

derangements at the initiation of screening confirms that we are initiating imaging too late. Therefore, periodic imaging should be started in childhood or early adolescence.⁵ The ideal screening test is gadolinium-enhanced magnetic resonance imaging (with a sensitivity and a specificity of 91% and 95%, respectively, to detect HCC)² at intervals of 3 to 4 years. In patients with contraindications to magnetic resonance, a computed tomography scan may be performed, although the risk of radiation exposure must be considered. Meanwhile, liver stiffness estimated by US and magnetic resonance elastography techniques may be used as a quantitative imaging biomarker for the detection, staging, characterization, and monitoring of liver fibrosis. However, the use of elastography in Fontan-associated liver disease is problematic because any altered hepatic stiffness beyond fibrosis, particularly the vascular congestion universally present in Fontan patients, may have an impact on the results.

For all these reasons, some authors recommend that patients with a Fontan procedure performed more than 10 years previously should undergo cardiac assessment, liver imaging, and even liver biopsy⁴ to stay ahead of neoplastic transformation. Even after heart transplant, patients who have undergone the Fontan procedure will require vigilant screening for HCC.

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Available online 18 January 2017

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1885-5857/

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Liver Imaging in Patients With Fontan Circulation. Response



Imagen hepática de pacientes con cirugía de Fontan. Respuesta

To the Editor,

Having read with interest the letter published by Martínez-Quintana et al. concerning my publication,¹ I would like to add some comments on liver imaging findings after Fontan surgery.

First, ultrasonography is the most commonly used imaging technique for initial and follow-up liver evaluation in these children, mainly due to the lack of radiation. Hepatic parenchymal changes after the procedure, known as Fontan-associated liver disease, include liver fibrosis and cirrhosis and hepatocellular carcinoma. Although ultrasonography usually detects late changes of fibrosis and cirrhosis (such as heterogeneous parenchymal echotexture or surface nodularity), recent publications show that other findings, such as hyperechoic lesions without surface nodularity detected by high frequency transducer, may represent the early stage of fibrosis.² These lesions were not demonstrated by computed tomography or magnetic resonance imaging and most patients (82%) showed normal biochemical hepatic function tests, despite the presence of hepatic parenchymal changes on imaging.²

Second, recent studies suggest that, considering that congestion is the primary or sole trigger of liver fibrosis in these patients, ultrasound elastography may eventually become a useful noninvasive, low-cost proxy assessment of Fontan hemodynamics and a clinical means of determining which patients are at highest risk of fibrosis development.³ Moreover, magnetic resonance elastography might prove particularly useful to evaluate progression of liver disease and have important prognostic value.⁴ Furthermore, some

authors conclude that magnetic resonance elastography allows earlier detection of fibrogenesis than biomarkers.⁵

Finally, with regard to contrast computed tomography and magnetic resonance imaging, heterogeneous enhancement is a common finding in cirrhotic liver. Hypervascular liver nodules are an additional important finding in patients with longstanding Fontan circulation (20%–30%), also known as focal nodular hyperplasia-like lesions.⁴ The main differential diagnosis of hypervascular nodules in a cirrhotic liver should be hepatocellular carcinoma but there are few reported cases of hepatocellular carcinoma in these patients.⁴ In fact, according to a recent publication, there are only 11 case reports of hepatocellular carcinoma after the Fontan procedure in PubMed.⁶

In conclusion, although ultrasonography and laboratory screening at regular intervals should be the first-line tests in the long-term evaluation of these patients, elastography and contrast studies are useful tools that should also be considered in follow-up.

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Available online 21 February 2017

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