IMAGES **in PAEDIATRIC CARDIOLOGY**

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Abstract
We present two cases of severe aortic coarctation detected in adulthood and who underwent successful relief by transcatheter stent deployment using a new covered stent, Optimus® stent (AndraTec GmbH Koblenz, Germany). One patient is a 46-year old female with resistant arterial hypertension, in whom coarctation was suspected on a follow-up transthoracic echocardiogram for bicuspid aortic valve disease and subsequently confirmed on magnetic resonance imaging. The second patient is a 68-year old male whose coarctation was diagnosed incidentally at coronary angiography being performed as part of the preoperative work-up for severe aortic stenosis. Suprasternal views to assess aortic arch and flows in the proximal descending aorta should be a standard part of every transthoracic echocardiogram. Treatment of aortic coarctation should be considered irrespective of patient's age.

MeSH: Aortic coarctation, aortic aneurysm, bicuspid valve, echocardiogram, endovascular procedure

Case 1
A 46-year old woman with resistant hypertension was first diagnosed with bicuspid aortic valve (BAV) with fused right and left coronary cusps during a transthoracic echocardiogram (TTE), and subsequent transoesophageal echocardiogram (TOE), performed for work-up of suspected endocarditis. She had associated mild aortic stenosis and mild/moderate regurgitation. There was effacement of the sinotubular junction and dilatation of aortic root and ascending aorta up to 4.9cm, in keeping with bicuspid aortopathy. No evidence of endocarditis was found. A suprasternal assessment was not performed on this occasion. Clinically, she had an ejection systolic murmur best heard at the right upper sternal border, which radiated to both carotids. Her blood pressure was 170/80mmHg in both arms and there was no appreciable radiofemoral delay. Mean blood pressure on 24-hour ambulatory monitor was 142/68mmHg, despite three antihypertensive agents.
The routine follow-up TTE performed a few months after the first scan showed unchanged BAV haemodynamics and aortic dimensions. Suprasternal assessment was performed on this occasion, and, although the views were suboptimal and the peak velocity in the proximal descending aorta was only 1.7m/s, a clear diastolic tail was evident on spectral Doppler and there was non-pulsatile flow in the abdominal aorta raising (Figure 1) the possibility of concomitant aortic coarctation.

Figure 1: TTE Spectral Doppler on flow pattern in the abdominal aorta showing non-pulsatile continuous flow pattern, strongly suggestive of a diagnosis of aortic coarctation. Subsequent magnetic resonance imaging (MRI) of the aorta confirmed aortic coarctation with a discrete focal stenotic segment with a minimum diameter of 6mm at the aortic isthmus together with dilated collateral vessels arising from the aorta around the coarctation site. Phase contrast flow mapping just distal to the coarctation revealed peak velocities of more than 4m/s. The transverse arch was of normal caliber at 22-24mm (Figure 2).

