Cardiology in the Young

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Transcatheter closure of an acquired post-operative aorta to right ventricle shunt in a child with complex univentricular heart

Raymond N. Haddad[1], Damien Bonnet[1,2], Diala Khraiche[1] and Sophie Malekzadeh-Milani[1]

[1]M3C-Necker, Hôpital Universitaire Necker-Enfants malades, Assistance Publique-Hôpitaux de Paris (AP-HP), Paris, France and [2]Université de Paris, Paris, France

Brief Report

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Author for correspondence:
Dr R. N. Haddad, MD, Service de cardiologie congénitale et pédiatrique, centre de référence des malformations cardiaques congénitales complexes - M3C, Hôpital Universitaire Necker-Enfants malades, 149, rue de Sèvres, 75015, Paris, France. Tel: +33 7 53 15 95 04; Fax: +33 1 44 49 47 30.
E-mail: raymondhaddad@live.com

Abstract

KONAR-Multifunctional™ VSD Occluder (Lifetech, Shenzhen, China) is one of the most recent additions to the armamentarium of device closure interventions offering special features to tackle complex cardiac anatomies. Herein, we report the first use of the KONAR-MFO in an 8.5-year-old female patient (27 kg/129 cm) with stage III palliated univentricular heart to close an acquired post-operative tunnel-like communication (5 mm long × 2.6 mm large) between the right anterior non-coronary aortic sinus and the rudimentary right ventricular cavity. The shunt was diagnosed two and a half years after bulboventricular foramen surgical enlargement. The 5× 3 mm KONAR-MFO was retrogradely implanted under ultrasound and biplane fluoroscopic guidance. Immediate and 12-month follow-up confirmed successful outcomes with complete shunt closure and preserved aortic valve competence.

Acquired aortocardiac shunts are extremely rare paediatric heart lesions.[1–4] Transcatheter closure is an attractive treatment modality, yet the anatomic complexity and morphological variability of these anomalies make them challenging to close using device occluders not originally designed for this purpose.[2–5]

Case presentation

An 8.5-year-old female patient (27 kg/129 cm) was referred to our institution for a progressively worsening aortic regurgitation that has been monitored since her last heart surgery during regular outpatient follow-up. The patient had type IIc tricuspid atresia with aortic coarctation that has been initially palliated with pulmonary artery banding and aortic repair in the neonatal period, and finally with a fenestrated total cavopulmonary connection and bulboventricular foramen enlargement (through right ventriculotomy) at the age of 6 years.

Upon admission to our unit, the patient had NYHA functional class II and physical examination showed a diastolic murmur of grade 4/6 along the left parasternal region. Chest X-ray and basic blood tests were normal. Baseline ultrasound showed a mild-to-severe eccentric aortic regurgitation (PHT of 450 ms, end-diastolic flow velocity of 0.35 m/second in the aortic isthmus). The ultrasound study was unable to accurately grade the regurgitation and depict its precise mechanism motivating a cardiac MRI that showed an anteriorly located aortic regurgitation and graded it as mild (regurgitation fraction of 25%). However, invasive diagnostic aortogram revealed normal aortic valve functioning and delineated an abnormal tunnel-like communication of 5.0 mm long× 2.6 mm large between the right anterior non-coronary aortic sinus and the rudimentary right ventricular cavity (Fig 1a). A complimentary pre-intervention planning CT angiogram confirmed catheterisation findings and excluded coronary arteries involvement. Full infective endocarditis workup came negative and the lesion was thereby considered as post-operatively acquired.

