

Unanticipated Pseudocoarctation Highlights the Importance of Visualizing Aortic Arch Anatomy Before Transfemoral Transcatheter Aortic Valve Implantation

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A 71-year-old gentleman with background hypertension, dyslipidemia, ex-smoker, permanent atrial fibrillation, and chronic obstructive lung disease presents to our institution with exertional dyspnea (New York Heart Association class III–IV) for aortic valve replacement assessment. Cardiovascular history includes mechanical mitral valve replacement (St. Jude Medical 25 mm) alongside coronary artery bypass (venous grafts to right coronary artery and obtuse marginal) in 2004, cerebrovascular accident in 2008 with multiple transient ischemic attacks since (latest 2012), and subacute subdural hematoma requiring evacuation (2013).

As part of the transcatheter aortic valve implantation work-up, he underwent a transthoracic echocardiogram, which showed a calcific, degenerative severe aortic stenosis (mean gradient, 45 mmHg; max aortic valve velocity, 4.3 m/s; and aortic valve area, 0.8 cm²) with associated moderate regurgitation. There was preserved left ventricular function with concentric hypertrophy. The mechanical mitral valve was functioning well with a mean transvalvular gradient of 5 mmHg and no paravalvular leaks. There were no prominent mechanical mitral valve pivot guards protruding in the left ventricular outflow tract (Figure 1A–1C). Multislice computed tomography scan revealed a tricuspid aortic valve with extensive fibrocalcific degeneration of the cusps. Annulus perimeter was 68 mm and annulus area was calculated at

3.46 cm². Minimum ilio-femoral diameter was 5.8 cm (both sides). Cardiac computed tomography revealed patent grafts and moderate stenoses at the mid and distal segments of the left anterior descending artery. After discussion with the heart team (Euroscore II, 8.8%; Logistic Euroscore, 22.62%; STS, 7.4%; and STS MOM of 34.9%), decision was made for a transfemoral transcatheter aortic valve implantation of an Edwards SAPIEN 3 (Edwards Lifesciences, Irvine, CA) 23 mm valve.

After obtaining an aortogram via the left radial artery, the operators encountered difficulty in advancing a normal J-wire/pigtail to the root via the right femoral access. Its course was obstructed at the level of the ligamentum arteriosum (Figure 2C). At that time, careful review of the computed tomography revealed a kinking of the aortic arch at that level (Figure 2A and 2B; Movie I in the Data Supplement) with reduction in the isthmus caliber (diameter, 9×14 mm) and associated minimal dilatation of the proximal descending thoracic aorta; there was a posterior origin of the left subclavian artery with associated ectasia at its proximal and medium segments. No collateral circulation or rib notching was seen. Eventually, operators managed to cross the pseudocoarctation using a Storz 0.35" wire (Cordis; Johnson and Johnson, Miami Lakes, FL) and an Amplatz left 1 (Cordis; Johnson and Johnson) catheter (Figure 2D). When crossing the kinking, no

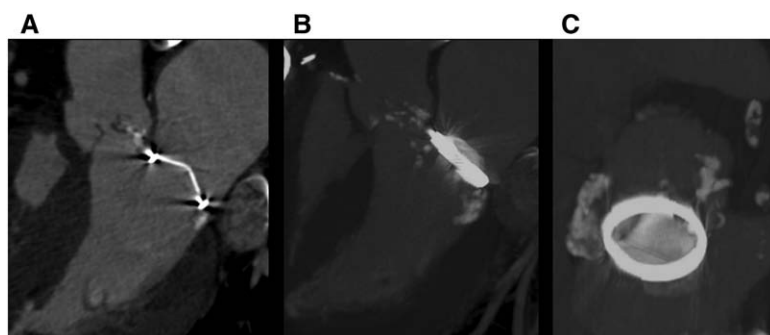


Figure 1. Oblique sagittal (A) and 3-dimensional volume reconstructed (B, oblique sagittal; C, oblique transverse) computed tomographic images demonstrating no significant protrusion of the mechanical mitral valve pivot guards in the left ventricular outflow tract.

