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Images in Cardiovascular Medicine

Right Coronary Artery Narrowing by Stretching on Aortic Root Aneurysm

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Case Description

A 35-year-old woman with Marfan syndrome presented to the emergency department reporting a 2-day history of chest pain and dyspnea. Transthoracic echocardiography revealed grade IV/IV aortic valve regurgitation, a markedly dilated aortic root, increased systolic and diastolic internal dimensions (49/67 mm) of the left ventricle, and a mild decrease in left ventricular ejection fraction (51%) in the Teicholz measurement. An enhanced computed tomography (CT) scan showed a 9.0-cm diameter in the sinotubular junction and a 9.9-cm diameter in the sinus level. Computed tomography coronary angiograms showed severe proximal stenosis of the right coronary artery (RCA) (Fig. 1A and B). The next day, an urgent modified button Bentall procedure was performed with a 23-mm On-X mechanical valve conduit (On-X Life Technologies) for the aortic root aneurysm. The dilated right aortic sinus pulled the RCA ostium upward (Fig. 1C). The RCA ostium was mildly stenotic compared with the artery lumen but freely passed through a 3.5-mm dilator (Fig. 2A). Although a longstanding stretch elongated the RCA, the artery was not redundant after the modified button Bentall procedure (Fig. 2B). Postoperative CT coronary angiograms showed minimal ostial stenosis without stenosis of the proximal RCA (Fig. 2C). In a 6-month postoperative echocardiogram, the left ventricular ejection fraction had improved from 38% immediately after surgery to 51%, and the systolic and diastolic dimensions of the left ventricle decreased to 40 and 53 mm, respectively.

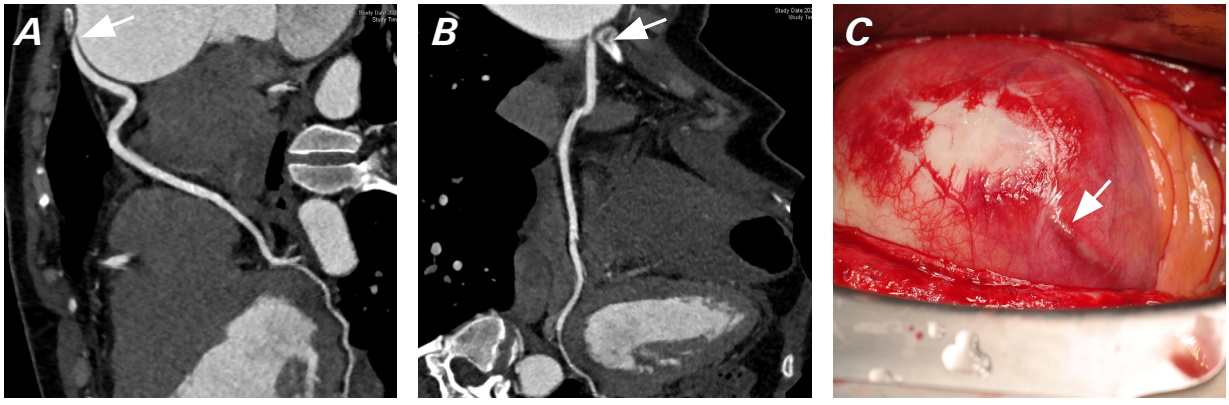

Fig. 1 A) and **B)** Preoperative computed tomography coronary angiogram shows a proximal RCA stenosis. **C)** Operative findings show the stretched RCA on the dilated sinus.

RCA, right coronary artery.

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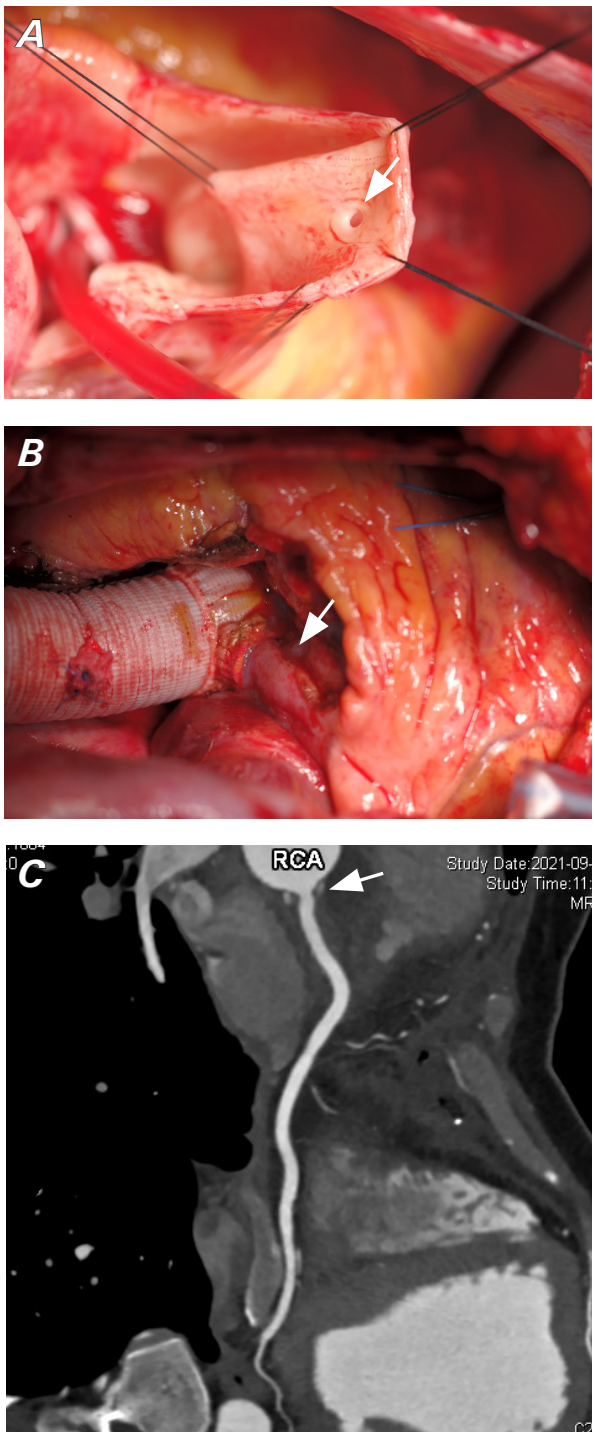

Fig. 2 A) Operative findings are a patent right coronary ostium and **B)** suitable length of the right coronary button (white arrow). **C)** Postoperative computed tomography coronary angiogram shows minimal stenosis of the RCA.

RCA, right coronary artery.

Abbreviations and Acronyms

CT	computed tomography
RCA	right coronary artery

Comment

Coronary diseases are rarely associated with Marfan syndrome, but there are reports of atherosclerotic disease[1] and aneurysms[2] associated with it. Other causes of proximal coronary stenosis—an ectopic origin and intramural coronary artery,[3] which are congenital lesions—can be considered, but the coronary anomaly was ruled out because of the position of the right coronary ostium and the absence of previous symptoms of ischemia.

Coronary CT angiography, echocardiography, and patient histories may be unreliable for diagnosing coronary artery anomalies.[3] In such patients, intravascular ultrasonography may be necessary to more accurately diagnose proximal RCA lesions.

The team planned the modified button Bentall procedure for this case and considered the possibility of a bypass graft for the RCA stenosis. The stretching of the RCA caused by the subcoronary sinus dilatation induced luminal narrowing in the proximal portion of the RCA. The luminal narrowing disappeared with the release of the stretched artery after the Bentall procedure.

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Conflict of Interest Disclosures: None

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