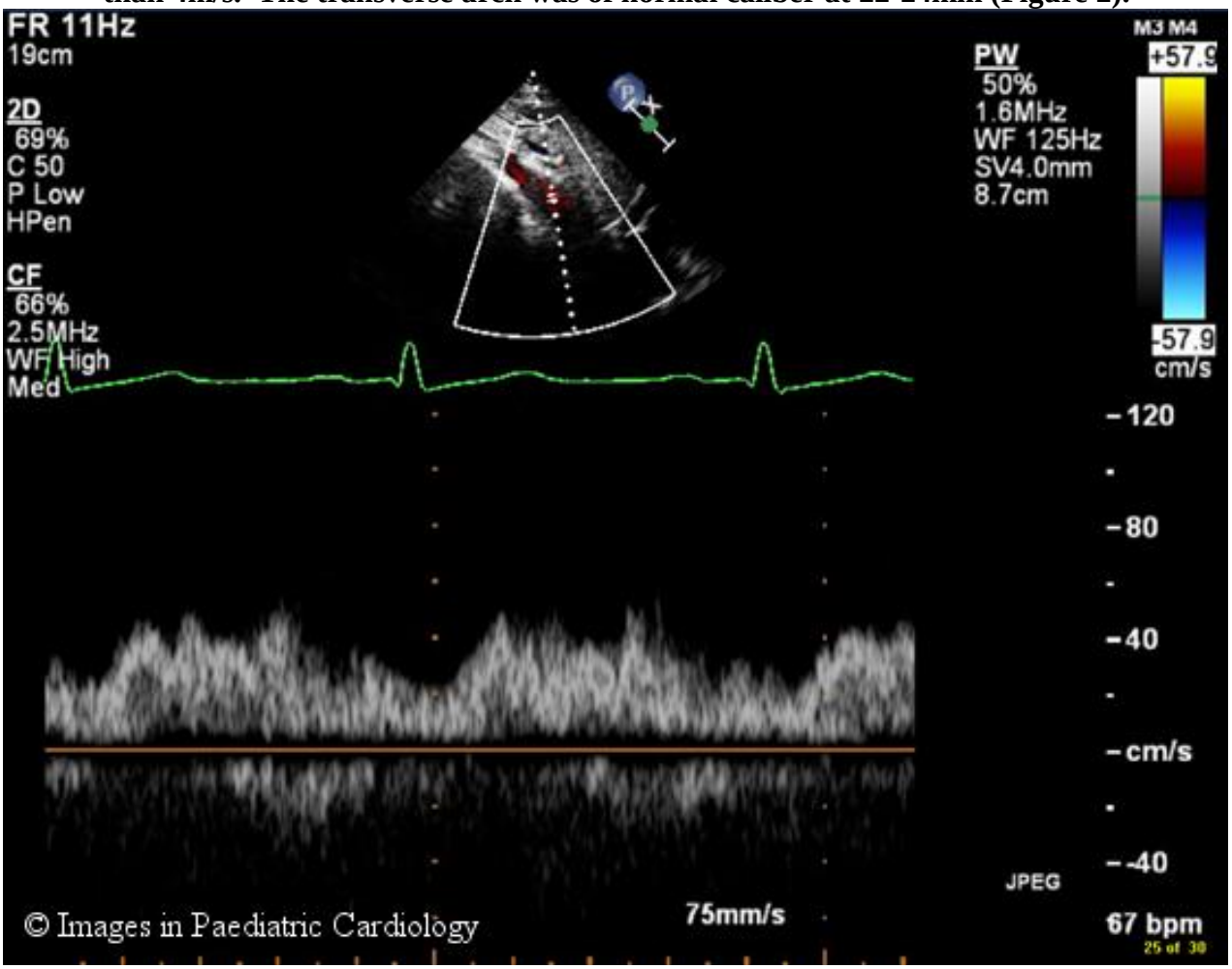
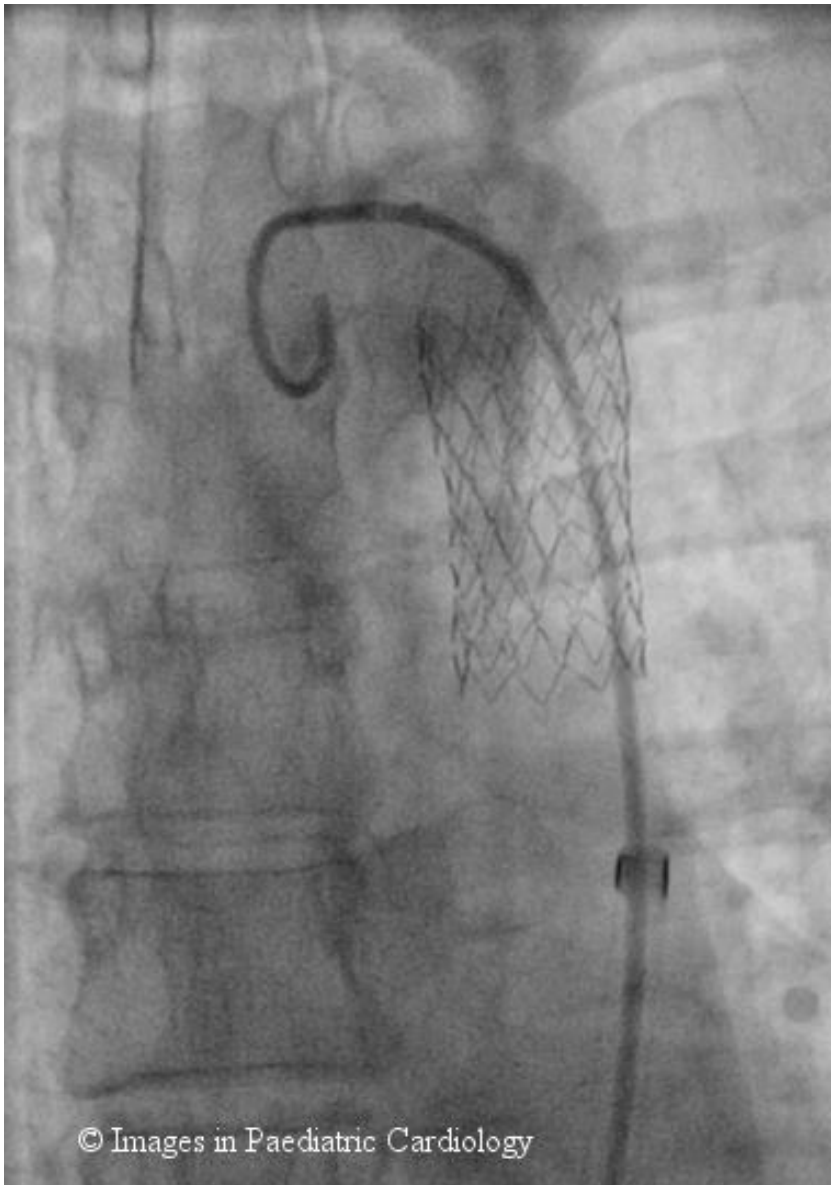


