Commentary: Just the tip of the iceberg…

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Central Message: Fontan-associated disease is an increasingly encountered and enigmatic problem.

Central Picture Legend: Manifestations of Fontan-associated disease.

The study by Kisamori et al is a reminder of the unsolved problems that are increasingly encountered in single ventricle palliation [1]. Cirrhosis, one of many markers of chronic Fontan-associated disease, is a bellwether of deeper and more insidious problems (Figure 1) [2].

The Fontan circulation can be defined as a circulation that lacks a subpulmonary pump. While Fontan palliation has offered decades of life for single ventricle patients, we are now confronted with its predicted long-term limitations [3]. So, what can we do?

Fontan circuit optimization is extremely important. An undersized extracardiac conduit (ECC) is functionally equivalent to IVC banding; graft size is critical as the patient grows into adulthood. Should we adopt a strategy of multiple Fontan re-interventions, i.e. planned “stage-4, -5” etc? If conduit upsizing is planned, when should it be performed, how durable will it be, and at what risk? The current debate regarding ECC vs lateral tunnel also factors heavily into this, with the pendulum shifting in favor of lateral tunnel [4].
Regardless how performed, we will remain saddled with an unsolved problem. No matter how effectively the Fontan circuit is optimized (ECC vs lateral tunnel, percutaneous IVC pathway expansion, and so on), it will always be associated with a power loss; it will never be associated with a power gain. Fontan optimization may represent ‘rearranging of the deck chairs’ with incremental long-term impact.

The Fontan cohort now represents the largest underserved segment of the congenital heart disease population. If we hope to make meaningful improvements, we should seek ways to restore the function of a subpulmonary pump – whether partial or complete. Any energy gain added to the Fontan circuit will shift the paradigm away from palliation toward functional cure [5].

References


WHAT YOU SEE

- Liver fibrosis and cirrhosis
- Plastic bronchitis
- Arrhythmia
- Ventricular Dysfunction
- Protein losing enteropathy

WHAT YOU DON’T SEE

- Renal dysfunction
- Transplantation
- Circulatory failure
- Atrioventricular valve regurgitation
- Hepatocellular carcinoma
- Impaired somatic growth
- Cognitive deficits
- Arteriovenous malformations
- Financial burden
- Lifetime surveillance
- Bone abnormalities
- Contraception and pregnancy
- Psychosocial challenges