

Fontan circulation: beyond cardiopulmonary assessment

Circulația Fontan: dincolo de evaluarea cardiopulmonară

Dear Editor

The Fontan procedure is today the last staged operation for children with congenital heart disease who cannot be offered a 2-ventricle repair, and it refers to any operation that results in the flow of systemic venous blood to the lungs without passing through a ventricle⁽¹⁾. Therefore, pathophysiology of the Fontan circulation leads to an obligatory systemic venous hypertension in which the superior and inferior vena cava and the pulmonary artery pressure are equal and generally in the range of 10 to 20 mmHg, 2 to 4 times that of normal.

Liver histology of patients with Fontan circulation usually begins with sinusoidal dilatation, parenchymal atrophy, and progressive sinusoidal collagen deposition and fibrosis in the perivenular distribution secondary to repetitive mechanical stretch and compression in the context of passive congestion and due to tissue hypoxia secondary to a low cardiac output. However, other factors such as systolic and a diastolic ventricular dysfunction, reduced basal oxygen saturation, hyperviscosity, shear stress or the release of vascular factors may promote hepatic changes in a not failing Fontan circulation⁽²⁾, and the development of ascites. Ascites, in such patients, is usually accompanied by a low hepatic vein wedge pressure and a low gradient between the wedged and the unwedged positions which could be in relation to the suboptimal flow dynamics seen in Fontan patients and the existence of congested hepatic sinusoids that act as “open tubes” favoring low transhepatic venous pressure gradients. These observations may be also responsible for the small number of Fontan patients with gastroesophageal variceal bleeding despite the hepatic involvement⁽⁶⁾. Meanwhile, a possible explanation for ascites is that the effect on the smallest portal veins and the sinusoids produces sufficient resistance at the sinusoidal level to favor ascites formation.

Cirrhosis may develop 11 to 15 years after a Fontan procedure and the incidence of cancer is of 1.5 to 5% per year on the basis of previous studies and the longer the time, the greater the risk of hepatic complications. Surveillance for hepatocellular carcinoma may be needed in such patients, especially if the alpha-fetoprotein level is elevated⁽³⁾. Also, the existence of dynamic contrast magnetic resonance imaging (MRI) exhibiting

typical arterial enhancement and venous/late washout is important as diagnosing hepatocellular carcinoma in patients with congenital heart disease is difficult because hyperenhancing nodules are often present and the typical arterial enhancing pattern of the hepatocellular carcinoma may not be obvious⁽⁴⁾. Finally, some authors recommend that patients who are >10 years out from their Fontan operation should undergo cardiac assessment, as well as a liver biopsy in order to stay ahead of neoplastic transformation⁽⁵⁾. Even after cardiac transplantation, patients who have undergone the Fontan procedure should require vigilant screening for hepatocellular carcinoma.

In short, Fontan patients, in addition to cardiopulmonary assessment to exclude dysfunction of the Fontan circulation, should be followed-up with analytical determination of liver enzymes and alpha-fetoprotein, abdominal ultrasound and liver MRI imaging and, if necessary, with liver biopsy to rule out the development of hepatocellular carcinoma.

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