

Active surveillance for coronary artery aneurysms and fistulas

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This editorial refers to ‘A giant coronary aneurysm with a coronary artery–pulmonary artery fistula: 14 years of follow-up with multimodality imaging’, by C. Nishino et al. <https://doi.org/10.1093/ehjcr/ytac195>.

The prevalence of coronary artery to pulmonary artery (CA-PA) fistulas accounts for <0.5% of all congenital heart disease¹ and can be identified in approximately 0.1–0.2% of patients during diagnostic coronary angiography. Indeed, while fistulas seem to be more common in paediatric patients with concomitant congenital heart disease, the finding of the malformation in adult patients in the absence of structural heart disease or symptoms is rare. A similar prevalence can be found with CA aneurysms, and given the impaired flow dynamics in coronary fistulas, aneurysms are more commonly identified in these patients, with an overall background prevalence of 0.35%.

In the recently published *Cardiovascular Flashlight* by Nishino et al.,² the authors report that a 49-year-old patient was referred for an echocardiogram having been found with a murmur. Evidence of a CA-PA fistula was subsequently identified, with confirmation of both a fistula and CA aneurysm on computed tomography. Interestingly, in this case, the authors report that the patient suffered with no cardiovascular symptoms. The patient was followed up under active surveillance over a period of 14 years, with surgery performed once the aneurysm had grown to 33 mm, with a positive outcome.

The presence of both a coronary fistula and aneurysm in the absence of symptoms is rare, with few other such reported cases in adults, especially with such large dimensions as described in this case.³ Often the malformations are the legacy of a previous percutaneous coronary intervention (PCI), myocardial infarction, trauma, or inflammatory vasculopathies.⁴ Fistulating coronary disease often presents with anginal symptoms owing to myocardial ischaemia through a coronary steal phenomenon, necessitating surgical ligation of the vessel and initiation of long-term antithrombotics owing to the risk of residual thrombus in the aneurysm stump⁵; however with an unoperated patent fistula, the optimal treatment regime in asymptomatic patients is unclear.

The second notable comment regarding this case is the decision to adopt a strategy of active surveillance of the malformation and ob-

serving the dimensions and evolution of symptoms over a 14-year period. Although a coronary fistula might also cause an additional volume/pressure overload of both the PA and the left ventricle, a gradual but continuous expansion during long-term observation in this particular case is worthy of attention.

Insights from the multicentre Coronary Artery Aneurysm Registry⁶ suggests that in the absence of atherosclerotic CA disease, the next most frequent cause of coronary aneurysms is connective tissue disease, which was not reported in this case. There is a relative paucity of data regarding the optimum timing of intervention on coronary aneurysms and whether closure should be undertaken percutaneously or surgically, although the rate of growth and absolute dimensions should be taken into account when deciding when and how to intervene. The prevalence and characteristics of coronary aneurysm and fistula subtypes are shown in *Table 1*.

Coronary aneurysms vary in prevalence by subtype, with saccular the most common making up approximately 50%, fusiform 40%, giant 5–10%, and mixed <5%, with the left anterior descending artery being the most frequently affected vessel. Strategies of treatment can be either percutaneous or surgical, with registry data identifying no significant difference between either treatment in terms of major adverse cardiovascular event.⁶ Importantly, PCI with drug-eluting stents is preferred over the use of bare metal stents, owing to a lower rate of in-stent restenosis. Ectasia has been introduced as a subtype of coronary aneurysm, with significance defined as diffuse dilatation of the vessel >1.5× the adjacent normal segment.⁷

The classification of CA fistulas is based on location. Indications for closure are dictated largely by risks of ischaemia/rupture and ventricular dysfunction. While the current recommendation for intervention in fistulating coronary disease is based on dimensions (>2× reference vessel diameter),⁸ randomized clinical trials pertaining to

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Table 1 Prevalence and subtypes of coronary artery aneurysms and fistulas

Coronary artery aneurysm prevalence	0.35% background prevalence identifiable during diagnostic coronary angiography
Coronary artery fistula prevalence	0.1–0.2% background prevalence identifiable during diagnostic coronary angiography
<i>Aneurysm subtype prevalence</i>	
Saccular	50%
Fusiform	40%
Giant	5–10%
Other/mixed	<5%
<i>Fistula subtype by site</i>	
LAD to pulmonary artery	<ul style="list-style-type: none"> • Small • Initial conservative strategy
LCx to coronary sinus	<ul style="list-style-type: none"> • Can result in fistula dilatation • Often require closure (surgical or percutaneous)
RCA to a venous structure	<ul style="list-style-type: none"> • Likely to result in fistula dilatation • Often require closure (surgical or percutaneous)
Acquired coronary fistula	<ul style="list-style-type: none"> • Require closure, less challenging than other subtypes percutaneously

the optimum closure strategy remain awaited, and an important unanswered question remains.

Nishino *et al.* elected to actively monitor their patient over a period of 14 years, and revascularize with surgical CA bypass grafting and

fistular resection. This *Cardiovascular Flashlight* highlights several points; firstly, that fistulating coronary disease warrants further study and inclusion into specialist registries, with a view to identify long-term risks of rupture and to further explore strategies of active surveillance. Secondly, the use of optimum medical treatments such as antithrombotics requires further investigation, and the assessment of long-term risks of percutaneous vs. surgical intervention should be evaluated.

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