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Images in Cardiology

Multiple Coronary Artery/Ventricular Microfistulas: A Late Complication of Fontan Surgery?

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A 16-year-old girl with a history of Fontan surgery for single right ventricle, common atrioventricular valve, common atrium, and pulmonary stenosis at 4 years of age was admitted because of Fontan circulatory failure (FCF). Angiography performed at 9 years of age revealed left coronary artery tortuosity and coronary artery/ventricular microfistulas (CAMFs) with a hypoplastic right coronary artery and an ejection fraction of 53% (Fig. 1A; Video 1, view video online). During treatment of FCF, she suffered a brain hemorrhage caused by a dural arteriovenous fistula of the transverse–sigmoid sinus. Despite extensive treatment, she died of sepsis. An autopsy revealed multiple dilated blood vessels with unique undulating shapes surrounded by sparse fibrosis, both grossly and histologically, within the ventricular myocardium from the subendocardium to the midwall (Fig. 1B-E), with an opening into the ventricular chamber (Fig. 1F). Vessel diameter varied from several hundred micrometers to 1 millimeter (approximately the same size as small arteries or veins); however, smooth muscle cells and elastic fibers were sparse in the vessel walls (Supplemental Fig. S1, A-D). Immunohistochemically, the vessels were lined by $CD31^{+}$ endothelial cells, and were negative for D2-40 (Supplemental Fig. S1, E,F). These pathological features were consistent with those of residual sinusoids, which cause CAMFs.

The prognosis of children with congenital heart disease has dramatically improved in recent years owing to advances in medical and surgical treatments. At present, the management of late complications in patients surviving until adulthood is a major concern. Several cases of CAMFs in univentricular circulation, with or without history of Fontan surgery, have been reported; however, their frequency and pathogenesis remain unclear.[1,2] To the best of our knowledge, this is the first case of CAMF identified by both angiography and pathological examination. Sinusoidal connections may develop and cause CAMFs late after Fontan surgery, which can affect FCF.

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Disclosures

The authors have no conflicts of interest to disclose.

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Supplementary Material

To access the supplementary material accompanying this article, visit the online version of the *Canadian Journal of Cardiology* at www.onlinecjc.ca and at https://doi.org/10.1016/j.cjca.2020.09.004.

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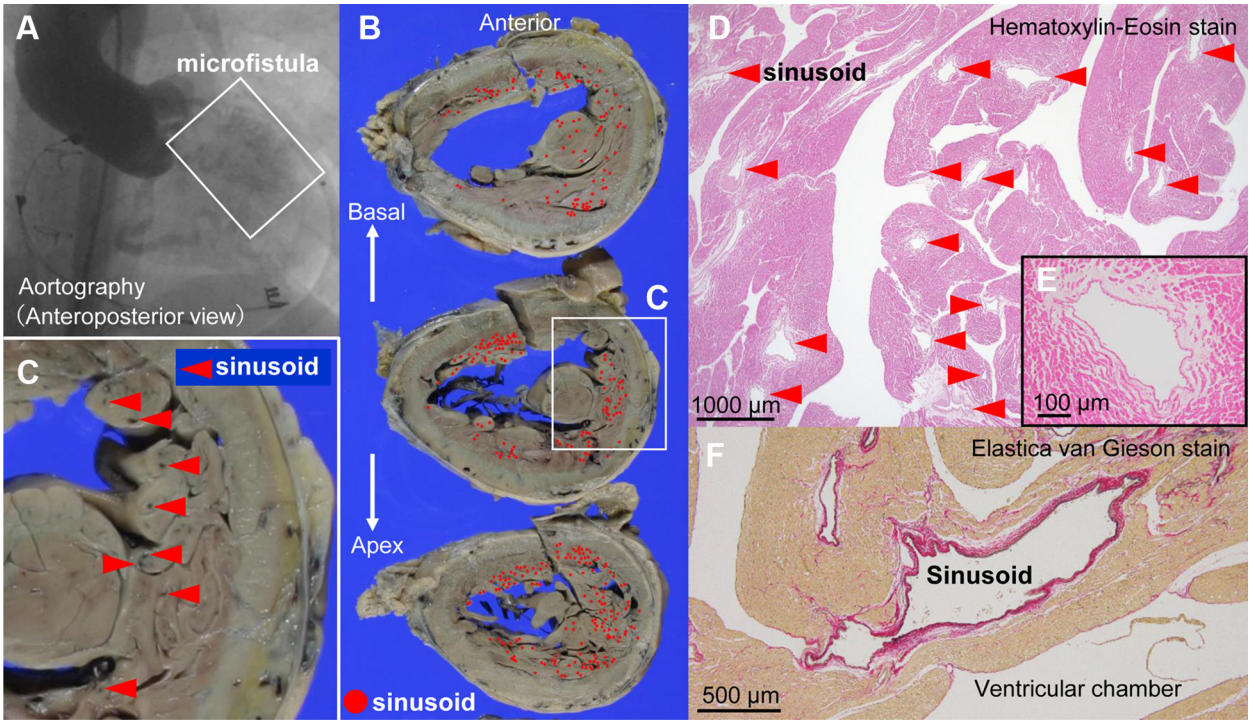

Figure 1. Aortography and pathology images. (**A**) Aortography showing coronary artery/ventricular microfistulas (CAMFs) (**white box**). (**B**) Mapping of CAMFs (**red circle**) in gross images of autopsied heart. (**C**) Enlarged Figure 1B, with **arrowheads** indicating CAMFs. (**D-F**) Histological images of CAMFs stained with hematoxylin-eosin (**D, E**) and Elastica van Gieson (**F**).

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