Appropriate informed consent was obtained for attempted device closure. The procedure was performed under general anaesthesia, biplane fluoroscopy, and transesophageal echocardiography. At the time of the procedure, the patient was under Warfarin therapy with INR ranging between 2 and 3. Antibiotic prophylaxis and systemic heparinisation were given as per institutional protocol. A bilateral 5-Fr arterial femoral access was obtained. Biplane aortogram was performed using a 5-Fr Pigtail diagnostic catheter (Cordis Corp., FL, United States of America) to relocate the defect that was easily crossed using a 5-Fr Judkins right coronary diagnostic catheter (Cordis Corp., FL, United States of America) in combination with an 0.032-inch/180 cm-long J-tip Radifocus® Hydrophilic Guidewire M (Terumo Corp., Tokyo, Japan). The catheter was then advanced into the rudimentary right ventricle and the wire was upgraded to a 0.035-inch/260 cm-long J-tip Stiff-type Radifocus® Hydrophilic Guidewire M (Terumo

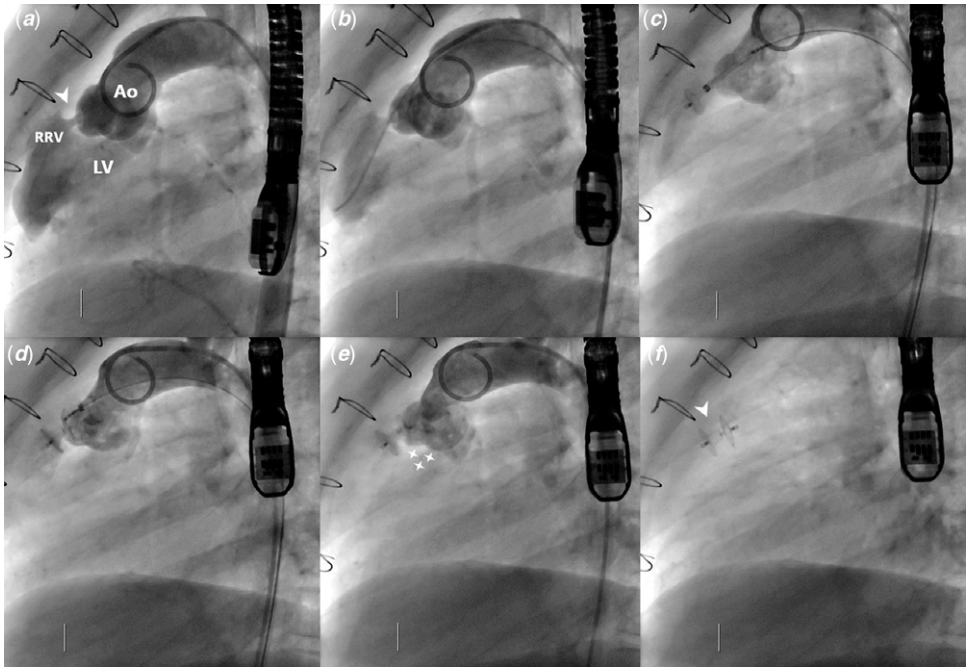

Figure 1. Diagnostic aortogram delineating a tunnel-like shunt (white pointed arrow) from the right anterior non-coronary aortic sinus to the rudimentary right ventricle (width: 2.61 mm; depth: 5.05 mm) (***a***). Consecutive sequences of the interventional closure of the shunt using a KONAR-MFO 5 × 3 mm connected from its right side (***b***–***d***). Control angiography before release showing complete shunt closure and normal aortic valve functioning (white stars) (***e***). Stable device after release (white pointed arrow) (***f***). Ao = aorta; LV = left ventricle; RRV = rudimentary right ventricle.

