CASE REPORT

Is endovascular therapy the right choice for treatment of functional compression of anomalous right coronary artery arising from left coronary sinus with interarterial course?

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SUMMARY

We report a case of successful percutaneous coronary intervention (PCI) for reversible ischaemia owing to an anomalous right coronary artery arising from the left coronary sinus with malignant interarterial course and not associated with obstructive coronary artery disease (CAD). PCI for treatment of functional compression of the ostioproximal intramural segment of the anomalous vessel, though described in the literature, is technically challenging, requiring appropriate hardware selection; the peculiar anatomical milieu, in the absence of atherosclerotic CAD, lends itself to an uncertain long-term outcome following endovascular therapy with stenting.

BACKGROUND

The clinical importance of an anomalous coronary artery arising from opposite sinus (ACAOS) arises from its association with sudden cardiac death, myocardial ischaemia, arrhythmias and syncope.¹ Sudden death mostly happens before 35 years of age, with the increased risk of sudden cardiac death being associated with four risk factors which are interarterial course (IAC) between the aorta and pulmonary artery, slit-like coronary orifice, acute angle of take-off of the anomalous coronary artery from the aorta and the presence of aortic intramural coronary arteries.² As the clinical course of left-ACAOS is more malignant than that of right-ACAOS³⁻⁴ surgical correction is especially recommended for left-ACAOS that involves a large territory at risk.⁵⁻⁶ Nevertheless, treatment in patients with right-ACAOS remains controversial because most of these cases have a benign outcome. Surgery is still considered to be the gold standard procedure in right-ACAOS patients with reversible ischaemia. Since the long-term results of coronary bypass grafting (CABG) are not ideal, percutaneous coronary intervention (PCI) can be an effective alternative in such patients.

Even though stent angioplasty seems an interesting alternative to cardiac surgery, it is an often technically complex procedure with difficulty to reach and cannulate the ectopic artery's ostium. Furthermore, the ideal degree of stent dilation remains problematic and the type of stent (drug eluting or standard stent) is not established. We report a case of successful PCI for a symptomatic right-ACAOS with malignant interarterial course not associated with obstructive coronary artery disease (CAD). Limitations of various surgical repair techniques and technical

considerations in the absence of intravascular ultrasound (IVUS) support during PCI to right-ACAOS are briefly discussed.

CASE PRESENTATION

A 50-year-old man, hypertensive with no other cardiac risk factors, and a known case of bronchial asthma, was referred for cardiac evaluation for his chest pain on exertion and persisting symptom of dyspnoea on exertion New York Heart Association class II despite adequate therapy for asthma.

INVESTIGATIONS

ECG showed left ventricular hypertrophy; two-dimensional echocardiography revealed left ventricular (LV) ejection fraction of 60% and no regional wall motion abnormalities (RWMA). Exercise treadmill test was positive for inducible ischaemia at 6 metabolic equivalents (METs). He was taken up for coronary angiography which did not reveal any obstructive CAD but showed an anomalous right coronary artery (RCA) originating from the left coronary sinus (right ACAOS) (figure 1).

Figure 1 Anteroposterior caudal view showing right coronary artery (RCA) originating from left coronary sinus and simultaneous filling of left coronary artery. No obstructive coronary artery disease is noted in the anomalous RCA.

To cite: Vadivelu R, Bagga S. *BMJ Case Reports* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-007856

In view of a positive stress test with non-occlusive CAD, stress myocardial perfusion imaging (MPI) with technetium was undertaken to determine functional significance of the right ACAOS. Stress MPI was positive for stress-induced reversible perfusion defect in inferior and inferolateral segments of LV involving 25% of LV myocardium, at 5 METS (figure 2). Knowing that anomalous RCA has a variable relationship with great vessels, CT imaging of the great vessels and coronaries was carried out, which defined the interarterial course of the anomalous RCA (ARCA) traversing between the ascending aorta and the major pulmonary artery.

TREATMENT

Coronary revascularisation is often required in symptomatic patients with ACAOS. Though surgical management is considered to be the gold standard in ACAOS, it has its own demerits. Since the patient refused surgery, the patient was taken up for PCI with stenting to the ostioproximal part of the RCA (interarterial portion).

