

Evidence Discussion – Monitoring After Fontan Procedure (AB-26.4)

Individuals with single-ventricle physiology who have undergone the Fontan Procedure which redirects venous blood flow to the pulmonary circulation invariably develop liver complications, which can include the development of nodules and cirrhosis secondary to the altered vascular anatomy, and thus are at risk for hepatocellular carcinoma. In addition, the congestive hepatopathy associated with the Fontan procedure makes differentiation of focal liver lesions from congestive changes more challenging than other cirrhotic conditions. Thus, most institutions use MRI rather than US for monitoring in the setting of cirrhosis. In addition, the evaluation for HCC is challenging due to the vascular changes associated with the Fontan procedure, because the typical HCC pattern of delayed venous-phase contrast washout may not be appreciated within the background congestive hepatopathy. Thus, biopsy is usually required. Also, distinguishing dysplastic lesions from true HCC based on LiRADS criteria is very challenging as well. There are no current society endorsed guidelines, and institutions may vary in the monitoring of chronic liver disease in this patient population. The above algorithm represents an accepted approach and is consistent with the consensus from the Fontan-Associated Liver Disease proceedings from the American College of Cardiology Shareholders Meeting (2015) as well as the consensus of a multidisciplinary group of American Society of Transplantation members (2020).

Also see evidence discussion for AB-26.1.

References:

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