Figure 2: Cross-sectional image of Aorta on MR- The bicuspid aortic valve could be visualised. This image shows a dilated aortic root (up to 47mm) and ascending aorta (up to 51mm) with a normal calibre aortic arch (22-24mm) and tortuous proximal descending aorta (32mm). The aortic coarctation with a focal stenotic segment with a minimum diameter of 6mm at the aortic isthmus is also visualised.

Percutaneous stenting was performed through a right femoral artery approach. A gradient of 30mmHg between ascending and descending aorta was demonstrated under general anaesthetic conditions. A 38mm covered Optimus® stent (AndraTec GmbH Koblenz, Germany) was deployed at the coarctation site on an 18mm x 40mm Cristal balloon (BALT) followed by post-dilatation with complete abolition of the gradient across the site (Figure 3).

Figure 3: Fluoroscopy still image at the end of transcatheter coarctation stenting for patient 1 showing the deployed 38mm covered Optimus® stent (AndraTec GmbH Koblenz, Germany) in situ



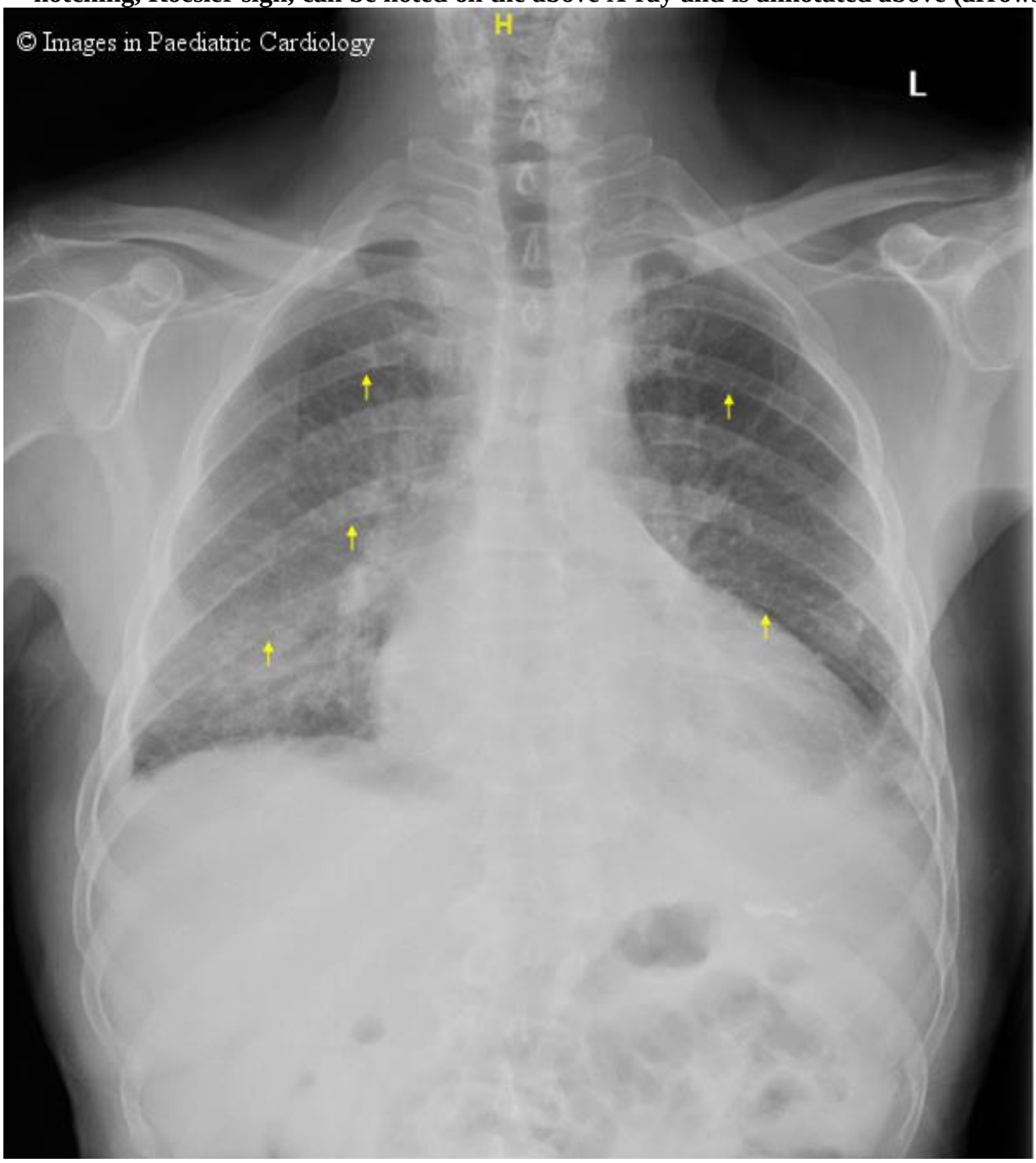
At outpatient review after the procedure, the patient's blood pressure control had improved dramatically and she it was possible to stop one of the antihypertensive agents. Repeat MRI at 10 months showed a patent stent with a peak forward velocity of 1.5m/s (Figure 4). She remains under close follow-up in view of the proximal ascending aortic dilatation which is likely to need to be addressed surgically in the future.

Figure 4: Post-stent insertion MR Angiography (MRA) - volume rendering technique with maximum intensity projection for aortic diameter measurements in MRA post-stent insertion. Dilated collateral vessels arising from the aorta around the coarctation site are seen on this image (top right). There is also significant dilatation of the proximal ascending aorta due to bicuspid aortopathy that will need monitoring and possible surgical referral in the future

Case 2

A 68-year old male with history of long-standing hypertension and recently diagnosed type II diabetes mellitus gave a one-month history of exertional dyspnoea, worsening orthopnoea and paroxysmal nocturnal dyspnoea. Clinical examinations revealed an ejection systolic murmur best heard at the right upper sternal border, which radiated to both carotids. He also had bibasal inspiratory crepitations in keeping with left ventricular failure (Figure 5). His blood pressure was 150/90mmHg in both arms.

igure 5: The chest X-ray showing venous congestion and increased peripheral markings in keeping with congestive heart failure. Small bilateral pleural effusions are noted. An area of air space shadowing is also seen in the right lower lung zone. Retrospectively, inferior rib notching, Roesler sign, can be noted on the above X-ray and is annotated above (arrows).



