A liver mass post-Fontan operation

N. RAJORIYA1, P. CLIFT2, S. THORNE2, G.M. HIRSCHFIELD1,3 and J.W. FERGUSON1

From the 1The Liver Unit, The New Queen Elizabeth Hospital, Birmingham B15 2TH, 2Department of Cardiology, The New Queen Elizabeth Hospital, Birmingham B15 2TH and 3NIHR Biomedical Research Unit and Centre for Liver Research, University of Birmingham, Birmingham, UK

Address correspondence to Dr N. Rajoriya, The Liver Unit, The New Queen Elizabeth Hospital, Birmingham B15 2TH, UK. email: neil.rajoriya@gmail.com

Learning Point for Clinicians

This case report raises awareness of a rare devastating complication of congenital heart diseases that require correction via the “Fontan” operation. In these patients, liver cirrhosis and its complications are now being recognised including hepatocellular carcinoma. With recognition, strategies for early treatment/interventions in a complicated population can be devised.

Case

A 19-year-old female was referred for urgent investigation of jaundice. She had a history of complex congenital heart disease with situs inversus, left atrial isomerism and juxtaposed atrial appendages, azygous continuation of the inferior vena cava and malposition of the great arteries with tricuspid atresia. In 1990, she had a successful atrio-pulmonary connection (‘Modified Fontan’ operation). She was followed up closely thereafter by the Cardiology team.

Eight years later, she had an oesophageal variceal haemorrhage requiring variceal band ligation. At the time, it was felt that her portal hypertension was related to her cardiac history with other causes of liver disease excluded. She remained on endoscopic surveillance with 2-yearly upper gastrointestinal endoscopies thereafter.

Discussion

Children with congenital cardiac defects lacking two effective pumping ventricles (e.g. tricuspid/pulmonary atresia, hypoplastic left or right heart syndrome) require operations to have this corrected eponymously called ‘Fontan operations’. After a Fontan procedure the systemic venous return is connected to the pulmonary arteries without the interposition of an adequate ventricle, and all shunts on the venous, atrial, ventricular and arterial level are interrupted. Advantages of this new circulation include improved systemic arterial saturations and reduction of the chronic volume overload on the single ventricle. The consequence of this operation is chronic systemic venous hypertension and a reduction of cardiac output both at rest and during exercise.

It has been postulated that as a result of the elevated venous pressure along with reduction in...
cardiac output, there will be a reduction in portal venous blood flow along with reduction of portal venous saturation. This in turn may lead to a dependence on the liver of the hepatic arterial buffer response (HABR) whereby the autoregulation mechanism leads to increase blood provision to the liver via the hepatic artery. Long-term HABR in the Fontan patient has been proposed to potentially play a role in the organ damage.\(^3\) In patients with late Fontan circulations, chronic venous congestion along with low cardiac output and chronic hypoxaemia (exaggerated in times of cardiovascular stress) may lead to hepatic inflammation and subsequent fibrosis and cirrhosis. Fontan-associated liver disease (FALD) is now recognized, with clinicians attempting to ascertain which patients will progress to fibrosis, develop portal hypertension and cirrhosis. With the development of cirrhosis comes the risk of HCC in a relatively young cohort of patients.

Ghaferi and Hutchins\(^4\) described in autopsy findings four patients surviving between 4 and 18 years post-Fontan operation with cirrhosis. In these, one had a hepatic adenoma (surviving 9 years) and another had an HCC (surviving to 18 years). Saliba \textit{et al.}\(^5\) described the occurrence of HCC in two patients with Fontan operations—one 18 another 22 years after operation. Neither was in an HCC-screening programme. Asrani \textit{et al.}\(^6\) described a series of four Fontan patients from 24 to 42 years of age developing HCC, thus the ages of development and time frame for development of HCC remain unknown but the process may take some time raising the question of HCC screening, and also awareness to diagnose patients with FALD and also those who go onto develop cirrhosis.

In conclusion, with the recognized potential development of cirrhosis in the Fontan cohort of patients, awareness of the inherent risk of HCC should be raised. This potentially may mean a new group of patients who indeed present to the adult medical teams with devastating complications of liver cirrhosis.

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\textbf{References}