Liver Fibrosis in Patients with Fontan Circulation Leads to Alteration in Portal Venous Hemodynamics

Hayashi T., Shindo T., Hirata Y., Shimizu N., Inuzuka R.
Department of Pediatrics, University of Tokyo Hospital, Tokyo, Japan

Introduction:
Inspiratory-to-expiratory flow rate ratio (Qin/Qex) of the portal vein (PV) increases in patients with functionally poorer Fontan circulation. This alteration in PV hemodynamics may lead to long-term gastrointestinal complications of Fontan circulation, including liver dysfunction and protein-losing enteropathy. Moreover, it is not uncommon for Fontan patients to develop liver fibrosis in the late postoperative period. Although it is hypothesized that liver fibrosis may further exacerbate PV hemodynamics by impeding splanchnic blood flow, the influence of liver fibrosis on PV hemodynamics has not been investigated. We aimed to evaluate the correlation between Qin/Qex of the PV and liver fibrosis.

Methods:
We studied 21 consecutive patients with Fontan circulation who underwent postoperative cardiac catheterization for various indications. Serum hyaluronic acid (HA) concentration was used as an indicator of liver fibrosis. Pulsed-wave Doppler recordings at the main portal trunk in the supine position was used to calculate the Qin/Qex; the median value was used to divide the patients into 2 groups.

Results:
The median age at examination was 5.3 years (range, 2.2–25.8 years), and the median time interval between Fontan procedure and examination was 2.0 years (range, 3 months to 19.7 years). Three patients were classified as NYHA functional class II, and the others as NYHA functional class I. The median Qin/Qex of the PV was 1.19 (range, 0.96–1.48). Patients in the high Qin/Qex group had significantly higher serum HA concentration than those in the low Qin/Qex group (median 27.1 [10–50.7] ng/mL vs. median 11.9 [10–25.2] ng/mL, respectively; p < 0.05). No significant difference was noted in laboratory data including total bilirubin, liver enzyme, brain natriuretic peptide levels, and platelet counts between both groups. Inferior vena cava pressure, superior vena cava pressure, systemic end-diastolic ventricular pressure, and Nakata index were similar for both groups.

Conclusions:
Fontan patients with higher Qin/Qex of the PV had significantly higher serum HA concentration. Liver fibrosis in Fontan patients leads to worsening of PV hemodynamics, which may exert further negative effects on Fontan circulation.