TTE confirmed a heavily calcified aortic valve with severe stenosis (aortic valve area of 0.9cm^2). The left ventricle was dilated with severe impairment of global systolic function (ejection fraction 30% by biplane Simpson's method) and a restrictive filling pattern on mitral inflow Doppler. Suprasternal views were technically difficult. A coronary angiogram was subsequently performed from a right femoral approach to assess for luminal coronary disease prior to management of the aortic stenosis. It was not possible to advance the catheter beyond the proximal descending aorta, after which right radial access was gained. Aortography confirmed the presence of a near

interruption of the aorta just distal to the origin of the left subclavian artery and a severely hypoplastic and tortuous isthmus. There was no significant luminal coronary disease. An ECG-gated cardiac CT angiogram confirmed the presence of severe coarctation distal to the origin of the left subclavian artery, with the aorta narrowing down to a narrow tortuous channel (minimum lumen diameter 3x3mm) (Figure 6). A dense network of collateral vessels in the mediastinum was noted (Figure 7).

Figure 6: A parasagittal reconstruction depicting the coarctation of the aorta on ECG-gated cardiac CT angiogram. Aortic dimensions are normal up till distal arch of aorta. There is severe coarctation of aorta distal to the origin of left subclavian artery at the isthmus, consisting of a narrow tortuous channel (minimum lumen diameter 3 x 3 mm) in the thoracic descending aorta. The descending aorta abruptly resumes normal calibre just below the level of the pulmonary bifurcation.

