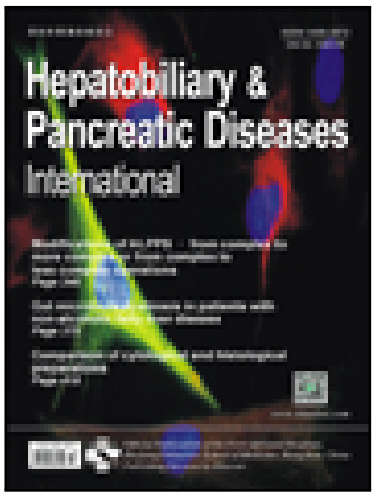

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Review Article
Running title: Glycogenic hepatopathy

Glycogenic hepatopathy

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BACKGROUND: Glycogenic hepatopathy (GH) is a disorder associated with uncontrolled diabetes mellitus, most commonly type 1, expressed as right upper quadrant abdominal pain, hepatomegaly and increased liver enzymes. The diagnosis may be difficult, because laboratory and imaging tests are not pathognomonic. Although GH may be suggested based on clinical presentation and imaging studies, the gold standard for diagnosis is a liver biopsy, showing a significant accumulation of glycogen within the hepatocytes. GH may be diagnosed also after elevated liver enzymes in routine blood tests. GH usually regresses after tight glycemic control. Progression to end-stage liver disease has never been reported. This review aims to increase the awareness to this disease, to suggest a pathway for investigation that may reduce the use of unnecessary tests, especially invasive ones.

DATA SOURCES: A PubMed database search (up to July 1, 2017) was done with the words "glycogenic hepatopathy", "hepatic glycogenosis", "liver glycogenosis" and "diabetes mellitus-associated glycogen storage hepatopathy". Articles in which diabetes mellitus-associated liver glycogen accumulation was described were included in this review.

RESULTS: A total of 47 articles were found, describing 126 patients with GH. Hepatocellular disturbance was more profound than cholestatic disturbance. No synthetic failure was reported.

CONCLUSIONS: GH may be diagnosed conservatively, based on corroborating medical history, physical examination, laboratory tests, imaging studies and response to treatment, even without liver biopsy. In case of doubt about the diagnosis or lack of clinical response to treatment, a liver biopsy may be considered. There is no role for noninvasive tests like fibroscan or fibrotest for the diagnosis of GH or for differentiation of this situation from nonalcoholic fatty liver disease.