Percutaneous Coronary Intervention in Anomalous Right Coronary Artery: Ready to Implement in Clinical Routine?

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Anomalous coronary artery originating from the opposite sinus of valsalva (ACAOS) is a rare inherited cardiac disease with a low prevalence of 0.26% in the general population.[1]

ACAOS with an interarterial course between the great arteries is historically called 'malignant' because it is associated with an anticipated higher risk of myocardial ischaemia and sudden cardiac death, especially affecting young patients during sports.

ACAOS is responsible for up to one-third of all sports-related sudden cardiac death.[2] However, this calculation represents the occurrence of ACAOS in patients who experience sudden cardiac death and not the proportion of people with the condition who will die because of it – in fact, the risk of patients living with ACAOS seems to be rather low.[3]

Although available data are limited and randomised controlled trials are lacking (because of ethical concerns), many experts in the field agree that a primary surgical approach should be the therapy of choice in ACAOS patients with an interarterial course.

However, with the increased use of non-invasive imaging, it can be assumed that the absolute number of patients diagnosed with the condition is rising, which reopens the discussion about the need for surgical correction of all ACAOS or whether some variants of the condition are only innocent bystanders.[3–5]

Another consideration is that the risk seems to decrease with ageing.[6] This could be explained by a selection bias towards low-risk patients or because of changes in shear stress with stiffening of the aorta in older people. An opposite argument is that, with increasing aortic diameter, ACAOS could become symptomatic later in life.

After an ACAOS has been detected, a careful haemodynamic evaluation is recommended to identify patients at risk of ischaemia.[7–9] The mechanisms leading to haemodynamic relevance in ACAOS are complex.[7,10,11] It has been suggested that a scissor-like mechanism where the coronary artery is compressed between the aorta and pulmonary artery is the culprit for ischaemia. Given the large difference in pressure between the two circulatory systems, this hypothesis seems to be unlikely, especially in absence of pulmonary hypertension or pathologically enlarged great vessels.

Therefore, the interarterial course instead acts as the surrogate for the crucial anatomical high-risk feature of the intramural course (coursing of the proximal vessel within the tunica media of the aortic wall).

Along with the intramural course, the vessel presents with a slit-like ostium, an acute take-off angle, an elliptic vessel shape and proximal narrowing (after the vessel leaves the intramural part). This forms a complex stenosis with fixed and dynamic components recently suggested by our group, where the intramural part is laterally compressed when stroke volume and aortic pressure increase.[8–12]

Elements of the fixed component are best compared to the concept of stenosis in coronary artery disease, with flow restriction due to decreasing cross-sectional area. The dynamic elements are driven mainly by the lateral compression of the intramural segment and can be unmasked only during exercise, as demonstrated by Angelini et al.[12]

The primary therapy in case of ACAOS with haemodynamic relevance is surgical revascularisation. Even though several techniques have been described, coronary 'unroofing' is the most common reported surgical procedure in the literature. The process is carried out to eliminate the intramural segment and slit-like ostium by opening up the intramural segment without translocating the coronary artery. Accordingly, it has been shown that the technique is the ideal approach in patients with long intramural segments.[13] In shorter segments, the intramural course can be eliminated but an acute take-off angle may persist and, in these cases, translocation might be preferred. What role percutaneous coronary

intervention (PCI) may play in ACAOS is widely unknown, especially with respect to the proximal segments.[14–16]

In this regard, we want to congratulate the authors Simon EJ, et al. for the case report, published in this volume, where PCI of a 69-year-old male patient presenting with R-ACAOS was performed.[21] In agreement with currently available data, unroofing was discussed but, because the patient had severe pulmonary comorbidities, a decision to carry out PCI was made. PCI was performed under intravascular ultrasound (IVUS) guidance, where the authors could confirm the lateral compression of the segment. The procedure was successful and resulted in resolution of the patient's symptoms.

Below, we would like to discuss the potential use of PCI and its pitfalls, technical challenges, outcomes and patient selection.

From a pathophysiological standpoint, PCI may eliminate the slit-like ostium, proximal narrowing as well as the lateral compression of the intramural segment. However, the evidence is low as the literature contains mainly case reports.[15]

The study by Angelini et al. is one of the few published studies on PCI being performed in ACAOS. Out of 67 patients with R-ACAOS (mostly middle-aged and older patients (mean age 48 ± 12 years), 42 underwent stent angioplasty of the intramural segment under IVUS guidance.[12] The cross-sectional area of the intramural segment was increased from 4.8 to 10.8 mm^2, and no pulsatility of the segment and no lateral compression were observed after the procedure. At follow-up of 5 years, no deaths had occurred and clinical symptoms had decreased in 74% of patients. Out of 10 patients with a clinically indicated follow-up coronary angiography, four presented with an in-stent restenosis. In one patient at 3 years' follow-up, the degree of in-stent restenosis was 40% and, at 6 years a stent thrombosis occurred, possibly due to stent kinking during phasic systolic bending.[12]

In another study by Darki et al. with 11 patients with R-ACAOS undergoing PCI, restenosis occurred in one patient (9%) after a mean follow-up time of 8.5 years, with favourable results otherwise.[17]

Regarding optimal patient selection for PCI in ACAOS, technical aspects have to be considered. Localisation of the ostium in R-ACAOS can be challenging, with an associated prolonged radiation exposure, increased contrast volumes or even leading to the erroneous diagnosis of total occlusion of the vessel; a pre-procedural coronary computed tomography angiography (CCTA) could be helpful in this context and allow the ostial location and the course of the anomalous vessel to be located. Based on the CCTA images, a preselection of potential matching catheters, as suggested by Ben-Dor et al., could increase the ability for rapid intubation of the anomalous ostium.[18]

Other technical considerations are optimal stent selection. Stent size could be estimated with the help of the segment just distal to the intramural course and covering the whole length with minimal protrusion into the aorta, as proposed by the ANOCOR working group.[19]

Drug-eluting stents have long been shown to reduce in stent restenosis in coronary artery disease lesions due to their antiproliferative nature; however, it is unknown whether this would apply also to stents for ACAOS lesions. One might argue that a more robust stent is needed in the intramural segment to resist lateral compression or kinking of the vessel, while allowing adjustment to the curvature of the intramural segment to decrease shearing within the stent. A similar problem exists in tortuous vessel stenting, where Han et al. suggested optimised stent geometry to reduce shearing forces on the material.[20]

To conclude, the primary therapeutic option in haemodynamically relevant R-ACAOS is surgical unroofing, if the anatomy is favourable with a long intramural course. The role of PCI of the proximal intramural part of the anomalous coronary artery is unclear; however, stenting might be an alternative approach in non-surgical candidates or older patients.

More data are needed on long-term stent patency rates and optimal stent choice to improve patient selection for PCI in the clinical setting of R-ACAOS. ❑

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