



Burnt out Takayasu arteritis and its challenges

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INTRODUCTION

Takayasu arteritis is an idiopathic rare large vessel primary vasculitis, more frequently occurring in females of child bearing age. It causes granulomatous inflammation of the aorta and its large branches. This can cause:

- **Stenosis/Occlusive symptoms:** Decreased/absent pulses, hypertension, claudication, risk of pulmonary thrombosis, cerebral infarction, blindness, and limb ischaemia.
- **Aneurysmal Symptoms:** Aortic regurgitation, vessel rupture, chronic pain
- **Constitutional symptoms:** Fever, Weight loss, "Burn out"

After the active inflammatory phase, the disease becomes 'burnt-out' causing occlusion in the aorta or large vessels. Most patients have prenatal counselling to discourage pregnancy. Life expectancy: median age at death is 38 years ⁽¹⁾. There are few case reports regarding management of pregnancy in this cohort

CASE REPORT

A 31 year old female (P1+1) presented to our preassessment clinic for delivery planning. Her obstetric history included a termination of pregnancy at 9 weeks and an ELCS at 32+5 weeks gestation due to severe maternal hypertension (when her disease was mild). She was diagnosed with Takayasu arteritis at the age of 20, stopped immunosuppression five years ago and was in the burnt out stage of her disease.

She was a high risk patient due to having symptomatic three vessel coronary disease, unstable angina, severe cardiovascular dysfunction with LV hypertrophy, claudication at 500m and a fixed 5mm width of her descending aorta with extensive collateralisation, such that her lower limbs derive their entire blood supply from her inferior epigastric vessels.

Despite taking seven antihypertensive agents, the patient had a baseline systolic blood pressure of 180-220. She declined termination of pregnancy with full understanding of the likelihood of maternal death due to cardiac disease or stroke. During her pre-operative planning, she was consulted with multiple specialities:

Vascular MDT: prophylactic axillo-bi-femoral not of benefit. No IR option. May need axillo-bi-fem intra-op.

Coronary MDT: if ACS, manage standard pathway. Not suitable for ECMO due to poor vasculature in the event of a cardiac event. Only ECMO option would be via sternotomy which makes decannulation impossible.

Delivery planning MDT: for consultant led elective LSCS. Midline laparotomy incision to be performed to reduce risk of devascularising legs. An emergency GA section for fetal distress not in best interest therefore NO antenatal CTGs to be performed. Patient had elective LSCS expedited to 28 weeks gestation as she was experiencing uterine steal symptoms, with chest pain and static fetal growth

