TRANSCATHETER INTERVENTION ON ADULT INTERRUPTED AORTIC ARCH WITH SEVERE BIVENTRICULAR FAILURE

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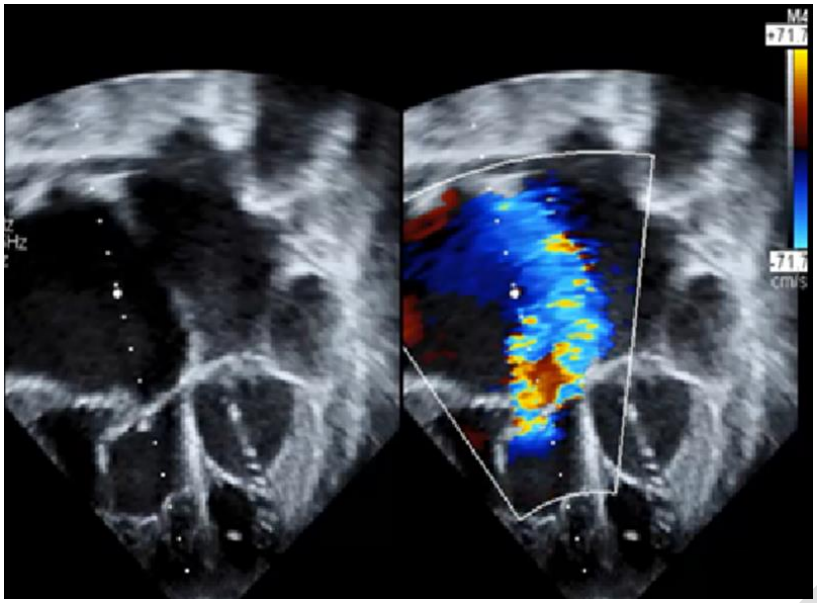
History and physical:

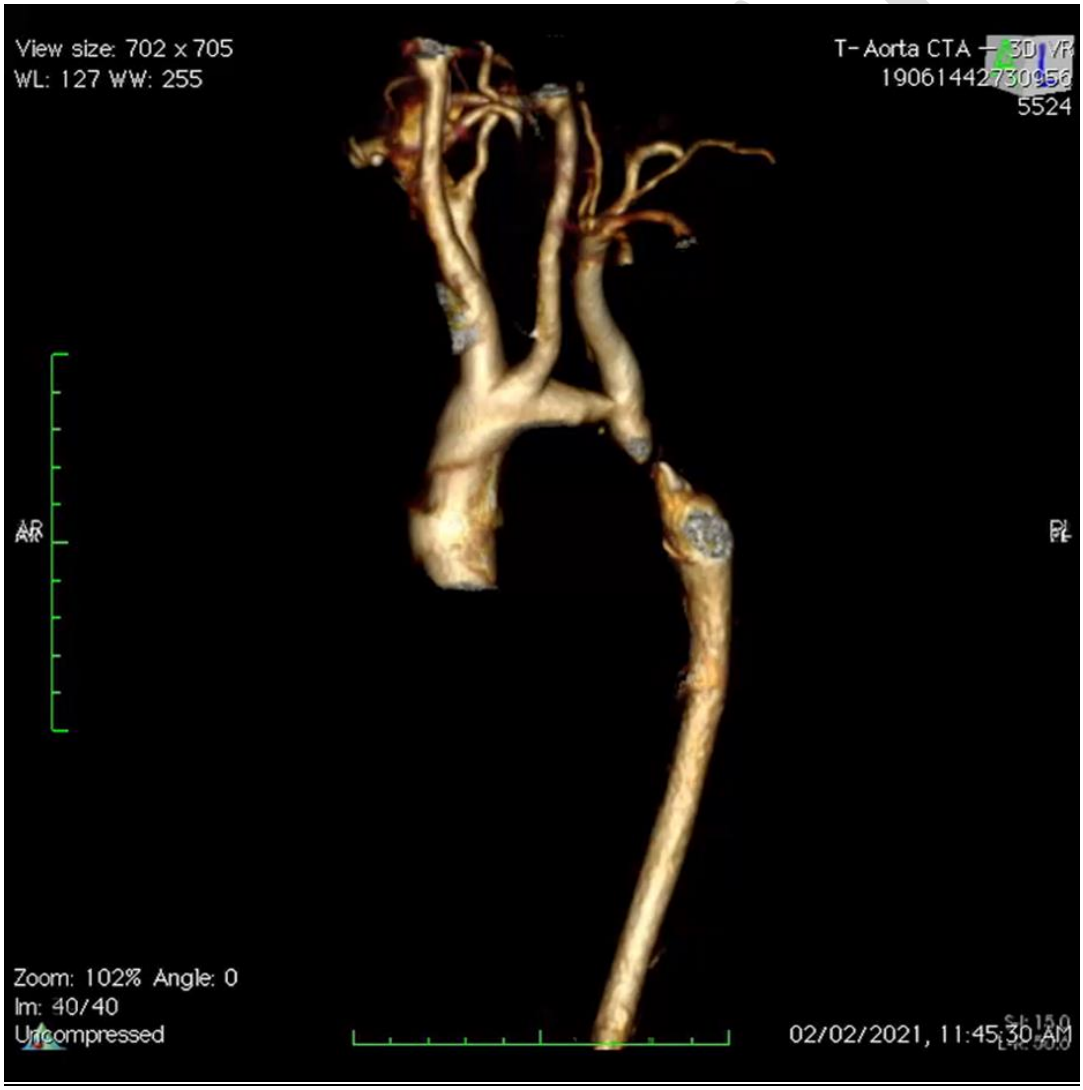
28-year-old lady presented 2 months after normal term childbirth with severe NYHA class III heart failure symptoms, including severe ascites, bilateral lower limb swelling, reduced effort tolerance, orthopnoea and PND. Peritoneal tapping was done draining 2L of transudate fluid. She was initially diagnosed as post-partum cardiomyopathy. Transthoracic echocardiogram showed mildly dilated LV with severely depressed LV function LVEF 29%, severe biatrial enlargement, severely dilated RV with severely depressed RV function TAPSE 1.1cm, severe TR with severe pulmonary hypertension PG 140mmHg, concerns for possible coarctation of aorta, no PDA, small pericardial effusion.

