




Specialist Vascular Surgeons

**Supravalvular aortic arch stenosis:
A report of progressive disease and endovascular hybrid repair in a child.**

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Supravalvular aortic arch stenosis: A report of progressive disease and endovascular hybrid repair in a child.

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Introduction

- SVAS – supraaortic stenosis
 - A disease of the arterial wall caused by reduced elastin (ELN)
- ELN arteriopathy
 - Usually results in thickening of the tunica intima and media, mostly affecting the ascending aorta.
 - Also reported in pulmonary and coronary arteries.
- SVAS may be non-syndromic or syndromic
 - Syndromic:
 - Complex genomic disorder that presents with neurobehavioural, craniofacial, cardiovascular and metabolic abnormalities.
 - Non-syndromic:
 - Caused by mutation of the ELN gene (auto-somal dominant inheritance)
 - ELN arteriopathy may result in discrete hourglass or diffuse long-segment stenosis of the supraaortic aorta.
 - Significant stenosis and high pressure gradients may result in hypertension, congestive cardiac failure, myocardial infarction and death.



Case Report

- 7 Year old male patient with complex, inherited, non-syndromic SVAS with numerous interventional procedures since birth.
 - Open surgical artery repair at 7 months for arch stenosis:
 - Bovine pericardial patch angioplasty of the ascending and descending aorta as well as the aortic arch via median sternotomy.
 - At age 7 he presented with refractory hypertension due to the SVAS with subsequent left ventricular hypertrophy and lower limb calf claudication.

Age-related anatomical abnormality, clinical presentation and intervention from birth to 7 years of age

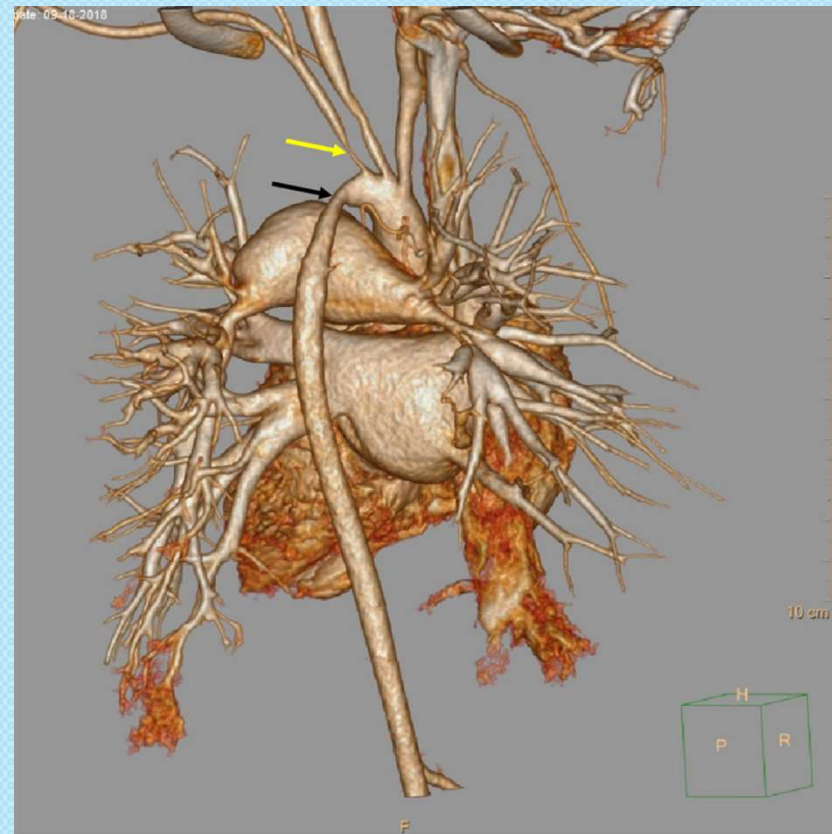
Age	Anatomical abnormality	Clinical presentation	Surgical intervention
2-days	Patent foramen ovale (PPO) Peripheral pulmonary stenosis (PPS)	Ejection systolic murmur (ESM)	Nil
2-months	PPS	ESM	Nil
6-7-months	SVAS stenotic ascending aorta, arch, descending aorta, and PPS. Biventricular hypertrophy	Hypertension ESM	Pericardial patch / Bovine patch of ascending aorta / arch / proximal descending aorta
15-months	PPS	Hypertension	Balloon angioplasty PPS
17-months	Stenosis of LCCA and LSCA. Stenotic arch, proximal descending arch, LCCA, LSCA, PPS	Hypertension	Balloon angioplasty PPS Complication: pulmonary haemorrhage
27-months & 3-years	Angiogram: progressive narrowing of arch and proximal descending aorta	Progressive hypertension Anti-hypertensive escalated	Nil
7-years	Stenosis of the arch and proximal descending aorta Stenosis of the ostia of the LSCA and LCCA	Hypertension Left ventricular hypertrophy Lower limb claudication	Hybrid surgery