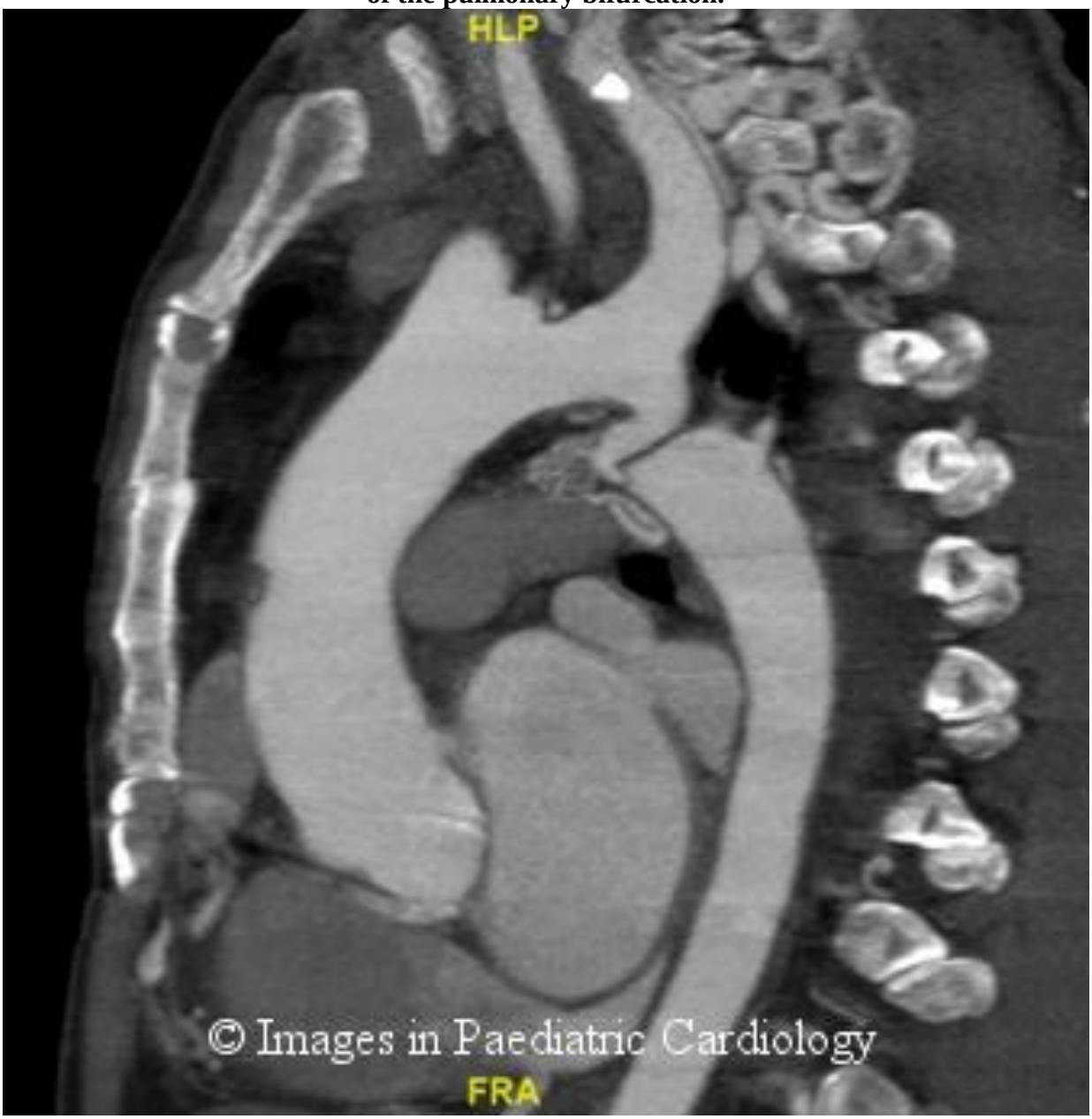


Figure 7: ECG-gated cardiac CT angiogram using volume rendering technique – A dense network of collateral vessels is observed in the mediastinum, as well as other large collaterals from the head and neck vessels. As a result of the high-pressure gradient across the area of constriction, there usually is formation of collateral pathways via the intercostal arteries. Collateral pathways (vertebral, internal thoracic, descending scapular arteries and thoracoacromial arteries) arise from the subclavian arteries and connect with the descending thoracic aorta.

A 38mm covered Optimus® stent (AndraTec GmbH Koblenz, Germany) was deployed at the coarctation site on an 18mm x40mm Cristal balloon (BALT) inflated to maximum volume after establishing a circuit from the left brachial to the left femoral artery as this facilitated crossing this complex coarctation for deployment from the femoral artery. Only a minimal gradient of 3mmHg across the coarctation site was noted at end procedure (Figure 8 and 9). TTE 2 months after the procedure confirmed a dramatic improvement in LV systolic function, with ejection fraction going up to 50%. Some months later, the patient underwent surgical valve replacement using a 21mm Carbomedics Top Hat® Supra-Annular prosthesis. He remains well and asymptomatic to date.

Figure 8: An ECG-gated cardiac CT angiogram post-stent insertion showing an optimally positioned stent at the coarctation site. A waist with a diameter of 13 mm by 13.4 mm is noted in the mid-portion of the stent

Figure 9: An ECG-gated cardiac CT angiogram post-stent insertion 3D reconstruction showing an optimally positioned stent at the coarctation site.

Discussion

Aortic coarctation is a complex disease of the vasculature accounting for 5–8% of all congenital heart defects. It typically occurs as a discrete stenotic lesion or as a long, hypoplastic aortic segment at the area of the ductus arteriosus insertion but it can be more complex with involvement of the isthmus