After obtaining right femoral arterial access, the ARCA was hooked with a Judkins left (JL) (Medtronic, Minnesota, USA)

3, 6 Fr guiding catheter (GC) using a simplified technique. Keeping a slight clockwise twist (so that it does not automatically cannulate the left coronary artery), the JL catheter was pushed deep in the left sinus of Valsalva. In doing so, the catheter took a U-turn on itself and its tip went up and cannulated the anomalously arising RCA. An ATW marker guiding wire (Cordis, Johnson & Johnson Inc, USA) was negotiated across the ARCA. This was specifically chosen in the absence of an IVUS available at shelf, so as to choose the approximate stent length to take care of the interarterial course of the artery based on evaluation of coronary CT angiogram. Stent Cypher Select (Cordis, Johnson & Johnson Inc, USA) 3×23 mm was implanted in the ostioproximal RCA at 12 atmospheres (atm) (figures 3 and 4) and postdilated with a non-compliant balloon Sprinter NC 3.5×12 mm (Medtronic Inc) at 18 atm. There were no periprocedural complications.

OUTCOME AND FOLLOW-UP

The patient has remained angina-free on follow-up. Twelve-month follow-up coronary CT angiogram showed the stent in situ in the interarterial segment of the ARCA without

Figure 2 Stress myocardial perfusion imaging with technetium was positive for stress-induced reversible perfusion defect in inferior and inferolateral segments of left ventricular (LV) (compare the bottom two rows at stress and rest) involving 25% of LV myocardium, at 5 metabolic equivalents and target heart rate of 97%.

Figure 3 Anomalous right coronary artery (ARCA) was hooked with JL GC 3, 6 Fr and ATW guiding wire was used to cross the lesion. Stent cypherselect 3×23 mm was implanted across the interarterial segment of ARCA.

any in-stent restenosis or fractures (figures 5 and 6). He had completed 37 months of follow-up with no significant symptoms and no restriction of daily activities. Stress technetium myocardial perfusion scan at 37 months did not reveal any reversible perfusion defect in the ARCA territory at 10 METS (figure 7).

DISCUSSION

Coronary artery anomalies have been reported in 1.3% of patients undergoing coronary angiography; however, the incidence may even be higher if detected by multislice coronary

Figure 5 Oblique CT coronary angiography imaging showing the stent in the proximal intramural segment of the anomalous right coronary artery.

computed angiography.^{7 8} Though familial clustering of anomalous coronary arteries has been described^{9 10} and animal studies in mice¹¹ have demonstrated Tbx1 gene defect in association with anomalous coronary arteries, a definitive gene for humans is as yet unknown.

Right-ACAOS is a rare congenital coronary artery anomaly, with an incidence varying from 0.107% to 0.92% among patients undergoing coronary angiography.^{7 12} Though a significant number of cases with right-ACAOS do not warrant intervention, treatment is indicated in the presence of disabling symptoms with reversible ischaemia, as documented by a nuclear stress test and a large dependent myocardial territory (more than a third of the total).¹³ The mechanisms of myocardial ischaemia have been hypothesised to be due to the

Figure 4 Left anterior oblique caudal view: final image after stent deployment in anomalous right coronary artery.

Figure 6 Volume rendered CT image showing detailed anatomy of origin and course of left coronary artery and anomalous right coronary artery and the stented region is seen as a bulge in between aorta and pulmonary artery.

Figure 7 Stress myocardial technetium perfusion scan showing near complete resolution of reversible perfusion defect is seen in the inferior segments (bottom two rows).

intramural compression of the ostioproximal part of ARCA within the aortic wall as demonstrated on IVUS imaging¹⁴ and, less commonly, secondary to acute-angled take off of the anomalous vessel from the opposite sinus with compression of the abnormal slit-like opening of the ostium, which is most often secondary to exercise-induced increased aortic pressure.¹⁵ The risk of sudden cardiac death (SCD), especially in the presence of an interarterial course between the aorta and pulmonary artery, is less as compared with left-ACAOS.⁴ Older patients (≥ 30 years) appear to have a lower risk of SCD, although some risk may still exist.

