



COARCTATION OF THE AORTA WITH AORTIC STENOSIS IN A PREMATURE PATIENT WITH TURNER'S SYNDROME

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History and physical:

33-week, 1.6 kg female with Turner's syndrome, long-segment coarctation of the aorta, and aortic stenosis. Baseline echocardiogram showed a left-sided aortic arch with aberrant right subclavian artery, severe, long-segment hypoplasia of the transverse arch with severe juxtaductal coarctation, small bidirectional PDA, normal LV size with mildly reduced function, normal MV with mild regurgitation. The aortic valve was unicuspid and measured 4.5 mm (Z-score -1.5) with a peak velocity of 2.2 m/sec in the presence of severe coarctation. The patient was started on PGE. The decision was made to proceed with cardiac catheterization for stent placement in coarctation via left carotid artery cutdown with assessment of the AV after relief of coarctation.

Indication for intervention:

Severe coarctation of the aorta, aortic stenosis

Intervention:

She was brought to the catheterization lab at one week of age and 1.7 kg. LCA cutdown was performed and a 4-Fr Prelude sheath was placed. Baseline angiography showed long-segment coarctation of the aorta that measured 2.8 mm in diameter and 12.5 mm in length. The transverse arch measured 4.0 mm and the proximal descending aorta measured 4.5 mm. The coarctation was stented using a 4 mm x 15 mm Vision stent which was deployed at nominal pressure. The aberrant right subclavian artery was intentionally jailed, as it originated at the site of the coarctation. Angiography and echocardiogram post-stent showed appropriately positioned stent with no residual obstruction. Echocardiogram showed a peak gradient of 44 mm Hg with mean gradient of 24 mm Hg across the AV. The decision was made to defer intervention on the AV at this time and monitor closely.

At 2 months of age and weighing 3 kg, the patient was brought back to the catheterization lab due to increased aortic valve gradient (peak 75-86 mm Hg, mean 38-50 mm Hg) and Vmax across the coarctation stent of 3.1 m/sec (peak 40.8 mm Hg, mean 16.3 mm Hg) on echocardiogram. Baseline hemodynamics showed a 7 mmHg gradient across the stent. Balloon dilation of the stent was performed using a 5 mm and 6 mm x 2 cm Sterling balloons with no residual gradient. Balloon angioplasty of the right subclavian artery was performed through the side cell of the stent using a 3 mm x 12 mm NCEmerge balloon with improvement of ostium caliber. Post-stent dilation AV gradient was 57 mm Hg. AV annulus measured 7 mm on echocardiogram and angiography. Balloon aortic valvuloplasty was performed using a 6 mm x 2 cm Tyshak mini balloon. Post-valvuloplasty, there was a 10 mm Hg residual gradient across the AV with no significant aortic insufficiency. She was discharged from the hospital 2 weeks later with stable echocardiogram.



Currently, she is 7 months old with most recent echocardiogram showing mild to moderate aortic stenosis (peak gradient 34 mm Hg, mean 20 mm Hg) with Vmax across the aortic stent of 3.2 m/sec. She is scheduled for elective repeat cardiac catheterization in the next month.

Learning Points of the Procedure:

Assessment of obstruction in series, stenting of coarctation in premature infants.

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