was conscious and

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rational, with a blood pressure of 110/70 mmHg, a heart rate of 120 beats per minute, oxygen saturation of 98% in room air, and a respiratory rate of 22 breaths per minute. He appeared dehydrated and unwell. He was not pale or icteric. His abdominal examination revealed no tenderness, ascites, or splenomegaly but noted mild hepatomegaly. His initial investigations showed glucose levels at 511 mg/dL (normal range: 70-90 mg/dL), sodium at 134 mmol/L (normal range: 136-145 mmol/L), potassium at 4.0 mmol/L (normal range: 3.5-5.1 mmol/L), arterial blood gas results indicating pH at 7.373, pCO<sub>2</sub> at 22.8 mmHg, pO<sub>2</sub> at 98mmHg, HCO<sub>3</sub> at 13.39 mmol/L, lactate at 2.4 mmol/L. Ketones were noted to be positive. He was initially treated for DKA with fluid replacement and an intravenous soluble insulin infusion, with further investigations in progress.

Further investigations, as outlined in table 1, were conducted during his ward stay while he was being treated for DKA. Evaluation for hepatomegaly revealed elevated liver enzymes, gamma glutamyl transferase, and alkaline phosphatase levels, prompting further evaluation. Screening for acute hepatitis, including hepatitis A antibody, hepatitis B surface antigen, and hepatitis C antibodies, returned negative results. His ultrasound scan revealed a liver size of 17.9 cm with uniform echogenicity and no ultrasonic evidence of hepatitis or chronic liver cell disease. Over the days during his ward stay, his liver enzymes drastically increased. However, with proper tight sugar control, they began to decrease (figure 1).

After discharge, we reviewed him with repeated liver enzyme reports and repeated ultrasound scans. The results showed a resolution of elevated liver enzymes and hepatomegaly.

## Discussion

GH is commonly reported with T1DM but can also be associated with type 2 diabetes mellitus.(5) It often occurs in individuals who are treated for frequent DKA with high doses of insulin because it alters the balance between glycogenesis and glycolysis, leading to the accumulation of glycogen in hepatocytes. It commonly presents with hepatomegaly and elevated liver enzymes with preserved liver synthetic functions, as indicated by normal serum albumin and INR levels. Altered liver enzymes necessitate further evaluation to exclude other possibilities such as NAFLD, viral hepatitis, and alcoholic hepatitis.

In our patient, he is not an alcoholic, and his serologies for viral hepatitis revealed negative results. To differentiate further from NAFLD, we conducted an ultrasound scan of the abdomen. Typically, ultrasound shows normal echogenicity with hepatomegaly, whereas NAFLD would show evidence of steatosis or cirrhosis. In our patient, his liver displayed hepatomegaly with normal echogenicity.

Liver biopsy is the gold standard investigation to diagnose GH. It reveals histological evidence of glycogen deposition with pale, swollen hepatocytes

