

## The role of the hepatic elastography in the follow up of patients after Fontan surgery

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**Introduction:** Fontan circulation causes liver congestion with hypoxic stress and local inflammation, that leads to chronic liver damage and fibrosis. Hepatic elastography can non-invasively estimate the level of organ fibrosis, measuring the liver stiffness (LS).

**Methods:** In order to evaluate usefulness of hepatic elastography in the follow-up of patients with Fontan circulation, we prospectively examined patients with complex congenital heart disease (CHD) who underwent Fontan surgery or Kawashima procedure in our centre between 2012 and 2017. Hepatic elastography was performed at 3, 6, 12 months after surgery and yearly thereafter. Echocardiographic and blood test data were also collected.

**Results:** 16 patients were enrolled (M:F = 1), with mean age at surgery of 6.46 (range 4.0-10.0) years. Native CHD are listed in Table 1. 14 patients underwent extracardiac Fontan surgery (fenestrated in 3 cases), while 2 of them Kawashima procedure. No patient had any preexisting hepatic illness.

Mean age at last follow-up was 12.8 (5-29) years. Echocardiography showed good flow in the Fontan circuit in all patients and a normal systolic ventricular function at follow-up. No one suffered from heart failure nor "protein losing enteropathy" during follow-up.

Mean LS value was 12.35 kPa (range 2.8-22.3). LS was higher in patients who underwent Fontan surgery than Kawashima procedure. Moreover LS was higher at third month after surgery and decreased at 6 and 12 months. It increased again at 5 years. No relevant variations of LS were detected thereafter. Native CHD seemed to be a determinant of the stiffness at 5 years, instead, as right single-ventricle patients showed the highest LS.

Increased LS was linked to hepatomegaly diagnosed by physical examination, but no relevant alteration was found in liver function tests. INR was prolonged in two patients on warfarin and otherwise normal.

**Conclusion:** Hepatic Elastography is a potential tool to monitor the onset and progression of congestive hepatopathy and fibrosis. Patients who underwent Fontan surgery showed an increase of LS, likely dependent on native CHD, without a relevant alteration of hepatic function. Lacking data regarding liver biopsy, it is not possible to establish whether the increased LS is due to hepatic congestion or fibrosis.

Native Congenital Heart Disease	Number (%)
Tricuspid atresia	5 (31)
DORV, PS/PA	3 (19)
ccTGA, PA	3 (19)
VSD with overriding and straddling tricuspid valve	2 (13)
TGA, VSD, CoAo, overriding and straddling tricuspid valve	1 (6)
Left isomerism, complete AVCD	1 (6)
Right isomerism, AVCD with RV dominance	1 (6)

AVCD: atrioventricular canal defect; ccTGA: congenitally corrected transposition of great arteries; CoAo: coarctation of the aorta; DORV: double-outlet right ventricle; PA: pulmonary atresia; PS: pulmonary stenosis; TGA: transposition of great arteries; VSD: ventricular septal defect