This case illustrates the alternative treatment with PCI for right-ACAOS for which surgical revascularisation is considered to be the standard of care,¹⁶ though a recent single centre study could not find a long-term survival benefit of surgical intervention on the ACAOS population.¹⁷ Surgical methods include aortocoronary bypass (CABG), ostial reimplantation, the unroofing technique and pulmonary artery (PA) translocation.^{18–20} CABG is preferred when there is associated atherosclerotic obstructive CAD and especially for cases that are not appropriate for unroofing or translocation.²⁰

Aortocoronary bypass grafting has inherent limitations with limited graft patency due to the inevitable competitive flow between the bypass and the native anomalous coronary artery. Both saphenous vein and internal mammary grafts have been used. Venous grafts are more prone to early failure and arterial grafts are vulnerable to vasospasm secondary to coronary steal phenomenon.²¹ Competitive flow harbingers graft failure and ligation of the native anomalous RCA has been suggested as an alternative to reduce this event, but ligation is met with serious adverse ischaemic events as the flow from bypass graft may not be sufficient in the immediate postoperative period.¹⁹ A minimally invasive, off-pump, right anterior thoracotomy approach for right internal mammary artery to ARCA has been recently reported to be successful in a small cohort of patients.²²

Since the long-term results of CABG are not ideal, alternative surgical procedures are increasingly considered as more favourable options. Unroofing and translocation procedures are appropriate and effective for certain morphological types, with both techniques showing favourable outcome at mid-term follow-up.^{18–20} Unfortunately, aortic insufficiency can result, even late after unroofing,²³ and myocardial ischaemia may persist, particularly in patients

with repaired ARCA with IAC.²⁴ The PA translocation procedure, in fact, converts high-risk variants (those passing between the great arteries) to low-risk variants (those that do not pass between the great arteries), without actually moving the coronary artery itself.²⁵ It is, of course, critical to document that other factors associated with coronary insufficiency, such as fixed obstructive coronary lesions or slit-like ostium, do not exist when this procedure is undertaken. Whether lateral or anterior translocation is chosen depends on the orientation of the coronary artery itself, and equally important, specific positioning of the aortic and pulmonary roots.

PCI of the proximal intramural segment of a patient with right-ACAOS though feasible is technically challenging. PCI has been reported to be suitable for revascularisation in the right ACAOS with superimposed atherosclerotic CAD in RCA.^{26–29} Primary PCI has also been reported to be feasible in ACAOS patients presenting with acute myocardial infarction secondary to atherosclerotic CAD.²⁸ PCI in right-ACAOS without superimposed CAD has been reported in adults, with short-term success.^{30–32} The main aim in this setting is to prevent interarterial ARCA from phasic compressive forces exerted by the great vessels. The radial strength of the stent protects the ARCA from dynamic compression during systole and also prevents flap-like mechanism of abnormal ostial opening.³²

However, the operator needs to understand the technical considerations for performing PCI for right-ACAOS. Difficult cannulation, suboptimal backup support with standard GCs owing to the ectopic juxtacommissural ostium and tangential direction of the anomalous vessels adds to the procedural complexities and increases procedural time. Appropriate GC selection and choosing extra support guidewires and using balloon catheter support to facilitate guidewire advancement are some of the technical considerations that need merit during PCI to right-ACAOS.³³ Switching from one access route to another (femoral to radial/brachial or vice versa), use of large GC like Amplatz GC (AL2,3), unusual GC-like Champ catheter or venous bypass GC, use of moderately stiff guidewires like PT (Boston Scientific, Minnesota, USA) can be tried in difficult situations.³³ We have been using a simple technique using JL GC, which has been well described in the literature, to hook the right ACAOS during PCIs for this vessel with an excellent technical success rate.³⁴

Although IVUS guidance is recommended for optimising PCI for right-ACAOS,¹⁴ the unavailability of this technology in our laboratory added further complexity to the procedure. Lack of adventitia and hypoplastic intramural segment associated with ACAOS are risk factors for aortic root dissection, especially encountered during aggressive coronary dilation. This feature of ACAOS leads to uncertainty about the ideal degree of stent dilation. The distal vessel reference diameter may be larger as compared with the proximal hypoplastic segment adding to the dilemma. IVUS guidance has been suggested as the main tool in such cases and the largest possible initial diameter should be aimed for prevention of restenosis.³¹

