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Absolute quantification of the hepatic glycogen content in a patient with glycogen storage disease by 13C magnetic resonance spectroscopy

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Using natural-abundance 13C magnetic resonance spectroscopy (MRS) on a conventional whole-body system operating at 1.5 T, the absolute hepatic glycogen concentration was noninvasively determined in a patient with type Ia glycogen storage disease. Furthermore, to assess the reliability of glycogen determination, hepatic glycogen content was assessed after an overnight fasting period in 35 healthy volunteers divided into two groups, one with a carbohydrate-rich diet, the other without any particular dietary preparation. In the patient, the glycogen concentration was found to be 458 mM. In the healthy subjects, average glycogen concentrations were 229 +/- 34 mM (mean +/- standard deviation) and 257 +/- 31 mM for the group without and with dietary preparation, respectively. The 13C-MRS results are in agreement with those obtained by conventional liver biopsy. 13C MRS in natural abundance may thus serve as a straightforward, fast, and noninvasive tool for quantification of the liver glycogen content in patients.

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