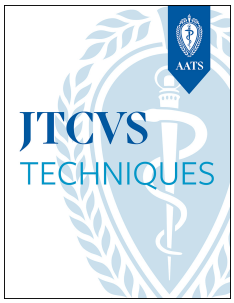

A young marathon runner with severe aortic coarctation and bicuspid aortic valve disease complicated by contained aortic rupture

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A young marathon runner with severe aortic coarctation and bicuspid aortic valve disease complicated by contained aortic rupture

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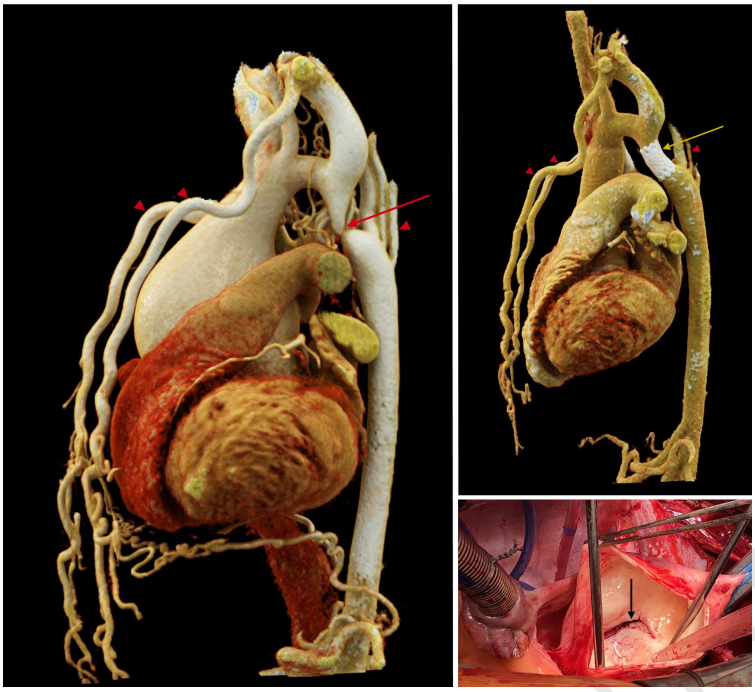
Central Message: 145 characters

Central Picture

Large aortic aneurysm in the presence of aortic coarctation, complicated by intimal tear



Central Message

Treatment of aortic aneurysms and coarctation has to balance risks of retrograde dissection, distal malperfusion and uncontrollable hypertension.

Key words

Aorta, coarctation, dissection, aortic rupture, bicuspid aortic valve

Case Report

A 17-year-old competitive athlete with no prior medical history was admitted after acute onset of chest and back pain. A week prior to the event he took part in a marathon, but was not able to finish due to spasms in his lower extremities.

Physical examination revealed a clinically stable patient with systolic and diastolic heart murmur and arterial hypertension. Echocardiography revealed presence of an aortic root aneurysm and a bicuspid aortic valve (BAV) with severe regurgitation, as well as pericardial effusion. Creatin-kinase (CK) levels were markedly elevated with 3372 U/l (reference value <190 U/l). Systolic pressure gradient between upper and lower extremities was 30 mmHg.

Computed tomography (CT) scanning revealed an 80 mm aneurysm of the ascending aorta with a lesion suggesting contained rupture at the level of the right pulmonary artery, as well as severe coarctation of the aorta distal to the left subclavian artery (Figure 1, panels A and C) with extensive collateral circulation (Figure 1, panels A, E, F).

Emergency surgery comprised aortic root replacement and replacement of the entire ascending aorta using hypothermic circulatory arrest (HCA) for the distal aortic anastomosis. The aortic valve was bicuspid, Sievers type 1 with R-L fusion pattern. A large tear in the posterior aspect of the ascending aorta confirmed the findings of the pre-operative imaging (Figure 1, panel G). Pressure monitoring during surgery was performed via both radial arteries as well as the left femoral artery. While the initial gradient was 30mmHg it dropped to 10mmHg (MAP) during CPB (Supplementary Figure). We prepared for double arterial cannulation (right axillary and femoral artery) but did not introduce a second line as the patient was cooling homogenously and lactate levels were stable.

Given the extensive collateralization, the coarctation was not addressed at the time of surgery. On POD 8, a balloon-expandable covered stent was implanted at the level of the proximal descending aorta, reducing the

systolic pressure gradient to <5 mmHg (Figure 1, panels C and D). Computed tomographic angiography after 4 weeks demonstrated good results (Figure 1, panel B).