The poor results obtained after stenting of muscular bridges suggest that restenosis may be more frequent in the presence of phasic compressive forces, which surely persist also in ACAOS patients, even after stent implantation. In view of this theoretically higher rate of restenosis, drug-eluting stents (DES) can be assumed to have an advantage over bare metal stents (BMS). However, since the intramural segment of this anomalous artery lacks an adventitia, the mechanism of restenosis may be different and hence the antirestenotic impact DES in this unusual milieu needs further investigation.

Learning points

- ▶ Right-ACAOS constitutes a group of coronary artery anomalies most commonly associated with myocardial ischaemia and, sometimes with sudden death, though the risk seems lower in older adults.¹⁷ The challenge lies in understanding the variability of their functional repercussions. The greater current challenge is to decide which cases require interventional treatment at all. Standard treatment remains surgical revascularisation. PCI offers an interesting alternative; however, several issues need consideration. Both modes of revascularisation require long-term and close follow-up of patients.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Perloff JK. Congenital anomalies of the coronary circulation. In: Perloff JK, ed. *The clinical recognition of congenital heart disease*, 6th edn. Philadelphia, PA: W.B. Saunders, 2012:530.
- 2 Lorenz EC, Mookadam F, Mookadam M, et al. A systematic over-view of anomalous coronary anatomy and an examination of the association with sudden cardiac death. *Rev Cardiovasc Med* 2006;17:205–13.
- 3 Eckart RE, Scoville SL, Campbell CL, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. *Ann Intern Med* 2004;141:829–34.
- 4 Basso C, Maron BJ, Corrado D, et al. Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. *J Am Coll Cardiol* 2000;35:1493–501.
- 5 Angelini P, Walmsley RP, Liberos A, et al. Symptomatic anomalous origination of left coronary artery from the opposite sinus of Valsalva: clinical presentations, diagnosis, and surgical repair. *Tex Heart Inst J* 2006;33:171–9.
- 6 Gersony WM. Management of anomalous coronary artery from the contralateral coronary sinus. *J Am Coll Cardiol* 2007;50:2083–4.
- 7 Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990;21:28–40.
- 8 Shi H, Aschoff AJ, Brambs HJ, et al. Multislice CT imaging of anomalous coronary arteries. *Eur Radiol* 2004;14:2172–81.
- 9 Devanagondi R, Brenner J, Vricella L, et al. A tale of two brothers: anomalous coronary arteries in two siblings. *Pediatr Cardiol* 2008;29:816–19.
- 10 Hung-Yu C, Wei-Hsian Y. Anomalous right coronary artery arising from left coronary sinus in two brothers. *Acta Cardiol* 2011;27:124–7.
- 11 Théveniau-Ruissy M, Dandonneau M, Mesbah K, et al. The del22q11.2 candidate gene Tbx1 controls regional outflow tract identity and coronary artery patterning. *Circ Res* 2008;103:142–8.
- 12 Angelini P, Villason S, Chan AV, et al. Normal and anomalous coronary arteries in humans. In: Angelini P, ed. *Coronary artery anomalies: a comprehensive approach*. Philadelphia, PA: Lippincott Williams & Wilkins, 1999:27–150.
- 13 Angelini P. Coronary artery anomalies: an entity in search of an identity. *Circulation* 2007;115:1296–305.
- 14 Angelini P, Velasco JA, Ott D, et al. Anomalous coronary artery arising from the opposite sinus: descriptive features and pathophysiologic mechanisms, as documented by intravascular ultrasonography. *J Invasive Cardiol* 2003;15:507–14.
- 15 Grollman JH Jr, Mao SS, Weinstein SR. Arteriographic demonstration of both kinking at the origin and compression between the great vessels of an anomalous right coronary artery arising in common with a left coronary artery from above the left sinus of Valsalva. *Cathet Cardiovasc Diagn* 1992;25:46–51.
- 16 Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *Circulation* 2008;118:2395–451.
- 17 Krasuski RA, Magyar D, Hart S, et al. Long-term outcome and impact of surgery on adults with coronary arteries originating from the opposite coronary cusp. *Circulation* 2011;123:154–62.
- 18 Mainwaring RD, Reddy VM, Reinhartz O, et al. Anomalous aortic origin of a coronary artery: medium-term results after surgical repair in 50 patients. *Ann Thorac Surg* 2011;92:691–7.

