

An incidental finding of a bicuspid aortic valve and pseudocoarctation of the descending aorta in a patient presenting with an acute coronary syndrome: a case report

Hari Vivekanantham *, Stéphane Cook , and Jean-Christophe Stauffer 

Department of Cardiology, University and Hospital of Fribourg, Chemin des Pensionnats 2, 1708 Fribourg, Switzerland

Received 15 January 2020; first decision 12 February 2020; accepted 9 April 2020; online publish-ahead-of-print 15 May 2020

Background

Aortic pseudocoarctation (PsCoA) is an elongation of the supra-isthmic aorta with kinking and low-grade narrowing. Consequently, no collateral circulation is found. It is frequently associated with other congenital heart anomalies, such as bicuspid aortic valve (BAV).

Case summary

We report the case of 60-year-old patient who presented to the ED with acute chest pain. Physical examination was remarkable for an elevated blood pressure (BP) and the presence of a systolic murmur on the left lower sternal border. An acute coronary syndrome was suspected and the patient underwent urgent coronary angiogram which showed an occluded obtuse marginal artery. Additionally, the presence of an aortic anomaly was noted during the aortography, and additional work-up, including a thoracic computed tomography angiography and transoesophageal echocardiography, revealed a BAV and a PsCoA of the descending aorta. The patient was discharged after optimizing BP control with oral medication and periodical follow-up was arranged.

Discussion

Aortic PsCoA may mimic true coarctation (CoA), but the absence of a haemodynamically significant descending aortic narrowing and of the typical clinical findings associated with aortic CoA, distinguishes both entities. Treatment is therefore conservative and based on the control of cardiovascular risk factors and in particular of arterial hypertension. Work-up should include imaging of the entire aorta and the search for associated congenital cardiac anomalies, which, if present, should be managed in consequence.

Keywords

Bicuspid aortic valve • Aortic pseudocoarctation • Aortic coarctation • Secondary hypertension • Acute coronary syndrome • Case report

Learning points

- Aortic pseudocoarctation is a rare entity, distinguished from true coarctation (CoA) by the absence of haemodynamically significant narrowing of the descending aorta and of the typical clinical findings found in aortic CoA.
- Associated congenital cardiac anomalies, such as bicuspid aortic valve, are frequent and should be looked for.
- Management is conservative and should focus on the optimal control of arterial hypertension.

* Corresponding author. Tel: +41 26 306 23 81, Email: hari.vivekanantham@h-fr.ch

Handling Editor: Monika Arzanauskaite

Peer-reviewers: Nikolaos Bonaros, Christoph Jensen, and Christoph Sinning

Compliance Editor: Max Sayers

Supplementary Material Editor: Ross Thomson

© The Author(s) 2020. Published by Oxford University Press on behalf of the European Society of Cardiology.

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact journals.permissions@oup.com

Introduction

Aortic pseudocoarctation (PsCoA) is a rare entity, belonging to the same spectrum of disease as aortic coarctation (CoA). As the haemodynamic significance of both of these diseases is different, management is accordingly not the same. Caregivers should be aware that these entities do not only concern a paediatric population as diagnosis can be missed until adulthood, in particular in the case of PsCoA.¹ Due to the frequent association with other congenital heart anomalies, such as bicuspid aortic valve (BAV), which increases the risk of serious cardiovascular complications, prompt detection and management is essential.

Timeline

Time	Events
Admission (Day 0)	<p>Presentation at the emergency department with retrosternal chest pain since the previous day</p> <p>The patient had symmetrically elevated blood pressure (BP) and a 3/6 systolic murmur with point maximum at the third left lower sternal border on cardiac auscultation</p> <p>The 12-lead electrocardiogram and lab results were suggestive of cardiac ischaemia</p>
Admission + 3 h	<p>Coronary angiography showed an occlusion of the obtuse marginal artery for which a strategy of conservative management was chosen</p> <p>Aortography showed a tortuous descending thoracic aorta (DA) as well as a dilated ascending aorta (AA) with an abnormal aortic valve opening</p>
Admission + 4 h	<p>Thoracic computed tomography angiography ruled out an aortic dissection and confirmed the presence of a 'kinking' of the DA distal to the subclavian artery</p>
Admission + 4.5 h	<p>Admission to the intensive care unit</p>
Day 1	<p>Transthoracic and transoesophageal echocardiography:</p> <ul style="list-style-type: none"> • Left ventricular hypertrophy with preserved left ventricular ejection fraction and no wall motion abnormalities • Bicuspid aortic valve with mild stenosis and mild regurgitation • Dilated AA
Day 7	<p>Patient discharged after optimizing BP control with oral medication</p> <p>Periodical follow-up arranged</p>