Discussion

Our patient exhibited the well-known triad of coarctation, BAV and aortic aneurysm.[1-4] Nevertheless, the presentation was remarkable as he has not only been asymptomatic to this day but even had been performing long-distance running for several years. He frequently complained of spasms in his lower extremities but attributed these to his strenuous training. The markedly elevated CK levels days after running suggests that he was indeed experiencing ischemia when exercising. In a recent study, mean CK levels 24h after a marathon were 1443±1533 U/L, whereas mean levels after 8 days were almost normalized with 166±19 U/L.[5] While many adult patients with coarctation suffer from difficulty to control hypertension, our patient exhibited only moderate blood pressure elevation during exercise testing 6 weeks after the event and monotherapy with an angiotensin receptor blocker.

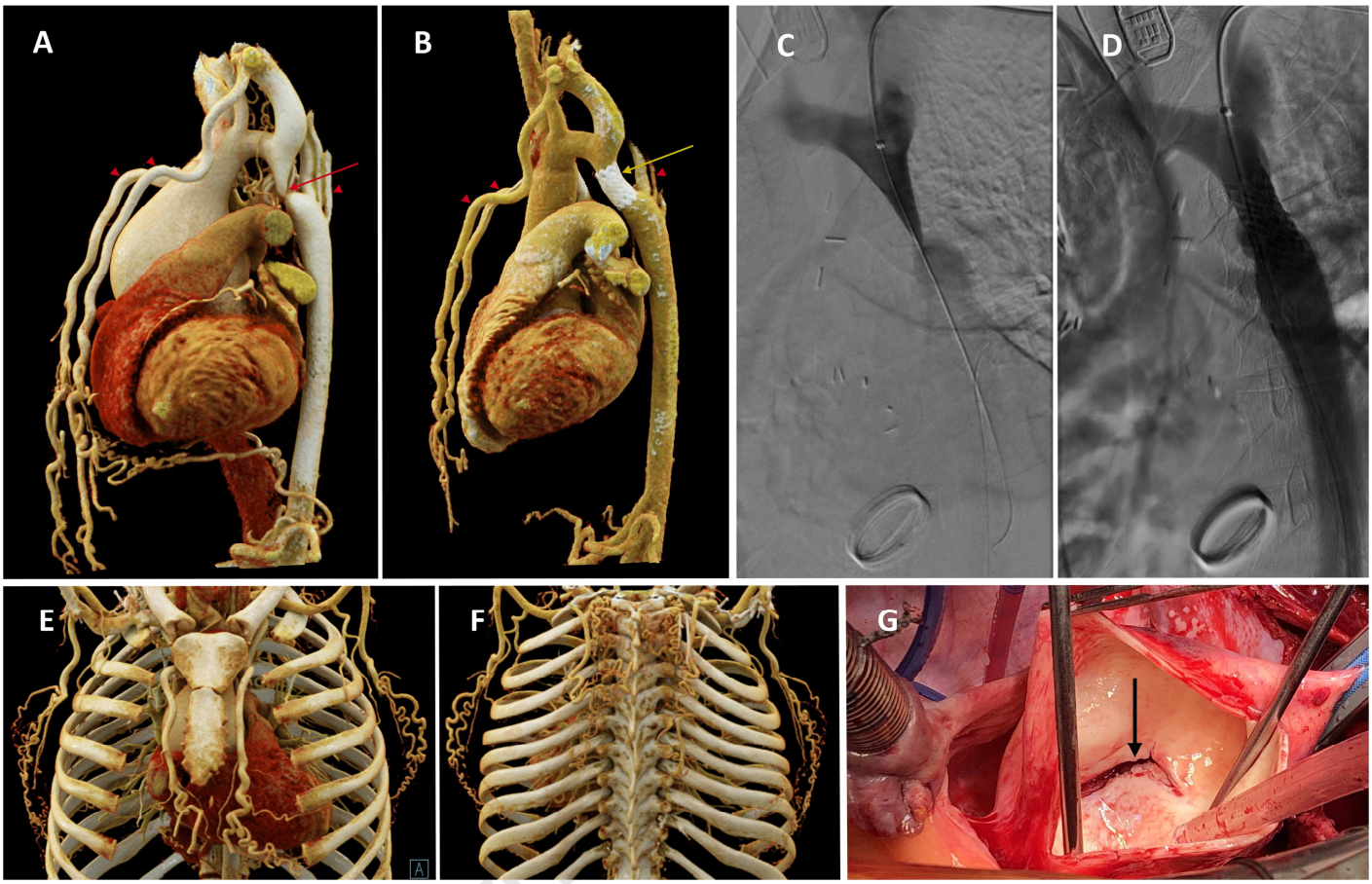
The patient presented with large aneurysm of the aortic root. The distal ascending aorta as well as the aortic arch were not enlarged or dissected but actually rather small. We did not perform a single-stage hybrid approach with stent deployment during HCA as lower body perfusion was not critical and a prolonged HCA could be avoided. A single-stage approach using a frozen elephant trunk (FET) was deemed unsuitable as arch replacement was not necessary and the radial force of the stentgraft portion of the FET would most probably not have been sufficient to open up the coarctation. Furthermore, the smallest FET available was 24mm and the risk of infolding would have been very high. Ascending-descending aortic bypass is usually reserved for situations in which a complex re-do should be avoided and not the procedure of choice in a patient with stenosis of a short segment.

