

University of Groningen

Large common left and right coronary artery to coronary sinus fistula

Hoendermis, E.S.; Waterbolk, Tjalling W.; Willems, Els; Zijlstra, F.

Published in:
Interactive Cardiovascular and Thoracic Surgery

DOI:
[10.1510/icvts.2006.132324](https://doi.org/10.1510/icvts.2006.132324)

IMPORTANT NOTE: You are advised to consult the publisher's version (publisher's PDF) if you wish to cite from it. Please check the document version below.

Document Version
Publisher's PDF, also known as Version of record

Publication date:
2006

[Link to publication in University of Groningen/UMCG research database](#)

Citation for published version (APA):
Hoendermis, E. S., Waterbolk, T. W., Willems, E., & Zijlstra, F. (2006). Large common left and right coronary artery to coronary sinus fistula. *Interactive Cardiovascular and Thoracic Surgery*, 5(6), 788-789. <https://doi.org/10.1510/icvts.2006.132324>

Copyright

Other than for strictly personal use, it is not permitted to download or to forward/distribute the text or part of it without the consent of the author(s) and/or copyright holder(s), unless the work is under an open content license (like Creative Commons).

The publication may also be distributed here under the terms of Article 25fa of the Dutch Copyright Act, indicated by the "Taverne" license. More information can be found on the University of Groningen website: <https://www.rug.nl/library/open-access/self-archiving-pure/taverne-amendment>.

Take-down policy

If you believe that this document breaches copyright please contact us providing details, and we will remove access to the work immediately and investigate your claim.

Downloaded from the University of Groningen/UMCG research database (Pure): <http://www.rug.nl/research/portal>. For technical reasons the number of authors shown on this cover page is limited to 10 maximum.

Case report - Congenital

Large common left and right coronary artery to coronary sinus fistula

Elke S. Hoendermis^{a,*}, Tjalling W. Waterbolk^b, Tineke P. Willems^c, Felix Zijlstra^a^aDepartment of Cardiology, Thoraxcenter, University Medical Center Groningen, University of Groningen, Hanzeplein 1, P.O. Box 30001, 9700 RB, The Netherlands^bDepartment of Thoracic Surgery, University Medical Center Groningen, The Netherlands^cDepartment of Radiology, University Medical Center Groningen, The Netherlands

Received 16 March 2006; received in revised form 28 July 2006; accepted 30 July 2006

Abstract

Coronary fistulas are vascular anomalies which in rare cases can cause hemodynamic problems with indication for intervention. We report about a 47-year-old man with a large coronary fistula arising from both, the left and right coronary artery. To our knowledge this is the first case described with this anatomy. The main coronary arteries were united at the crux cordis and drained through the coronary sinus into the right atrium. As a consequence of the longstanding volume overload the coronary arteries were extremely dilated. Also, both ventricles were dilated. Therefore, although the patient was asymptomatic, the obvious compromise of the ventricles due to volume overload was regarded as an indication for surgical intervention.

© 2006 Published by European Association for Cardio-Thoracic Surgery. All rights reserved.

Keywords: Congenital; Congestive heart failure; Coronary fistula

1. Case report

A 47-year-old man working on an off-shore dredge rig was referred for cardiomegaly found inadvertently on an X-ray, performed because of a rib contusion. He was asymptomatic and did not use any medication. A positive family history was his only risk factor for atherosclerosis and there was no family history of congenital heart disease, cardiomyopathy or sudden death. Physical examination revealed a continuous heart murmur at the left sternal border, third and fourth intercostal space.

The ECG showed sinus rhythm, a normal frontal axis, high voltages in V4–6, and normal repolarisation. Echocardiography showed a severely dilated, hyperkinetic left ventricle and mild dilation of both atria and the right ventricle. Echocardiography revealed flow in these cavities and one connection to the right atrium was found (Fig. 1b). To establish the diagnosis of a coronary fistula, a contrast-enhanced 64 slice-multidetector computer tomography (MDCT) and a cardiac catheterisation were performed. The MDCT (Fig. 2 and Video 1) demonstrated enormously dilated, tortuous left and right coronary arteries (20 mm at their origin), which merged at the crux cordis. A common entry into the coronary sinus was found from where an enormous flow drained into the right atrium. The calculated left to right shunt was 48% of the total

pulmonary flow (pulmonary to systemic flow was 2 to 1). Pulmonary hypertension was excluded. All coronary side branches had a normal caliber.

Nuclear imaging with TC-99M-Tetrofosmin showed no abnormalities indicative of myocardial ischaemia, and the patient had a normal exercise capacity.

The patient was operated through a median sternotomy, without the use of cardiopulmonary bypass. The left and right coronary arteries were ligated directly proximal to the point where they merged and entered the coronary sinus. The patient was treated postoperatively initially with heparin and started with acenocoumarol. There were no complications. Two months after ligation the echocardiogram showed a normalisation of the diameters of the left and right ventricle. Flow in the dilated left and right coronary arteries was still present, but had changed from

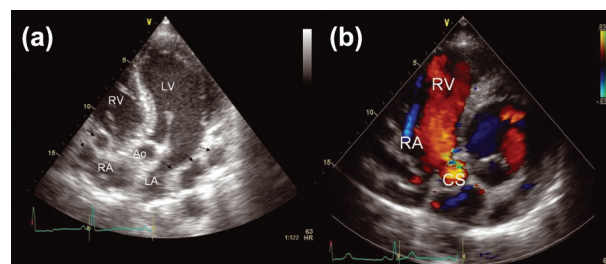


Fig. 1. (a) Echocardiogram apical 5-chamber view of the dilated left ventricle (LV) and right ventricle (RV) and dilated coronary arteries (arrows), RA = right atrium, LA = left atrium. (b) Parasternal short axis view. Fistula entering the right atrium (RA) via coronary sinus (CS).