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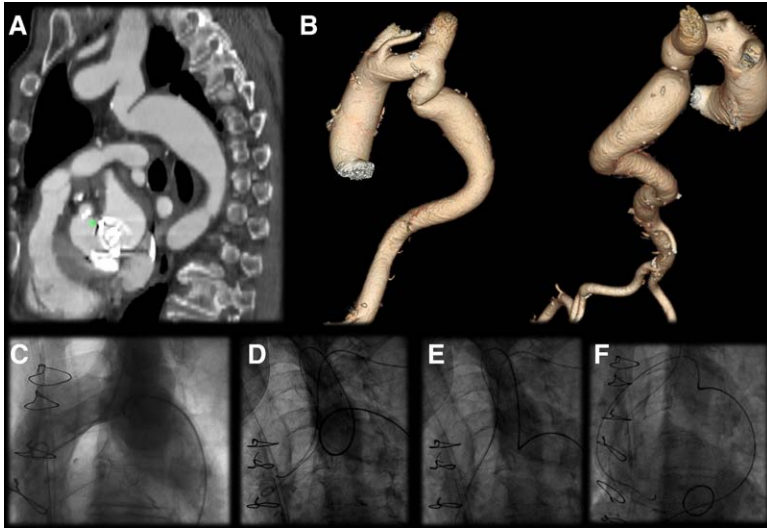


Figure 2. **A**, Sagittal reformatted computed tomographic image of pseudocoarctation shows that arch 1 lies high in the thorax extending to the thoracic inlet. The redundant descending aorta kinks at the level of the ductus ligament and extends into the apparent arch 2. **B**, Three-dimensional reconstruction of the aorta demonstrating the pseudocoarctation. **C**, Aortogram showing the pseudocoarctation and the difficulty in crossing it with a pigtail catheter. **D** and **E**, The pseudocoarctation was eventually crossed with a Storq 0.035'' wire and an Amplatzer left 1 catheter. **F**, The distinctive figure of 3 characteristic of pseudocoarctation has been adapted by the diagnostic AL-1 catheter and Storq wire.

significant gradient was observed. These findings were consistent with the diagnosis of aortic pseudocoarctation. After crossing the aortic valve with the Amplatzer left 1, the Storq wire was exchanged for a Lunderquist extra stiff guidewire (Cook Medical, Bjaaeverskov, Denmark), which straightened the pseudocoarctation and facilitated a relatively easy passage of the SAPIEN 3 Edwards Commander delivery system (Figure 3A and 3B). Before crossing the pseudocoarctation with the valve, the operators advanced a predilatation balloon (without predilating), to assess the manoeuvrability through the pseudocoarctation. The final result was excellent with optimal valve positioning without paravalvular leak (Figure 3C–3F). No difficulties were encountered during delivery system retrieval, using routine steps.

The term pseudocoarctation was introduced in 1952 by Dotter and Steiberg as an aortic lesion similar to coarctation, yielding the same radiological picture, with little or no obstruction. It is a rare anomaly consisting of redundancy of the aortic arch, leading to a tortuous course producing an S-shaped figure. Although it is an isolated anomaly in most cases, operators should be aware of potential associations of pseudocoarctation with anatomic anomalies of the head and neck vessels (prominent tortuosity and kink formation, as

well as dolichoectatic changes in the proximal segments) and cardiac anomalies, including bicuspid aortic valve, aortic stenosis (as in our case), ventricular septal defect, atrial septal defect, patent ductus arteriosus, and sinus of Valsalva aneurysm.¹ Kinking of the aorta and an acute angle between the descending aorta and aortic arch have been described as predisposing factors to fatal descending aorta perforation, caused during the delivery system advancement.² Operators should be cautious when encountering difficulties in advancing the delivery sheath because there is always the possibility the system is caught on atherosclerotic material or one of the head and neck vessels. In cases of complex aortic atherosclerosis or excessive arch tortuosity, a transapical approach should be favored to avoid embolic events or aortic dissection.³