- 19 Fedoruk LM, Kern JA, Peeler BB, *et al.* Anomalous origin of the right coronary artery: right internal thoracic artery to right coronary artery bypass is not the answer. *J Thorac Cardiovasc Surg* 2007;133:456–60.
- 20 Davies JE, Burkhart HM, Dearani JA, *et al.* Surgical management of anomalous aortic origin of a coronary artery. *Ann Thorac Surg* 2009;88:844–8.
- 21 Yanagawa B, Alghamdi AA, Chen RB, *et al.* Coronary artery bypass graft for anomalous right coronary artery. *J Card Surg* 2011;26:44–6.
- 22 Reddy RC, Takahashi M, Beckles DL, *et al.* Anomalous right coronary artery from the left sinus: a minimally invasive approach. *Eur J Cardiothorac Surg* 2012;41:287–90.
- 23 Van Son JA, Haas GS. Anomalous origin of left main coronary artery from right sinus of Valsalva: modified surgical treatment to avoid neo-coronary ostial stenosis. *Eur J Cardiothorac Surg* 1996;10:467–9.
- 24 Brothers JA, McBride MG, Seliem MA, *et al.* Evaluation of myocardial ischemia after surgical repair of anomalous aortic origin of a coronary artery in a series of pediatric patients. *J Am Coll Cardiol* 2007;50:2078–82.
- 25 Gulati R, Reddy VM, Culbertson C, *et al.* Surgical management of coronary artery arising from the wrong coronary sinus, using standard and novel approaches. *J Thorac Cardiovasc Surg* 2007;134:1171–8.
- 26 Ng W, Chow W. Successful angioplasty and stenting of anomalous right coronary artery using a 6 French left Judkin guide catheter. *J Invasive Cardiol* 2000;12:373–5.
- 27 Charney R, Spindola-Franco H, Grose R. Coronary angioplasty of anomalous right coronary arteries. *Cathet Cardiovasc Diagn* 1993;29:233–5.
- 28 Yip H, Chen MC, Wu CJ, *et al.* Primary angioplasty in acute inferior myocardial infarction with anomalous-origin right coronary arteries as infarct-related arteries: focus on anatomic and clinical features, outcomes, selection of guiding catheters and management. *J Invasive Cardiol* 2001;13:290–7.
- 29 Sohara H, Tsurukawa T, Kawabata K, *et al.* Pitfalls of intervention therapy in a patient with anomalous origin of the right coronary artery from the left sinus of Valsalva associated with organic stenosis. *J Cardiol* 1997;29:111–16.
- 30 Doorey AJ, Pasquale MJ, Lally JF, *et al.* Six-month success of intracoronary stenting for anomalous coronary arteries associated with myocardial ischemia. *Am J Cardiol* 2000;86:580–2.
- 31 Hariharan R, Kacere RD, Angelini P. Clinical articles: can stent-angioplasty be a valid alternative to surgery: when revascularization is indicated for anomalous origination of a coronary artery from the opposite sinus? *Tex Heart Inst J* 2002;29:308–13.
- 32 Giorgio M, Rosa P, Davide Z, *et al.* Radionuclide evidence for reversible ischemia after percutaneous treatment of anomalous right coronary artery with dynamic compression by great vessels. *J Cardiovasc Med* 2008;9:1134–7.
- 33 Rigatelli G, Cardaioli P. Endovascular therapy for congenital coronary artery anomalies in adults. *J Cardiovasc Med* 2008;9:113–21.
- 34 Sapra R, Kaul U, Singh B, *et al.* Simplified approach of cannulating anomalously arising right coronary artery from left sinus of Valsalva. *Cathet Cardiovasc Diagn* 1998;45:346–7.

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