Case presentation

A 60-year-old south-Asian male, with past medical history of poorly controlled blood pressure (BP) under an angiotensin II receptor

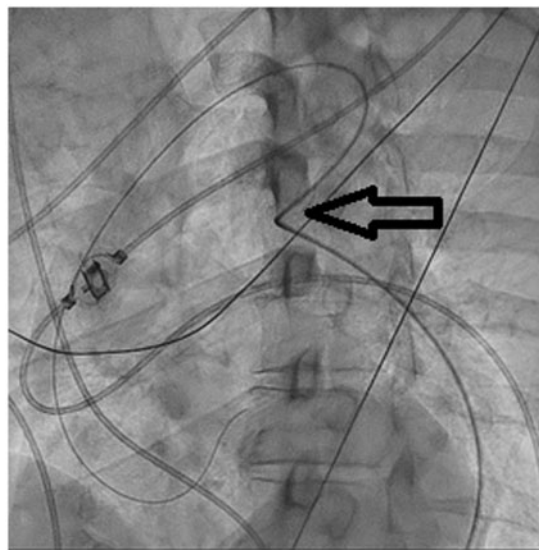


Figure 1 Fluoroscopic image, taken during the coronary angiography, showing the guidewire having an acute angle (arrow) as it was advanced in the descending aorta from a right femoral artery access.

antagonist (sartan), presented to the emergency department complaining of retrosternal chest pain that appeared the previous day. Vital parameters were relevant for elevated BP (left arm 178/108 mmHg, right arm 172/100 mmHg). Physical examination was remarkable for a 3/6 systolic murmur with point maximum at the third left lower sternal border. No radial-radial (RR) or radio-femoral (RF) delay was found. The dorsalis pedis and posterior tibial artery pulses were palpable bilaterally. Bilateral upper limb and thigh BP measurement did not show any discrepancies and the patient did not report any claudication. The 12-lead electrocardiogram showed a complete right bundle branch block and left anterior hemiblock, as well as negative T-waves from V4 to V6 suggestive of ischaemia ([Supplementary material online, Figure A](#)). High-sensitive cardiac Troponin T was elevated at 495 ng/L (normal value: <14 ng/L). As an acute coronary syndrome (ACS) was evident, the patient underwent urgent coronary angiogram through a right femoral artery access. The exam revealed an occlusion of the obtuse marginal artery ([Supplementary material online, Videos S1 and S2](#)). Due to the small calibre of the vessel and the absence of functional reperfusion in the ventriculography, a strategy of conservative management was chosen.

Furthermore, the descending thoracic aorta (DA) was noted to be tortuous when advancing the guidewire ([Figure 1](#)). Aortography confirmed this finding and additionally showed a dilated ascending aorta (AA) with an abnormal aortic valve opening ([Supplementary material online, Videos S3 and S4](#)). A thoracic computed tomography (CT) angiogram performed subsequently ruled out an aortic dissection and confirmed the presence of a 'kinking' of the DA distal to the subclavian artery, with a <50% of narrowing relative to the diameter of the aorta at the diaphragm level ([Figure 2A–C](#)). The diameter of the AA was measured at 40 mm. The patient was subsequently admitted to the intensive care unit.