Her vital signs: weight 35.5kg, height 135cm, right upper extremity BP 139/100mmHg, lower extremity BP 118/66mmHg, HR 95bpm, pre- / post-ductal SpO2 100%. On exam, she was small in stature, tachypnoeic, had an asymmetrically larger left chest with parasternal heave and had a grade III/VI pansystolic murmur over her left lower sternal border. She also had significant JVD, ascites, hepatomegaly and lower extremity oedema.

We followed up with a CT aortogram, which showed severe cardiomegaly and complete type-A interruption of her aortic arch, with an interrupted length of 6mm, mildly hypoplastic distal aortic arch and dense arterial collateralizations supplying her descending aorta.

Imaging:





Indication for intervention:

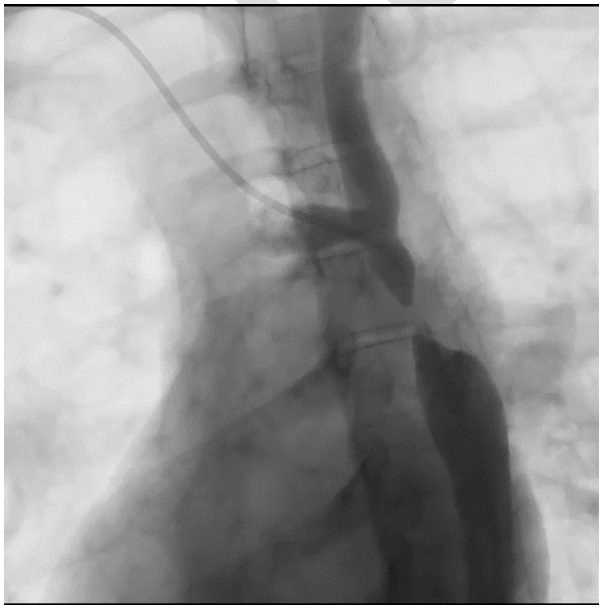
Urgent intervention was definitely warranted, however best modality of intervention was unclear, as both surgery and transcatheter caried significant risk. After a multi-disciplinary meeting discussion, due to risk of going on bypass, consensus was to attempt percutaneous treatment first, with surgical team on standby in case of complications.

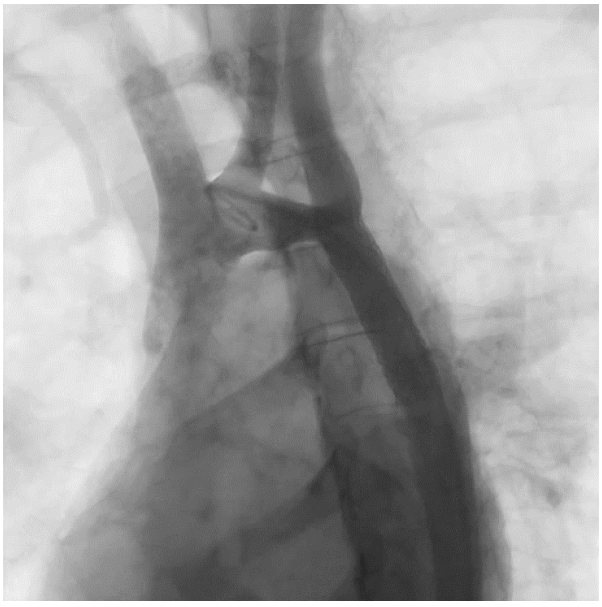
Intervention:

Right heart study performed confirmed pulmonary hypertension (PAp 98/55/74mmHg, PCWp 22mmHg, PVRi 10.3wu.m2). Simultaneous angiograms on the upper and lower segments of the interrupted arch via femoral artery and radial artery cannulation confirmed short segment type-A arch interruption. Proximal arch measured 18.2mm, distal arch 9.0mm, interruption 6.1mm, descending aorta at diaphragm 10.8mm. There was a 54mmHg transaortic gradient.

With a 10mm PFM Multi-Snare placed in the upper pouch as a target, the interrupt segment was recanalized with a Baylis RF wire advanced across with short pulses. The wire was snared and pulled out of the radial artery to create a secure loop, and the interrupted segment was crossed with a 2.7F Progreat microcatheter and 5F Terumo GlideCath co-axially. An Amplatzer stiff wire was parked in the right subclavian artery, and a 9F Mullins sheath was advanced across the interrupted segment. A 12x49mm Bentley BeGraft covered stent was deployed. Post-stenting, we noted partial jailing of the LSCA. Over two V18 wire, one in the ascending aorta and one across the jailed LSCA, two 8x30mm Valver balloon were inflated simultaneously to dilate and flare the proximal stent as well as relief the jailing.

Post-intervention, there was good flow across the aortic stent and LSCA. There was a 10mmHg residual gradient primarily over the hypoplastic arch.





Learning points of the procedure:

Interrupted aortic arch can be treated percutaneously in the cath lab with good long-term results.