GASTROINTESTINAL COMPLICATIONS IN PATIENTS WITH A FONTAN CIRCUIT

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Background: Patients undergoing the Fontan procedure have been noted to develop gastrointestinal complications including congestive hepatopathy and cirrhosis. Screening for hepatic dysfunction in survivors of the Fontan operation has been recommended.

Methods: The electronic Adult Congenital and Pediatric Cardiac databases from 2 tertiary facilities serving a 3-state region were reviewed and all Fontan survivors > 18 years old were identified. Patients were followed in either a pediatric cardiology clinic or subspecialized ACHD clinic. Demographic, radiographic, laboratory, and surgical data were reviewed to determine whether screening for hepatic dysfunction during adolescence or adulthood was performed, the type of screening procedure used, and the results of such testing. Cirrhosis was noted to be present or absent based on the results of radiographic imaging or biopsy specimen. Surgical and clinical correlates of cirrhosis were sought.

Results: Routine testing for hepatic dysfunction was performed in 51 of 70 (73%) adult survivors of the Fontan procedure. Despite similar ages (27.3 v 26.2, p=0.9), patients followed in an ACHD rather than pediatric cardiology clinic were more likely to undergo testing (100 vs 12%, p<0.0001). At a mean age of 25.6 + 6.4 years, 26 of 51 (51%) patients had cirrhosis. A liver mass requiring intensified monitoring for hepatocellular carcinoma was present in 12 (23%). Abnormal transaminases were present in 15% of patients and did not correlate with presence of cirrhosis (p=0.13) while an elevated A2 macroglobulin did (77 v 22%, p=0.01). The presence of cirrhosis did not relate to time from Fontan procedure (20 + 5 v 19 + 5 yrs; p=0.9) or type of Fontan (p=0.31). Of the 16 patients (23%) who died or required cardiac transplant in adulthood, 7 (44%) died or were listed as a result of hepatic dysfunction.

Conclusions: Despite surgical advances, cirrhosis remains common in adults following Fontan procedure in childhood. Universal screening is not yet routine. The presence of normal transaminases is insufficient to rule out liver disease. Hepatic dysfunction is an important cause of mortality in these patients and efforts must be made to standardize screening.