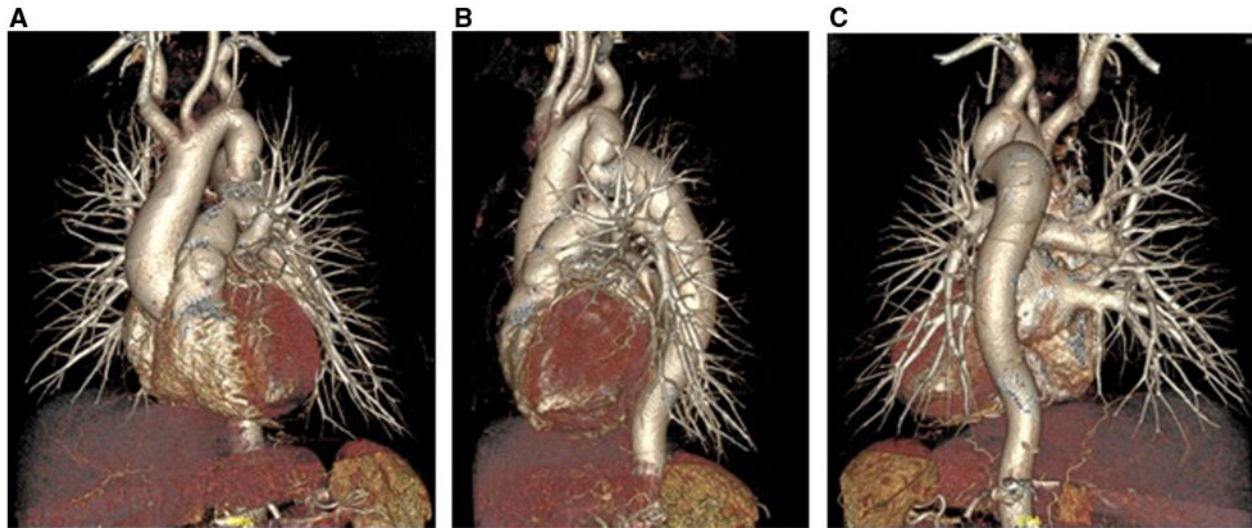


Figure 2 (A–C) Volume-rendered 3D reconstructions of a thoracic computed tomography angiogram showing a tortuosity or 'kinking' of the descending aorta, distal to the left subclavian artery, without any developed collateral circulation.

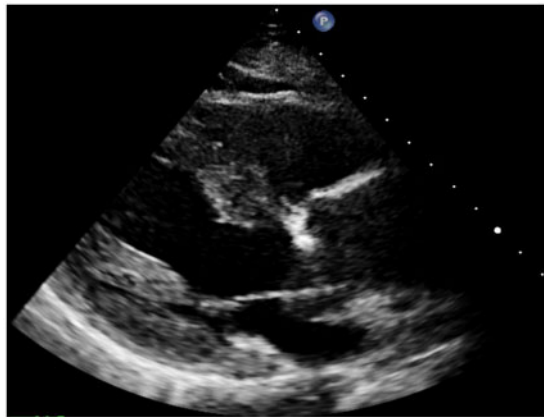


Figure 3 Transthoracic echocardiography parasternal long-axis view showing hypertrophy of the anteroseptal and inferolateral left ventricular walls, as well as a calcified aortic valve.



Figure 4 Transoesophageal echocardiography short-axis view of the aortic valve showing a Type I L-R (raphe between the left and right coronary sinus) bicuspid aortic valve.

A transthoracic echocardiography performed on the following day showed a concentric left ventricular hypertrophy (LVH) without any wall motion abnormalities (*Figure 3*) (*Supplementary material online, Videos S5 and S6*). The aortic valve was severely calcified with mild stenosis and mild regurgitation. The diameter of the AA was measured at 40 mm as well. A transoesophageal echocardiography performed later confirmed a Type I L-R BAV (raphe between the left and right coronary sinus) according to the Sievers classification,² with a planimetric opening area of 1.8 cm² (*Figure 4*) (*Supplementary material online, Video S7*). The narrowest diameter of the DA was measured at 11 mm as in the CT (*Figure 5*) (*Supplementary material online, Video S8*). As the angle

between the proximal and distal part of the DA was too acute, no Doppler gradients were measurable.

The patient could be discharged a week following admission under dual antiplatelet therapy, statin, low-dose sartan, and low-dose beta-blocker, with optimal BP control (at discharge: left arm 132/78 mmHg, right arm 128/70 mmHg) under this regimen.