Pre-operative computed tomography (CT) angiogram

Findings:

- High-grade stenoses of the:
 - mid-arch
 - Proximal descending aorta
 - Left common carotid artery (LCCA)
 - Left subclavian artery (LSCA)
 - Pulmonary artery segmental branches
- The ascending aorta, brachiocephalic artery and right common carotid artery appeared free of stenoses.

Posterior oblique view of the aorta and great vessels showing high-grade stenosis of the proximal descending aortic (black arrow) and proximal LSCA high-grade stenosis (blue arrow)



Hybrid procedure – single stage

- Due to re-stenosis of the arch and ostial stenoses of the LCCA and LSCA, a hybrid procedure was done.
 - Extra-anatomical bypass:
 - Standard right to left CCA bypass and a RCCA to LSCA bypass using 6mm polytetrafluoroethylene (PTFE).
 - No carotid shunting used as the external carotid and vertebral arteries were adequate in maintaining cerebral perfusion.
 - Ligation of the stenosed proximal LCCA and LSCA.
 - Endovascular procedure:
 - Standard vascular access of the right femoral artery.
 - Angiography confirmed adequate cerebral perfusion.
 - Standard endovascular stent grafting from the brachiocephalic artery to the proximal descending aorta using an 11mmx100mm nitinol Gore Viabahn covered stent.
 - Residual stenosis of 50% (sustained pressure gradient of 40mmHg) following balloon dilatation.
 - Deployed a second 14x45mm stainless steel Z-Med stent graft.
 - The arch diameters were 10mm proximally and 4mm at the narrowest arch segment and 11mm at the mid-descending aorta.
 - The 2nd stent graft was placed as an inlay within the 1st stent graft.
 - A 10mm high pressure balloon was used to expand both stent grafts adequately.



Outcome:

Immediately post-operative

- No post-op complications.
- Improved calf claudication.
- His blood pressure appeared to improve to normal values for his age.
 - He was discharged with no anti-hypertensive medications.



Outcome: Follow-up

- 2 Months:
 - Normotensive → 108/47mmHg, normal range for height percentile = 113-116/72-77mmHg.
 - Anti-hypertensives introduced at age 6 were stopped after the procedure.
 - Claudication symptoms had resolved.
- 1 Year:
 - No clinical recurrence of hypertension or calf claudication
 - No further anti-hypertensives required.
 - Scripted Aspirin 75mg/daily.
 - No neurological symptoms reported.