or the transverse arch. Rarely, it may be present in the ascending, descending, or abdominal aorta. Coarctation of the aorta is frequently associated with other lesions, including bicuspid aortic valve (BAV), subvalvular, valvular, or supravalvular AS, mitral valve stenosis, or complex congenital heart defects.[4]

There is an increased propensity to certain complications as a result of abnormal haemodynamics and wall shear stresses imposed by the coarctation of the aorta. Coarctation causes significant left ventricular afterload, compensatory hypertrophy, left ventricular systolic dysfunction and formation of collateral pathways via the intercostal arteries.

A thorough cardiac examination may elicit clinical features suggestive of aortic coarctation. It is imperative to keep an open mind and routinely assess for upper body systolic hypertension, lower body hypotension, blood pressure gradient between the upper and lower extremities, radiofemoral delay, a systolic murmur or continuous bruit loudest in the interscapular region.

Transcatheter stent implantation, often using covered stents especially in older patients where calcification may be present or the lesion very tight, has become the treatment of choice for the management of aortic coarctation in older children and adults, offering shorter operative and recovery times compared to surgical repair. Several surgical techniques have been employed for the management of coarctation over time, with the most commonly utilized technique in recent years being extended resection and end-to-end anastomosis but this is nowadays used in infants only.

In conclusion, these cases highlight the importance of a comprehensive examination and echocardiogram looking beyond the pathological aortic valve. A full echocardiographic assessment to delineate the aortic root, proximal ascending aorta, aortic arch and proximal ascending aorta is mandatory in all patients.

Competing interests: None declared.

References

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