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Abdominal Imaging Surveillance in Adult Patients After Fontan Procedure: Risk of Chronic Liver Disease and Hepatocellular Carcinoma☆

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A B S T R A C T

Purpose: To assess the prevalence of chronic liver disease (CLD) and hepatocellular carcinoma (HCC) in adult patients who had surveillance imaging after Fontan procedure.

Methods: Institutional review board–approved, Health Insurance Portability and Accountability Act–compliant retrospective study evaluated electronic medical records including radiology reports and clinical notes for adult patients after Fontan procedure between January 1993 and January 2016. Abdominal ultrasound, computed tomography, and magnetic resonance imaging reports were reviewed for changes of CLD and HCC. Existence of concomitant viral hepatitis was also recorded.

Results: A total of 145 patients (male: 78 and female: 67) had surveillance imaging after Fontan procedure. In all, 78% (113/145) had ongoing imaging surveillance (median follow-up 3.05 years, IQR: 0.75-5.3 years); 19% (21/113) had an initial normal study and remained normal throughout follow-up; 19% (21/113) had an initial normal study with subsequent imaging reporting changes of CLD; and 62% (71/113) had existing changes of CLD on initial study. HCC was identified in 5 patients (median 22 years post-Fontan, IQR: 10-29 years), 4 of which had a normal initial study. Only 1 patient with HCC had concomitant viral hepatitis C infection.

Conclusion: Radiologists should be aware that CLD is exceedingly common in post-Fontan cardiac physiology, and surveillance imaging is warranted given the risk of HCC.

Introduction

Because of advanced surgical techniques and improved health care delivery, many patients with congenital heart disease (CHD) are now surviving well beyond childhood into their adulthood.[1] In particular, the Fontan procedure has been shown to prolong life expectancy in patients born with a single cardiac ventricle who cannot undergo biventricular repair.[2] Unfortunately, these patients are prone to developing cardiac-related hepatic issues as they age, presumably due to postsinusoidal increased venous pressure in the liver from post-Fontan cardiac physiology that can result in chronic hepatic congestion, chronic liver disease (CLD), portal hypertension, and even hepatocellular carcinoma (HCC).[3-7]

Estimating the true prevalence of CLD among patients who underwent a Fontan procedure remains a major challenge. Although hepatic biopsies can accurately determine the presence and stage of CLD in patients after the Fontan procedure, biopsy is an invasive procedure with associated risks and is typically performed only in symptomatic patients limiting its usefulness as a screening tool.[8] In addition, fibrosis may not be uniform within the liver, which may lead to sampling error and variability in the results of parenchymal biopsy. Therefore, noninvasive imaging methods, that is, ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI) may be considered viable options for the screening and detection of CLD and HCC in the post-Fontan population.[9-12]

To our knowledge, there is sparse literature evaluating the actual prevalence of CLD in the adult post-Fontan population detected on surveillance imaging.[3,13,14] In addition, only a few case reports or case series describe the presence of HCC in the post-Fontan patient population.[4,5,15-17] Therefore, radiologists may not be aware that adult patients with CHD who underwent a Fontan procedure should be carefully screened for CLD and its sequelae, including the presence of HCC.

As a large tertiary care center with a robust cardiac center, our institution sees a moderate cohort of adult patients who underwent a Fontan procedure. These patients typically undergo routine

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abdominal imaging surveillance with US, CT, or MRI or a combination of modalities to monitor the development of CLD and HCC. The purpose of this retrospective study is to assess the presence of CLD and HCC in this patient population as detected by imaging surveillance.

Materials and Methods

Patient Selection

Institutional review board approval was obtained and a waiver of informed consent was granted for this Health Insurance Portability and Accountability Act–compliant retrospective study. Our department's cardiology patient electronic medical records were searched to identify patients with CHD who underwent a Fontan procedure. Subsequently, the radiology picture archive communications system was searched to identify patients who had abdominal radiological imaging follow-up after Fontan procedure for the clinical indication of CLD surveillance between January 1, 1993 and January 1, 2016. Patients younger than 18 years or those without abdominal imaging reports available for review were excluded.