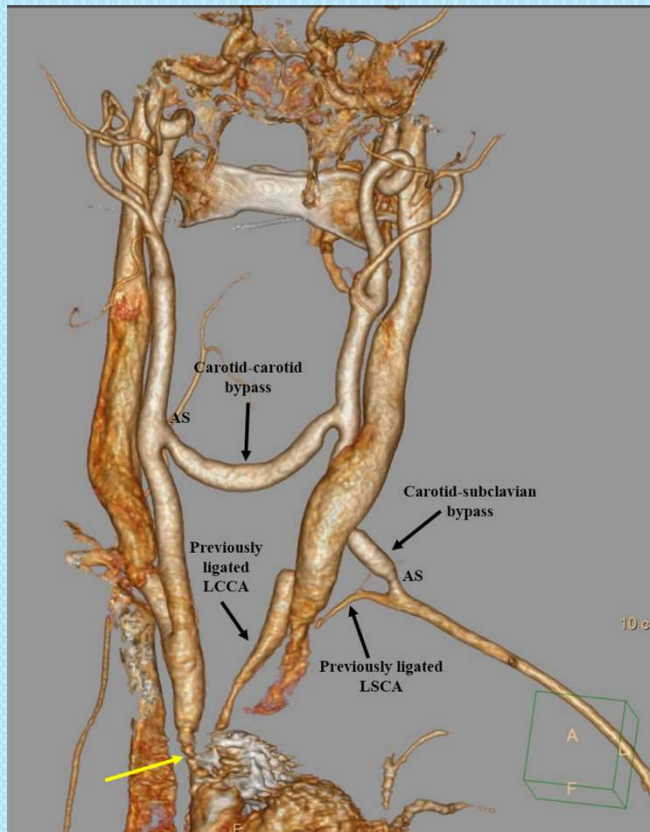


CT angiogram follow-up

- 6 – Months:
 - Patent extra-anatomical bypasses and adequately expanded stent grafts.
- 1 Year:
 - High-grade stenosis at the ostium of the brachiocephalic artery.
 - Anastomotic stenoses of the previous bypass grafts.
 - The aortic stent grafts (SG-SG) remained relatively patent.

CT angiogram follow-up

Anterior posterior view showing the results of the hybrid procedure (black arrow) and progressive stenosis of the brachiocephalic ostium (yellow arrow).
AS = anastomotic stenosis.



Sagittal view after the hybrid procedure revealing patent stent grafts in the aortic arch.





Discussion

- Highlights the clinical consequences of ELN arteriopathy affecting the supra-avalvular aorta, the branches of the arch and pulmonary arterial branches
- Progressive arteriopathy:
 - Patch aortoplasty at 7 months (open surgery).
 - Restenosis of the arch at 7 years (hybrid surgery).
 - Multiple new ostial and anastomotic stenoses at 8 years (1 year follow up CT).



Discussion

- Open surgery is regarded as the gold standard for supraaortic coarctation, but reports indicate high rates of re-stenosis and re-operation.
 - Imamura et al (2010) Surgery for SVAS
 - 49 patients (age range 24 days to 24 years)
 - 25 years follow up
 - 86% underwent patch repair
 - 33% of re-operation due to stenosis.



Discussion

- **Shahanavaz et al (2020)** Balloon expandable stents on residual stenosis and hypertension
 - 57 patients (age range 4days – 42 years)
 - 7 Centres
 - 37 patients followed up
 - 8 patients required re-intervention.
- **Brown et al (2002)** Standard patch aortoplasty
 - 101 patients
 - 38 year follow up
 - 14% requited re-operation at least once
- **Deo et al (2012)** Single-patch repair
 - 53 year follow up
 - 32 re-intervention procedures in 16 patients
 - Risk factors: young age at initial surgery and aortic stenosis.



Discussion

- Currently no comparative studies between endovascular intervention and open surgery in the treatment of non-syndromic SVAS patients.
- This case highlights the progressive stenotic complications associated with ELN arteriopathy.



Discussion

- Endovascular intervention may be better suited in high risk patients that have had multiple open surgical procedures.
- Adult endovascular interventionalists need to be aware of ELN arteriopathy and its progressive nature.
- Endovascular strategies may need to be modified for younger patients.
- In this case the stent-in-stent procedure allow for subsequent balloon angioplasty should it be needed.
- Proper endovascular planning and anticipation of progressive multi-vessel stenosis is important.



Conclusion

- Endovascular intervention using stent grafts may be considered an alternative to open surgery in aortic arch re-stenosis due to ELN arteriopathy.
- An extra-anatomical bypass may be necessary to facilitate stent graft placement.
- Although this procedure was feasible in this 7 year old and appeared to alleviate the symptoms, lifelong monitoring and possible subsequent intervention is mandatory.