The lonely ventricle in chronic Fontan circulation

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“On paper,” the Fontan circulation should achieve the necessary objectives for palliation of patients with a single ventricle. The volume load expected of the single ventricle is restored to normal, such that ventricular output, minus certain systemic-to-pulmonary collateral flow, is equal to systemic demand, and cyanosis is relieved, so that secondary erythrocytosis and the resultant coagulopathy should not cause stroke or other disability. And yet the Fontan circulation continues to disappoint. Many patients have disorders of lymphatic circulation, such as protein-losing enteropathy, plastic bronchitis, and hepatic congestion, with these apparent sequelae of central venous hypertension occurring despite measured central venous or Fontan pressures that would cause no apparent harm in biventricular circulation. In their presentation of an interesting anecdotal case in this issue of the *Journal*, Van Puyvelde and coauthors¹ explore the contribution of chronic compliance changes of the single ventricle in the Fontan circulation.

In this case, a transplant offered the opportunity to measure directly the relationship of passive volume to pressure in the explanted intact single ventricle. The observed sharply elevated pressure-volume relationship relative to measurements taken of an unrelated specimen lends support to this group's previously published concept of diastolic functional change in the chronically underloaded systemic ventricle.² They and others have attributed this change to a constant deficiency of ventricular preload, caused by the restrictor effect of the pulmonary circulatory bed. How ventricular filling pressure passes from low to high, as in the presented case, represents an apparent paradox. Perhaps the explanation lies in the absence of change in systemic ventricular volume accommodation, wherein the dampening effect of the restrictor (the Fontan circulation) is substituted for the dynamic effect of the pulmonary ventricular pumping mechanism, which can multiply the pulmonary throughput, and therefore systemic ventricular preload, in response to exercise. The “lonely ventricle,” being separated from the ordinarily variable demands of physiologic functioning, withers in its functional capacity like the proverbial coal separated from the fire.

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Central Message

In this article, Van Puyvelde and coauthors explore the long-term effects of Fontan physiology on the functional capacity of the single ventricle.

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Is there a way around this effect? In the case presented by Van Puyvelde and coauthors,¹ protein-losing enteropathy seems to have been precipitated by elective closure of a deliberate fenestration 1 year after Fontan completion. By allowing a “way around” the restrictor of the passive pulmonary circulation, a fenestration may expose the systemic ventricle to a more dynamic range of volume handling demands, perhaps maintaining better function. In fact, the benefit of early conversion from Glenn circulation, which is in effect identical to a very large fenestration, is increasingly questioned. Alternative strategies may delay the Fontan completion or forgo it altogether: these range from delaying Fontan completion until at least the onset of significant and progressive cyanosis to the addition of a systemic-to-pulmonary shunt to alleviate cyanosis at the expense of constant increased ventricular volume demand.

The report of Van Puyvelde and coauthors¹ contributes one piece in the puzzle of long-term palliation for the patient with a single ventricle. It is a thought-provoking reminder that the Fontan circulation is highly unphysiologic, and that more remains to be learned about this challenging group of patients.

References

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