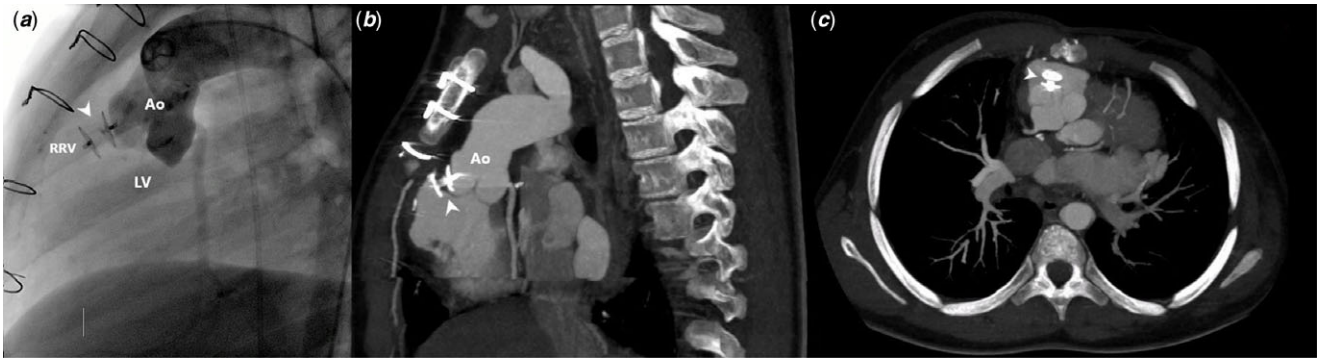

Figure 2. One-year follow-up control imaging with ascending aortogram (***a***) and multi-planar modified CT-scan reconstructions (***b***, ***c***). Note device stability, complete shunt closure, and preserved aortic valve competence (white pointed arrow). Ao = aorta; LV = left ventricle; RRV = rudimentary right ventricle.

Corp., Tokyo, Japan). A 5 × 3 mm KONAR-MFO was selected and was connected to the delivery cable from its right side. The catheter was exchanged with a 5-Fr/80 cm SteerEase™ delivery sheath (Lifetech, Shenzhen, China) through which the device was advanced and sequentially deployed under fluoroscopy guidance (Fig 1b–d). Device stability, shunt closure, and normal aortic valve function were checked on ultrasound and control angiography before releasing the device (Fig 1e and f). No early complications were observed. The procedure required 25 minutes of fluoroscopy. The patient received 48 hours of therapeutic heparin and was safely discharged with no residual shunt or aortic regurgitation. The patient was prescribed to continue warfarin with oral aspirin therapy. One year follow-up confirmed excellent results with complete shunt closure and preserved aortic valve competence (Fig 2).

Discussion

Aortocardiac shunts have been previously described.[1–5] Aetiologies are numerous and treatment strategy remains debatable.[1–6] Surgery is most commonly considered as the primary treatment option, yet interventional device closure has been repetitively reported as a safe and effective alternative with attractive advantages. Multiple devices have been implanted and the majority of the results are encouraging.[1–6] The recently introduced KONAR-MFO has been smartly designed, combining technical features of previous occluder devices, to tackle encountered difficulties.[7–9] Emerging reports are consecutively positive and clinical applications have been mainly limited to ventricular septal defects closure.[9] This is the first use of the KONAR-MFO to percutaneously treat an iatrogenic aorta to right ventricle shunt occurring in a complex child following intracardiac repair.

Iatrogenic aorta to right ventricle fistulas has been commonly described following transcatheter or surgical aortic valve replacements and ventricular septal defect repairs.[1–5] The most commonly suggested mechanisms of this complication are ischaemic necrosis, aneurysm rupture, aggressive surgical debulking, and erosion between the implanted material and the aortic wall in late-onset perforation of the aortic wall.[1–6] Our patient underwent an opening of the accessory cavity beneath the aortic valve to enlarge the bulboventricular foramen. Based on the time frame sequence of the

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events and the aseptic status of the patient, we retrospectively advocate that an iatrogenic surgical injury to the area of contact between the right anterior non-coronary aortic sinus and accessory cavity could have been the likely cause for such a particular fistulous-like communication in our patient. Our case showed the utility of the aortogram as a complimentary exam to non-invasive diagnostic tools in the straightforward mapping of the aortic valve region. Diagnostic aortogram was very useful in adjusting the diagnosis and in providing the required information for the planning of the intervention.[4,10] Multi-modality imaging and fusion technology can also be helpful for pre-procedural planning, device choice, and pre-procedural guidance, yet these technologies are not always necessary to achieve the intended outcomes.[4]