*Corresponding author. Fax: +31 50 3614884.

E-mail address: e.s.hoendermis@thorax.umcg.nl (E.S. Hoendermis).

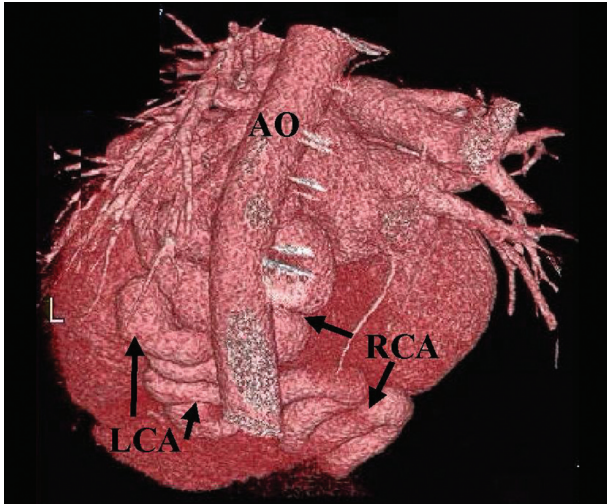


Fig. 2. MSCT-scan demonstrating the extremely enlarged coronary arteries merging at the posterior side of the heart at the crux cordis. RCA=right coronary artery, LCA=left coronary artery, AO=Aorta.

a turbulent to a laminar pattern, demonstrating a normalised flow velocity. The coronary sinus also showed laminar flow. The patient is currently in a good condition and has started working again off shore using a self-control system for INR measurement.

2. Discussion

Coronary fistulas are rare vascular anomalies. Usually they consist of a mostly small, abnormal communications between a coronary artery and a cardiac chamber. In two-thirds of the cases fistulas arise from the right coronary artery [6]. More than 90% enter the right atrium, the coronary sinus or the right ventricle [6]. Large volume shunt flow may cause aneurysmal enlargement of the feeding coronary artery and dilation and/or functional compromise, not only of the receiving cardiac chamber, but also of other chambers as a consequence of the high volume circulation. Pulmonary hypertension may occur.



Video 1. Enormously dilated left and right coronary artery uniting at the crux cordis and feeding a common fistula into the coronary sinus.

Myocardial ischemia may be induced by the steal phenomenon. Other complications can be infective endocarditis, thromboembolic events and in rare cases rupture of the fistula [1,4,5].

Most coronary fistulas are small and do not need therapeutic intervention, however, surgical or catheter intervention is indicated, when the patient is symptomatic or when a shunt is particularly large, and thus associated with risk of complications. Surgical intervention for coronary fistulas, performed in later adult life, carries higher morbidity and mortality than in young adults [3,7]. In asymptomatic patients with large fistulas the decision between interventional and conservative treatment has to be taken individually. Percutaneous intervention with transcatheter coil occlusion is increasingly the therapeutic intervention of choice [3,5], but was no option in this case because of the excessive size and the site of the shunt. The left and right ventricle have returned to normal size within two months after surgical intervention. In this case it was remarkable that both main coronary arteries were feeding the same fistula. Nevertheless, the myocardial blood supply through coronary branches of normal caliber was not impaired. Our main concern was the risk of thromboembolic complications postoperatively after reducing the high flow velocity in extremely dilated coronary arteries. For that reason we started anticoagulation with acenocoumarol immediately after closure of the shunt, hoping that this regimen would minimise the risk, although it is not supported by clear evidence that acenocoumarol is superior to antiplatelet therapy in aneurysmal coronary artery disease [8]. In the presented case surgical intervention could restore a normal coronary and systemic circulation in a patient with a large fistula arising from both the left and right coronary artery.

References

- [1] Valvuranakis M, Bush CA, Boudoulas H. Coronary artery fistulas in adults: incidence, angiographic characteristics, natural history. *Catheter Cardiovasc Diagn* 1995;35:116-120.
- [2] Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levine FH. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature, and delineation of management. *Circulation* 1997; 99:849-854.
- [3] Liberthson RR. Congenital anomalies of the coronary arteries. In: Gatzoulis MA, Webb GD, Daubeney PEF. *Diagnosis and management of adult congenital heart disease*. Churchill Livingstone, 2003:425-431.
- [4] Tirilomis T, Aleksic I, Busch T, Zenker D, Ruschewski W, Dalichau H. Congenital coronary artery fistulas in adults: surgical treatment and outcome. *Int J Cardiol* 2005;98:57-59.
- [5] Perry SB, Rome J, Keane JF, Baim DS, Lock IE. Transcatheter closure of coronary artery fistulas. *J Am Coll Cardiol* 1992;20:201-209.
- [6] Kouchoukos NT, Blackstone EH, Doty DB, Hanly FL, Karb RB. Congenital anomalies of the coronary arteries: coronary arteriovenous fistula. In: Kirklin, Barratt-Boyes *Cardiac Surgery*. 3rd edition. Churchill Livingstone 2003;1241-242.
- [7] Kamiya H, Yasuda T, Nagamine H, Sakakibara N, Nishida S, Kawasuji M, Watanabe G. Surgical treatment of congenital coronary artery fistulas: 27 years experience and a review of the literature. *JK Card Surg* 2002;17:173-177.
- [8] Hart JJ, Joslin CG. Coronary artery ectasia. *Kans Med* 1998;98:6-9.