Discussion

The clinical findings in our patient, especially the absence of a pulse delay (RR and RF) and BP discrepancies between the four extremities, suggested the absence of significant blood flow obstruction through the kinked DA. Consolidating this hypothesis, no collateral

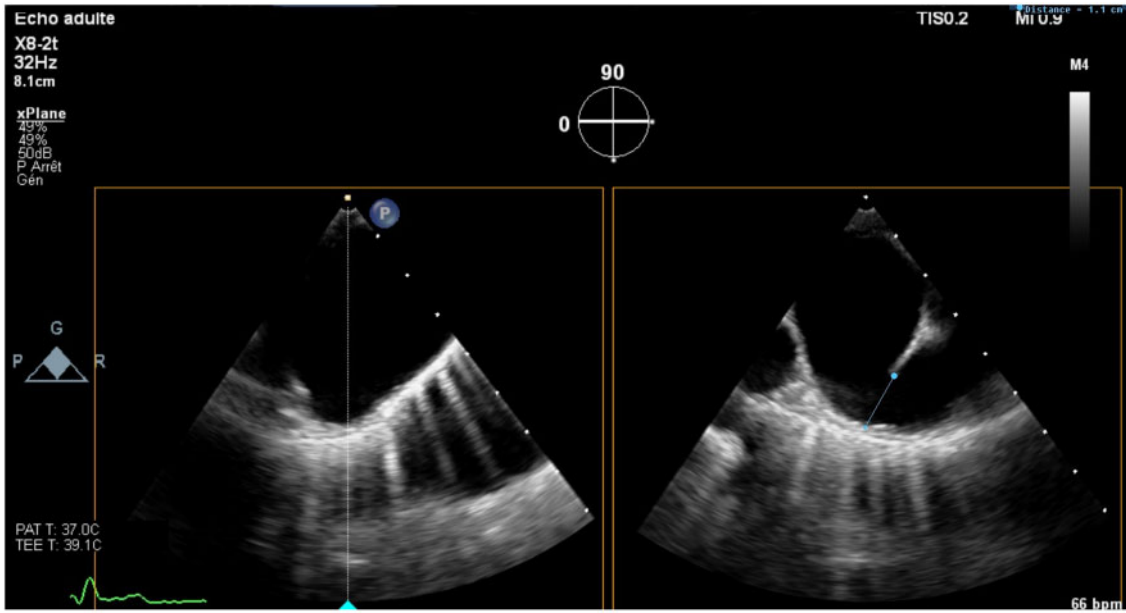


Figure 5 Transoesophageal echocardiography using simultaneous biplane imaging (X-plane) showing the narrowing of the descending aorta, measured at 11 mm.

circulation was found in the thoracic CT angiography. All in all, the diagnosis of PsCoA was made.

Coarctation and PsCoA are part of a common spectrum of disease, with the difference being the functional repercussion of the aortic narrowing. While true CoA is defined by congenital narrowing of the DA at the aortic isthmus with clinical features such as upper body systolic hypertension, lower body hypotension, a BP gradient between upper and lower extremities, RF pulse delay, and palpable collateral circulation,³ these are typically not found in PsCoA.¹ The mean age of diagnosis of PsCoA is 43 years,¹ while CoA may go undiagnosed as late as in the adult life if it is not severe or if collateral circulation develops. Untreated CoA can lead to a compensatory LVH due to the increased afterload and hence to hypertensive vascular complications like premature coronary artery disease.³

Bicuspid aortic valve is the most common congenital heart disease with a prevalence of 1–2%.⁴ The prevalence of CoA is of 0.03%³ and that of PsCoA is unknown but probably even less, as a recent systematic review¹ only reported 18 published cases of PsCoA in the adult population. Both CoA and PsCoA have reported association with BAV.^{4–9} The Type I L-R BAV is the most common pattern¹⁰ and is additionally the only type reported in CoA.¹¹ About 50–75% of patients with CoA have a BAV, whereas only 7% of patients with BAV have a concomitant CoA.⁴ This association increases the risk of developing aortic dilatation and dissection, compared to BAV alone. If an association between a specific type of BAV with PsCoA exists or not is, to our knowledge, unknown.