We chose KONAR-MFO as the most suitable device for this case. Indeed, the central disk perfectly fitted across the tunnel achieving immediate and stable complete shunt closure. The articulating soft right retention disk conformed to the aortic sinus curvature without interfering with the aortic valve function or the nearby coronary arteries. This is the first report of closure of this rare cardiac lesion using this particular device.[9] The procedure was technically smooth. The tunnel was easily accessed from the aorta using proper wires and the device was promoted directly into position. The retrograde approach allowed straightforward delivery avoiding technical complications of the arterio-venous circuit and reducing irradiation exposure.[11] The anatomic complexity of these rare lesions makes them sometimes challenging to close using devices not originally designed for this purpose, yet we set additional proof of the efficacy of transcatheter treatment.

Conclusion

Retrograde transcatheter closure of acquired aorta to right ventricle shunt is a feasible and safe procedure with very good outcomes. The new KONAR-Multifunctional™ VSD Occluder offers high anatomical conformability and procedural flexibility to effectively treat this complex type of heart lesion.

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Author contributions. RH collected clinical data, designed illustrative material, and took the lead in writing and revising the entire manuscript. All authors have read and approved the final version of the manuscript.

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008. The patient's legal guardians signed informed consent for the reported procedures and this publication.

References

1. Rojas P, Amat-Santos IJ, Cortés C, et al. Acquired aseptic intracardiac shunts following transcatheter aortic valve replacement: a systematic review. JACC Cardiovasc Interv 2016; 9: 2527–2538.
2. Nakamura K, Passeri JJ, Inglessis-Azuaje I. Percutaneous closure of acute aorto-right ventricular fistula following transcatheter bicuspid aortic valve replacement. Catheter Cardiovasc Interv 2017; 90: 164–168.
3. Petrou E, Iakovou I, Leontiadis E, Kourkoveli P, Rammos S, Karatasakis G. Percutaneous transcatheter closure of aorto-right ventricular fistula using the Amplatzer duct occluder. Hellenic J Cardiol 2016; 57: 185–187.
4. Nagi Haddad R, Ly R, Iserin L, Malekzadeh-Milani S. Aorta-to-right ventricle neoshunt closure using an Amplatzer Duct Occluder II device. J Card Surg 2021; 36: 2156–2159.
5. Pate GE, Al Zubaidi A, Chandavimol M, Thompson CR, Munt BI, Webb JG. Percutaneous closure of prosthetic paravalvular leaks: case series and review. Catheter Cardiovasc Interv 2006; 68: 528–533.
6. Kerkar PG, Lanjewar CP, Mishra N, Nyayadhish P, Mammen I. Transcatheter closure of ruptured sinus of Valsalva aneurysm using the Amplatzer duct occluder: immediate results and mid-term follow-up. Eur Heart J 2010; 31: 2881–2887.
7. Boukaram L, Haddad RN, Daou L, Saliba Z. Transcatheter closure of congenital Gerbode-type ventricular septal defect using the new multifunctional occluder device. J Card Surg 2021; 36: 2986–2988.
8. Haddad RN, Daou LS, Saliba ZS. Percutaneous closure of restrictive-type perimembranous ventricular septal defect using the new KONAR multifunctional occluder: midterm outcomes of the first middle-eastern experience. Catheter Cardiovasc Interv 2020; 96: E295–E302.
9. Haddad RN, Saliba Z. Interventional device closure of perimembranous ventricular septal defects: challenges, pitfalls, and advancements in device technology. Interv Cardiol 2021; 13: 109–114.
10. Alam M. Transesophageal color flow Doppler features of aorta to right ventricle fistula. Chest 1993; 103: 1907–1908.
11. Haddad RN, Rizk C, Saliba Z, Farah J. Percutaneous closure of ventricular septal defects in children: key parameters affecting patient radiation exposure. Am J Cardiovasc Dis 2021; 11: 65–72.

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