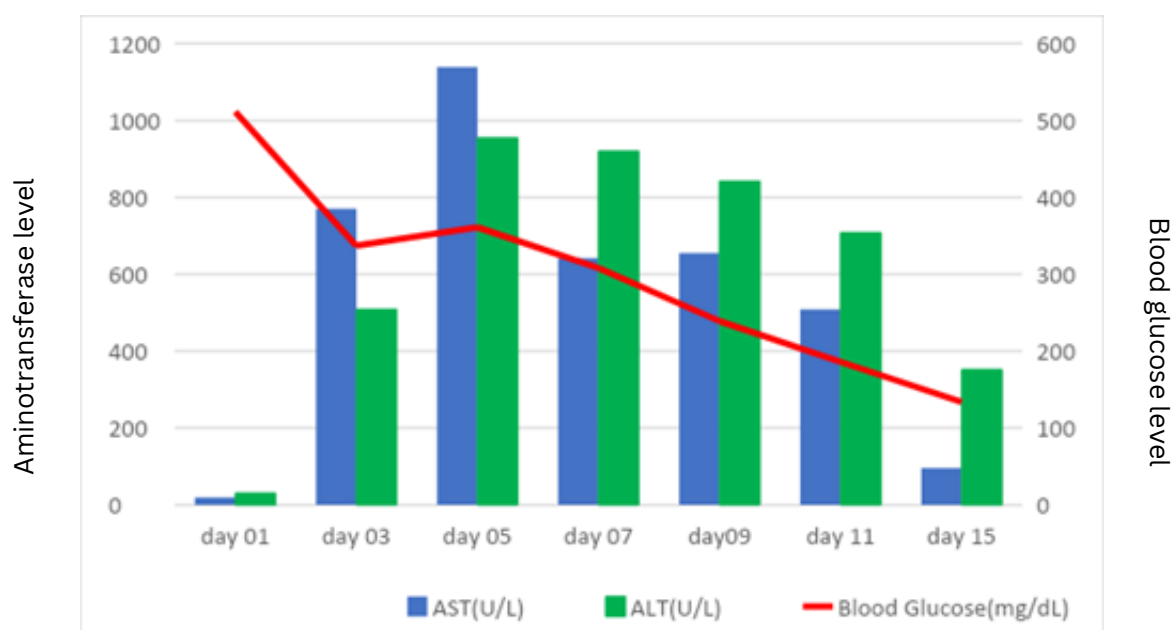


Figure 1 - Association between aminotransferases and blood glucose levels

**Table 1** - Summary of investigations

Investigation	Results				Reference range
	Day 1	Day 3	Day 6	Day 9	
WBC (x10 <sup>9</sup> /L)	9.77	9.52	5.84		4.5-11
Haemoglobin (g/dL)	14.1	13.5	12.6		11-13
Platelets (x10 <sup>9</sup> /L)	369	407	423		150-400
AST (U/L)	19.5	770	1139	509	10-35
ALT (U/L)	31	509	955	708	12-35
ALP (U/L)		732	366	391	300-500
Gamma GT (U/L)		385	630	639	5-40
Total bilirubin (mmol/L)		6.0	6.5	4.0	4-18
Total protein (g/L)		62	66	67	72-83
Serum albumin (g/L)			29	31	34-50
PT (seconds)		14	14.2		10-13
INR		1.26	1.04		<1.0
Serum creatinine (mg/dL)	0.9	1.12	0.53		<1.2
Blood urea (mg/dL)	5.5	8.7	3.3		5-20
Sodium (mmol/L)	134	139	138		136-145
Potassium (mmol/L)	4.0	3.52	5.2		3.5-5.1
CRP (mg/dL)	2.0	0.1			0-5
Blood culture		No growth			
Urine culture		No growth			
UFR albumin		Trace			NIL
UFR pus cells		6-8			NIL
UFR red cells		Nil			NIL

**WBC**- white blood cell, **AST**- aspartate transaminase, **ALT** - alanine transaminase, **ALP**- alkaline phosphatase, **gamma GT**- gamma glutamyl transferase, **PT**- prothrombin time, **INR**- international normalised ratio, **CRP**- c-reactive protein, **UFR** urine full report

and confirms glycogen deposition by PAS staining.<sup>(1)</sup> However, based on the clinical presentation, serology reports, and ultrasound scan findings, we were able to make the diagnosis of GH in the context of limited resources.

Our patient is involved in sports and hence leaves for school early in the morning, and stays at school for practice after regular school hours, often returning home late. This demanding schedule caused him to miss frequent doses of insulin, ultimately leading to frequent DKA admissions. Frequent DKA admissions and exogenous insulin therapy triggered GH in our patient.

The mainstay of treatment is tight sugar control, which leads to clinical and biochemical improvement. However, it can recur with poor glycemic control if not managed properly. Therefore, the mainstay of prevention is good adherence to treatment and regular monitoring of blood sugar levels.

## Conclusion

When treating T1DM patients, we should be aware of the reversible, rare cause for elevated aminotransferases such as GH because timely diagnosis and treatment with proper sugar control can improve the disease. Although liver biopsy is the gold standard test, diagnosis can also be made using clinical, biochemical and imaging parameters.

## Declarations

### Conflicts of interest

The authors declare that they have no conflicts of interest to be addressed regarding this case report.

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