Specific guidelines regarding management of PsCoA are lacking, and therefore management should follow the 2010 ESC guidelines on true CoA.^{3,12} Indications for repair are a non-invasive pressure difference >20 mmHg between the upper and lower limbs with upper limb hypertension, pathologic BP response to exercise or LVH [class

of recommendation (CoR) I, level of evidence (LoE) A]. In hypertensive patients with a $\geq 50\%$ narrowing of the aortic lumen relative to the reference aortic diameter at the diaphragm level, intervention should be considered (CoR IIa, LoE C). The invasive measurement of a peak-to-peak gradient across the CoA orifice by catheterization, with a value of >20 mmHg being haemodynamically relevant, is not considered in the 2010 ESC guidelines. Concerning the management of BAV with associated aortic root dilatation, the 2014 ESC guidelines⁴ recommend surgery of the AA in case of an aortic root or AA diameter of >45 mm if aortic valve replacement is planned (CoR I, LoE C), or if the diameter is >50 mm with the presence of risk factors such as CoA or hypertension (CoR I, LoE C).

Our patient had LVH as a sign of increased afterload. To determine if he is suffering from essential hypertension or secondary hypertension due to the PsCoA or both, is difficult. The fact that BP could be adequately controlled under low-dose sartan/beta-blocker bi-therapy is reassuring but does not support any of the aetiologies. A regular monitoring of BP at home with a target of <140/90 mmHg as per the 2018 ESC guidelines was advised.¹³ As the risk of aortic dilatation and progression to aneurysm formation and eventually rupture is even higher in this patient due to the combination of a BAV, AA dilatation, and PsCoA, a stricter BP target of <130/80 mmHg should, in our opinion, be considered. Avoidance of isometric exercise comprising a high static load was pragmatically recommended (CoR III, LoE C, if aortic >40 mm in the presence of a BAV or elastopathy). Screening for BAV of first-degree relatives was advised (CoR IIa, LoE C).

A beta-blocker therapy was begun in the setting of the ACS. Of note, it may also be considered in patients with BAV and dilated aortic root >40 mm (CoR IIb, LoE C). Beta-blockers and sartans—in particular losartan—have shown efficacy in preventing progression of

aortic root dilatation in patients with Marfan syndrome^{14,15} and their use is therefore extended to other aetiologies.

A periodical clinical and echocardiographic follow-up, including a baseline imaging assessment of the entire aorta (CoR I, LoE C), was scheduled. Annual surveillance of the aorta for the development of an aortic aneurysm is recommended.¹ An uncontrollable hypertension or progression of the LVH, despite adequate medical therapy, should prompt a surgical correction of the PsCoA, as the contribution of the latter to the increased afterload would, in this case, be manifest.

Conclusion

Pseudocoarctation is a rare diagnosis which shares a common spectrum of disease with CoA. It consists of a 'non-severe' CoA, with lower impact on the left ventricular afterload and hence lacks adaptive mechanisms such as the development of collateral circulation. The presence of concomitant BAV or of other congenital heart diseases is not unusual, and should be investigated. As specific guidelines for the management of PsCoA are lacking, decisions should be made based on a case by case evaluation, following the 2010 ESC guidelines on CoA.³

One should bear in mind that CoA, and PsCoA by extension, are rare causes of secondary hypertension¹⁵ that should be considered in case of a suggestive clinical picture. Prompt detection and management can prevent deadly cardiovascular complications.

Lead author biography



Dr Hari Vivekanantham has achieved his medical studies at the University of Geneva, Switzerland, in 2014. He underwent 3 years of residency in internal medicine followed, as of 2017, by a 2-year residency in the cardiology department of the Inselspital - University and Hospital of Bern, Switzerland. He is currently achieving his cardiology residency in the cardiology department of the University and Hospital

of Fribourg, Switzerland. He has particular interest in electrophysiology and pacing.

Supplementary material

[Supplementary material](#) is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