Instead, a two-stage hybrid approach was chosen for addressing the coarctation, as lactate levels declined after re-perfusion was started and it became clear that distal perfusion was sufficient. Delaying the second procedure

in selected cases might be beneficial, e.g. in terms of kidney function. In case the dissection would have extended into the stenosed segment, immediate stenting would have been necessary.

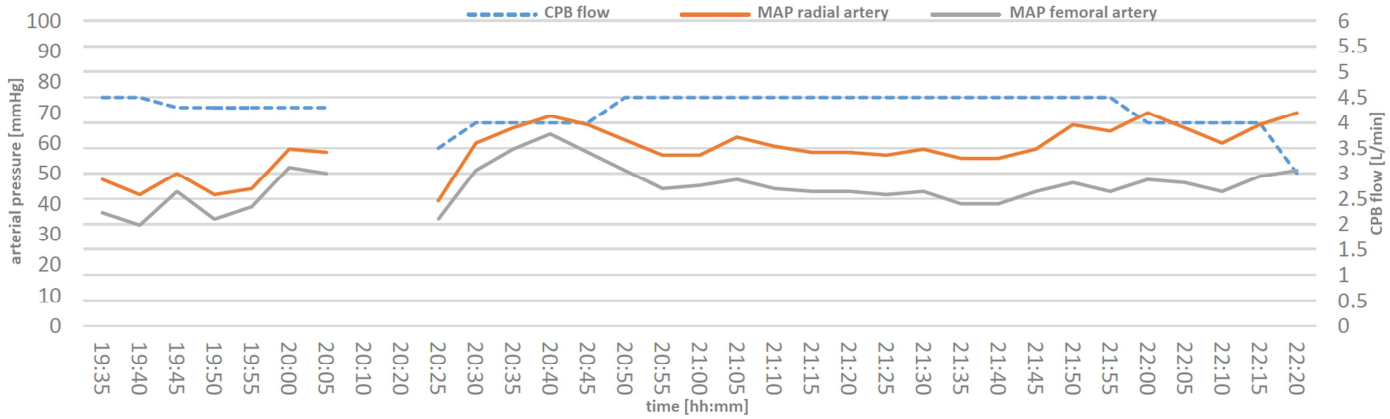
In patients with large aneurysms of the ascending aorta and concomitant aortic coarctation, the risk of retrograde type A dissection during percutaneous intervention of the coarctation has to be balanced against the risk for distal malperfusion during cardiopulmonary bypass and hypothermic circulatory arrest, as well as the difficulty to control hypertension in the perioperative period. Considering a stepwise approach is reasonable.

Figure 1



Panel A: Aortic coarctation (red arrow) with strong collateralization (red arrowheads) and large aneurysm of the ascending aorta. Panel B: after replacement of aortic root and aortic hemiarch, followed by aortic covered stent implantation (yellow arrow). Panel C: aortic angiography at level of aortic coarctation before (panel C) and after aortic covered stent implantation (panel D). Panels E and F: the strong collateralization with prominent anterior and dorsal arterial vessels. Panel G: intraoperative picture of ascending aortic aneurysm, with large intimal tear (black arrow).

Supplementary Figure



Pressure and flow chart during cardiopulmonary bypass. Note the steady pressure gradient between radial and femoral artery during cardiopulmonary bypass. CPB: cardiopulmonary bypass. MAP: mean arterial pressure.

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