□ LETTERS TO THE EDITOR □

Challenges in Screening for Hepatocellular Carcinoma in the Glycogen Storage Disease Type 1a Population

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To the Editor We read with interest the case report by Ochi et al. (1). The authors have provided excellent multi-modality imaging of a case of hepatocellular carcinoma (HCC) which arose in a patient with previously undiagnosed Glycogen Storage Disease (GSD) type 1a. We agree that the key roles of imaging in the GSD-1 population are identifying early-stage HCC and adenomas at risk for malignant degeneration.

It is well known that hepatocellular adenomas (HCA) are common in patients with GSD-1, and in rare instances patients develop HCC (2). However, differentiating HCC from benign HCA by imaging can be quite difficult (3). Thus, the challenge in caring for these patients is to provide a screening program which is adequately sensitive for detecting HCC, but simultaneously specific enough so that benign HCA are not mistaken for HCC. These needs must be further balanced against the costs of screening and the risks of radiation exposure. Of the many imaging techniques described by Ochi et al., ultrasound (US), contrast-enhanced ultrasound (CEUS), and magnetic resonance imaging (MRI) do not use ionizing radiation.

In our clinical experience, HCA arising in patients with GSD-1 are often small, numerous, widely distributed, and nonspecific in sonographic appearance. Routine CEUS evaluation may prove challenging, since ultrasound contrast agents disperse during imaging, and often only a single lesion can be characterized with each contrast bolus. Therefore many patients require repeated contrast boluses in order

to adequately characterize their lesions, with associated increases in imaging time and cost. Additionally, small lesions located deep within the liver may be challenging to characterize due to difficulties obtaining adequate acoustic windows and sonographic penetration.

We advocate the use of contrast-enhanced MRI for both screening and diagnosis of these lesions. While initially more expensive, a contrast-enhanced MRI is much more rapid and in total less costly than initial screening with conventional US followed by CEUS of all candidate lesions, which may involve extended or multiple patient visits to the imaging center. MRI is also less subject to operator-dependent variability in the detection and measurement of lesions than US. The ultimate challenge is to identify high-risk adenomas and early HCC when they are resectable, since advanced stage lesions appear to be uniformly fatal (4). However at the current time, no imaging modality can reliably distinguish benign adenoma from premalignant adenoma or HCC in this setting, and the final diagnosis can only be made pathologically.

The authors state that they have no Conflict of Interest (COI).

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