Data Collection

The electronic medical record was searched for each adult patients with CHD who underwent a Fontan procedure with abdominal imaging (US, CT, or MRI) results available at our institution. Demographic data including sex, age at time of Fontan procedure, and age at time of initial abdominal imaging study were recorded. Dates and modality of additional abdominal imaging studies available for each patient were also recorded. All abdominal MRI included noncontrast and postcontrast multiphasic sequences including arterial, portal venous, and delayed phases. Abdominal imaging reports of initial study and any follow-up abdominal imaging studies were reviewed for the mention of hepatic abnormalities including early or late CLD, portal hypertension, or HCC. Relevant findings were stratified into 3 categories —*normal*: no radiographic findings of CLD, portal hypertension, or HCC; *changes of CLD*: coarsened or heterogenous echotexture (US), lobar redistribution (CT/MRI), evidence of fibrosis (CT/MRI), or evidence of portal hypertension such as splenomegaly or ascites due to CLD (US, CT/MRI), and varices (CT/MRI) without HCC; and *presence of HCC*. The presence of HCC was determined by classic imaging features of HCC of precontrast T1-hypointense signal with arterial enhancement and washout.[18] The clinical records of patients found to have HCC by imaging were also assessed for supporting tumor markers (increased alpha fetoprotein levels) and subsequent treatment regimens including radiofrequency ablation or chemoembolization on future imaging. All patients were also assessed for the presence of concomitant viral hepatitis infection such as hepatitis C.

Statistical Analysis

Statistical analysis included calculation of median (MDN) and interquartile range (IQR) of time between Fontan procedure and initial abdominal imaging study as well as length of abdominal imaging follow-up. Prevalence of normal, changes of CLD, and the presence of HCC on initial imaging study was recorded as a percentage of the patient population. Similarly, percentage of patients who remained normal throughout follow-up period, who demonstrated changes of CLD, and who obtained a new diagnosis of HCC throughout the follow-up period was calculated.

Table 1
Initial imaging findings identified by modality type

Earliest imaging findings	Normal	CLD	HCC	Total
Modality				
US	51	64	0	115
	44.3%	55.7%	0%	100.0%
CT	1	10	0	11
	9.1%	90.9%	0%	100.0%
MRI	3	16	0	19
	15.8%	84.2%	0%	100.0%
Total	55	90	0	145
	37.9%	62.1%	0%	100.0%

Results

A total of 145 patients met the inclusion criteria. The median age was 27.0 years (IQR: 24-33.5 years), with slightly more males than females (M: 78, F: 67). Median age at Fontan procedure was 4 years (IQR: 2-6.75 years). Initial abdominal imaging study available at our institution was approximately 19 years (MDN) post-Fontan (IQR: 17-23). A total of 559 imaging studies were performed on 145 patients during the follow-up period. Median number of imaging studies per patient was 3 (IQR: 2-5).

On initial imaging study, 38% (55/145) had normal imaging and 62% (90/145) demonstrated changes of CLD including liver heterogeneity, volume redistribution, portal hypertension, and cirrhosis (Table 1). None of the patients had imaging findings of HCC on initial study. US was the most common imaging modality (115/145) at initial imaging, followed by MRI (19/145) and CT (11/145). On initial US, 44% (51/115) had normal findings whereas the rest showed features of CLD. On initial MRI and CT, the majority showed features of CLD (16/19 and 10/11, respectively) and only 4 cases were normal (Table 1).

In all, 22% of patients (32/145) did not receive any further follow-up imaging. Table 2 describes the initial imaging findings of these patients; 78% (113/145) had ongoing imaging surveillance with a MDN follow-up of 3.05 years, IQR of 0.75-5.3 years; 42% of patients (47/113) were followed up with a single type of imaging modality (US = 42, MRI = 5); 48% of patients (54/113) had follow-up imaging with 2 modalities (US/CT = 21, US/MR = 32, CT/MRI = 1], whereas 11% of patients (12/113) were scanned with all the 3 imaging modalities (US/CT/MRI) during surveillance.

Of the patients with follow-up imaging, 19% (21/113) had an initial normal study and remained normal throughout follow-up regardless of imaging modality. In comparison, 19% (21/113) demonstrated an initial normal study with subsequent imaging studies documenting new imaging findings of CLD with 4 of these patients obtaining a new diagnosis of HCC on subsequent imaging. A total of 62% (71/113) had existing changes of CLD on initial imaging and one of these patients was diagnosed with HCC on subsequent imaging. Of note, 83% (94/113) of patients were negative for hepatitis C, 12% (13/113) were untested, and 5% (6/113) had hepatitis C viral infection.

Table 2
Initial imaging findings of patients without imaging follow-up

Imaging findings	*N*	%
Normal	13	40.6
CLD	19	59.4
HCC	0	0
Total	32	100.0