References

- Singh S, Hakim FA, Sharma A, Raina Roy R, Panse PM, Chandrasekaran KB, Alegria JR, Mookadam F. Hypoplasia, pseudocoarctation and coarctation of the aorta—a systematic review. *Heart Lung Circ* 2015;**24**:110–118.
- Sievers HH, Schmidtke C. A classification system for the bicuspid aortic valve from 304 surgical specimens. *J Thorac Cardiovasc Surg* 2007;**133**:378–388.
- Baumgartner H, Bonhoeffer P, De Groot NMS, de Haan F, Deanfield JE, Galie N, Gatzoulis MA, Gohlke-Baerwolf C, Kaemmerer H, Kilner P, Meijboom F, Mulder BJM, Oechslin E, Oliver JM, Serraf A, Szatmari A, Thaulow E, Vouhe PR, Walma E; Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC); Association for European Paediatric Cardiology (AEPIC); ESC Committee for Practice Guidelines (CPG). ESC Guidelines for the management of grown-up congenital heart disease (new version 2010): the Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC), endorsed by the Association for European Paediatric Cardiology (AEPIC). *Eur Heart J* 2010;**31**: 2915–2957.
- Erbel R, Aboyans V, Boileau C, Bossone E, Di Bartolomeo R, Eggebrecht H, Evangelista A, Falk V, Frank H, Gaemperli O, Grabenwöger M, Haverich A, Iung B, Manolis AJ, Meijboom F, Nienaber CA, Roffi M, Rousseau H, Sechtem U, Sirnes PA, von Allmen RS, Vrints C. 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases: the Task Force for the diagnosis and treatment of aortic diseases of the European Society of Cardiology (ESC). *Eur Heart J* 2015; **36**:2779–2926.
- Lochaya S, Kaplan B, Shaffer AB. Pseudocoarctation of the aorta with bicuspid aortic valve and kinked left subclavian artery: a possible cause of subclavian steal. *Am Heart J* 1967;**73**:369–374.
- Kavanagh-Gray D, Chiu P. Kinking of the aorta (pseudocoarctation): report of six cases. *Can Med Assoc J* 1970;**103**:717–720.
- Angelini GD, Kulatilake EN, Hayward M, Ruttley MS. Pseudocoarctation of the aortic arch associated with bicuspid aortic valve lesion: is it a surgical entity? *Thorac Cardiovasc Surg* 1985;**6**: 36–37.
- Tung R, Siegel RJ. Aortic pseudocoarctation associated with a stenotic congenitally bicuspid aortic valve. *Am J Cardiol* 2007;**100**:157–158.
- Cordeiro F, Silva Carvalho S, Ferreira A, Moreira JL. Aortic pseudocoarctation: a very rare finding. *BMJ Case Rep* 2018;**2018**:bcr-2017-222924. doi: 10.1136/bcr-2017-222924.
- Lung B. Genetic diseases of the aorta: aortic diseases related to Marfan syndrome and other genetic abnormalities. In AJ Camm, TF Lüscher, G Maurer, PV Serruys, eds. *The European Society of Cardiology (ESC) Textbook of Cardiovascular Medicine*, Vol. 2. 3rd ed. Oxford: Oxford University Press; 2019. p2609–2611.
- Ciotti GR, Vlahos AP, Silverman NH. Morphology and function of the bicuspid aortic valve with and without coarctation of the aorta in the young. *Am J Cardiol* 2006;**98**:1096–1102.
- Sondergaard L, Charakida M. Coarctation. In AJ Camm, TF Lüscher, G Maurer, PV Serruys, eds. *The European Society of Cardiology (ESC) Textbook of Cardiovascular Medicine*, Vol. 1. 3rd ed. Oxford: Oxford University Press; 2019. p810–814.
- Williams B, Mancia G, Spiering W, Agabiti Rosei E, Azizi M, Burnier M, Clement DL, Coca A, de Simone G, Dominiczak A, Kahan T, Mahfoud F, Redon J, Ruilope L, Zanchetti A, Kerin M, Kjeldsen SE, Kreutz R, Laurent S, Lip GYH, McManus R, Narkiewicz K, Ruschitzka F, Schmieder RE, Shlyakhto E, Tsioufis C, Aboyans V, Desormais I; ESC Scientific Document Group. 2018 ESC/ESH Guidelines for the management of arterial hypertension: the Task Force for the management of arterial hypertension of the European Society of Cardiology (ESC) and the European Society of Hypertension (ESH). *Eur Heart J* 2018;**39**:3021–3104.
- Chiu HH, Wu MH, Wang JK, Lu CW, Chiu SN, Chen CA, Lin MT, Hu FC. Losartan added to beta-blockade therapy for aortic root dilation in Marfan syndrome: a randomized, open-label pilot study. *Mayo Clin Proc* 2013;**88**:271–276.
- Shores J, Berger KR, Murphy EA, Pyeritz RE. Progression of aortic dilatation and the benefit of long-term beta-adrenergic blockade in Marfan's syndrome. *N Engl J Med* 1994;**330**:1335–1341.