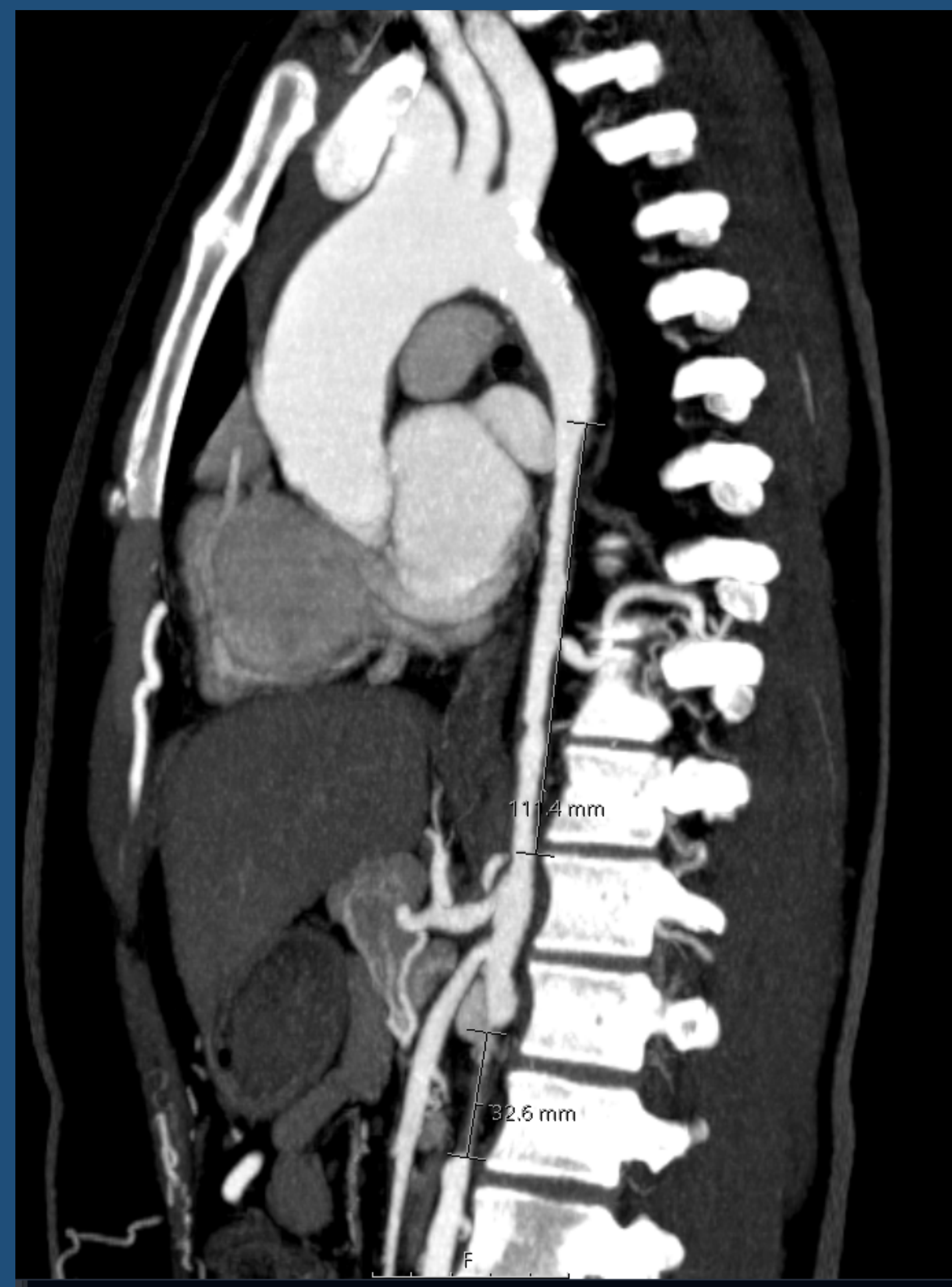


Fig 1. CT Aorta, sagittal view
Distal to the origin, there is a 5mm calibre stenosis of the descending aorta. There is a modest restoration in calibre of aorta between celiac trunk and renal arteries measuring 1.1cm

ANAESTHETIC DILEMMAS:

- **WHERE to do?** Decision not for a co-located cardiac tertiary centre as would not be ECMO candidate
- **BP CONTROL – must keep sbp > 180**
- **Risk of CVA**
- **What is rescue plan if ischaemic legs?**
- **MOH**

The patient was brought to ITU 24 hours prior to surgery for invasive monitoring and to assess her response to IV hypotensive agents.

The patient underwent a cardio stable general anaesthetic (whilst informing neonatal team of high opioid dosage). GA allowed a perioperative transoesophageal echocardiography to be performed to guide filling and drugs choice. Operation occurred in a Vascular theatre with surgical team on standby.

Intra-operatively, her sbp was kept > 180 as she has no gut perfusion below that. The baby was taken to NICU and doing well. She was extubated and had normal lower limb Dopplers post-operatively.

DISCUSSION

This case underscores the extraordinary collaborative working and early discussions about rescue therapies in order to safely manage the consequences of Takayasu arteritis.

There were many examples of shared decision making in this case: from initially counselling the patient about terminating this pregnancy, through to discussing rescue therapies in the event she has ACS or ischaemic limbs. This occurred over months of planning with different medical and surgical teams and gauging second opinions from other tertiary centres. It is an example of how early well-documented planning could be useful for patients in this cohort in the future.

REFERENCES

(1) Comarmond C, Saadoun D, Nizard J, Cacoub P. *Pregnancy issues in Takayasu arteritis*. Semin Arthritis Rheum. 2020 Oct;50(5):911-914. doi: 10.1016/j.semarthrit.2020.08.001. Epub 2020 Aug 9. PMID: 32911287.

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