In our case, apart from the pseudocoarctation, a mechanical mitral prosthesis was present. In one of the largest case series⁴ of transcatheter aortic valve implantation in patients with previous mitral valve prosthesis (n=40), device success as per Valve Academic Research Consortium criteria was achieved in 97.5% of cases, suggesting that TAVR can be performed both effectively and safely in the majority of patients with previous mitral prostheses. However, several case reports have demonstrated valve maldeployment and embolization principally

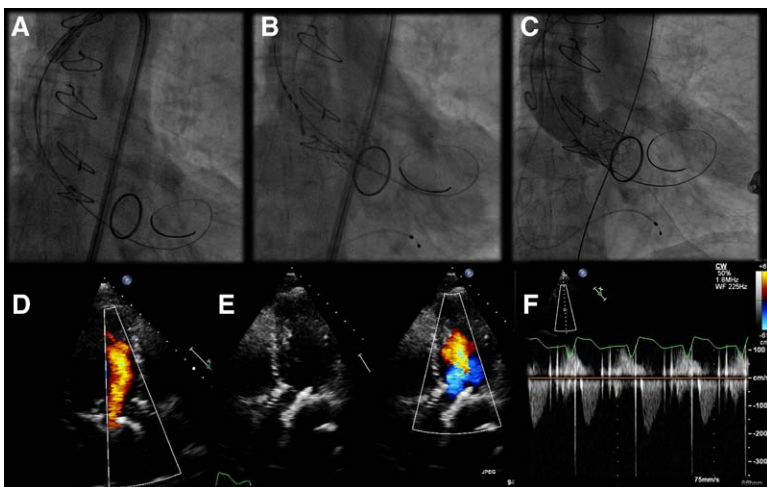


Figure 3. **A**, The pseudocoarctation was eventually straightened using a Lunderquist extra stiff guidewire. This facilitated easy passage of the Edwards Commander delivery system. **B**, Deployment of the Edwards SAPIEN 3 23 mm valve. **C**, Final aortogram showing optimal valve position with no paravalvular leak. **D**, Three-chamber and **(E)** 5-chamber apical transthoracic echocardiographic views after valve implantation demonstrating absence of paravalvular leak. **F**, Continuous wave Doppler across the aortic valve post-transcatheter aortic valve implantation demonstrating a peak gradient of 16 mm Hg (V_{max} , 197 cm/s) and a mean gradient of 9 mm Hg.

when the inflating balloon impinges onto the adjacent mitral prosthesis: specifically, the prominent prosthetic housing cage, the pivot guard (mechanical valve), or the commissural struts (bioprosthesis). Echocardiography and computed tomography are useful in identifying unfavorable characteristics that should raise alarm, such as prominent commissural struts, pivot guard, or rigid housing cage in close proximity to the aortic annulus within the left ventricular outflow tract. In the current case, we did not identify any of these alarming features and indeed the balloon inflation was uncomplicated.

New generation valves with lower profile sheaths and delivery systems facilitate transcatheter aortic valve implantation via the transfemoral route even in patients with challenging anatomies, such as pseudocoarctation. Operators, however, need to pay meticulous attention to the whole length of the aorta before deciding the optimal access site. Attention is often drawn to the femorals, the iliacs, the abdominal aorta, the aortic root and particularly the aortic annulus; the current case highlights the importance of including the aortic arch and descending aorta in our detailed pretranscatheter aortic valve implantation access assessment. If such an assessment had been performed most probably this patient would have undergone transapical valve implantation.

Disclosures

Dr Colombo is a consultant and minor shareholder of Direct Flow Medical (Santa Rosa, CA), Dr Latib serves on the advisory board for Medtronic and is a consultant and proctor for Direct Flow Medical (Santa Rosa, CA), and Dr Montorfano is a proctor for Edwards SAPIEN (Edwards Lifesciences, Irvine, CA). The other authors report no conflicts.

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