



# CPP

## Loeys-Dietz and Pregnancy

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## Case KL

Tertiary Referral:	1 <sup>st</sup> trimester, aortic root of 37mm
Presentation:	34year old, G <sup>2</sup> P <sup>0</sup> , no surgery, no treatment. Father abdominal aortic surgery in 50s. Normal habitus.
Genetic Screening :	Confirmed TGFB2 mutation.
Diagnosis:	Loeyz-Dietz.



# Loeyz-Dietz Syndrome

- LDS:** A syndrome of altered cardiovascular, craniofacial, neurocognitive and skeletal development.
- Significance:** Aggressive arterial aneurysms-mean age at death 26 years.
- Pregnancy:** High incidence of pregnancy complications.



## Back to case

First visit: Confirmed DCDA Twin pregnancy 9/40.

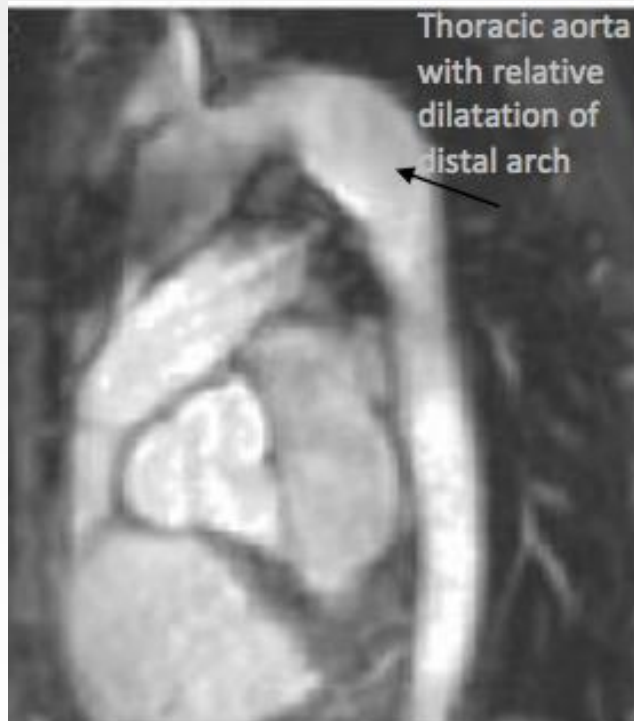
Confirmed mutation of TFGBR2 gene.

cMR: Aortic root at 42mm

Dilated proximal descending aorta

Tortuous iliac arteries

Follow up: Referred to pregnancy and heart disease team



Thoracic aorta  
with relative  
dilatation of  
distal arch



# How do you counsel?

How do you counsel? Rare

To discuss: Risk dissection.

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Data from Marfan : 1% (<4.0cm) 10% (>4.0cm).

In LDS: Aorta > 4cm offer surgery.



# Other risks associated with LDS-Case series

Risks:

Obstetric Haemorrhage

Uterine rupture

Aneurysms elsewhere e.g. cerebral



# Case Management

Counselled:	Risk dissection approx 15%.
Fetal Medicine:	Twin 2- Large Cleft and IUGR.
Cardiac assessment:	Echo/cMR 4-6 weeks.
Plan:	Close surveillance of BP.





## Decisions: MDT Discussion at 30 weeks regarding plans for delivery

Gestation?

Mode?

Who?

Where?

Analgesia?

Post-op stay?

The Baby!



## Delivery & Post Op course

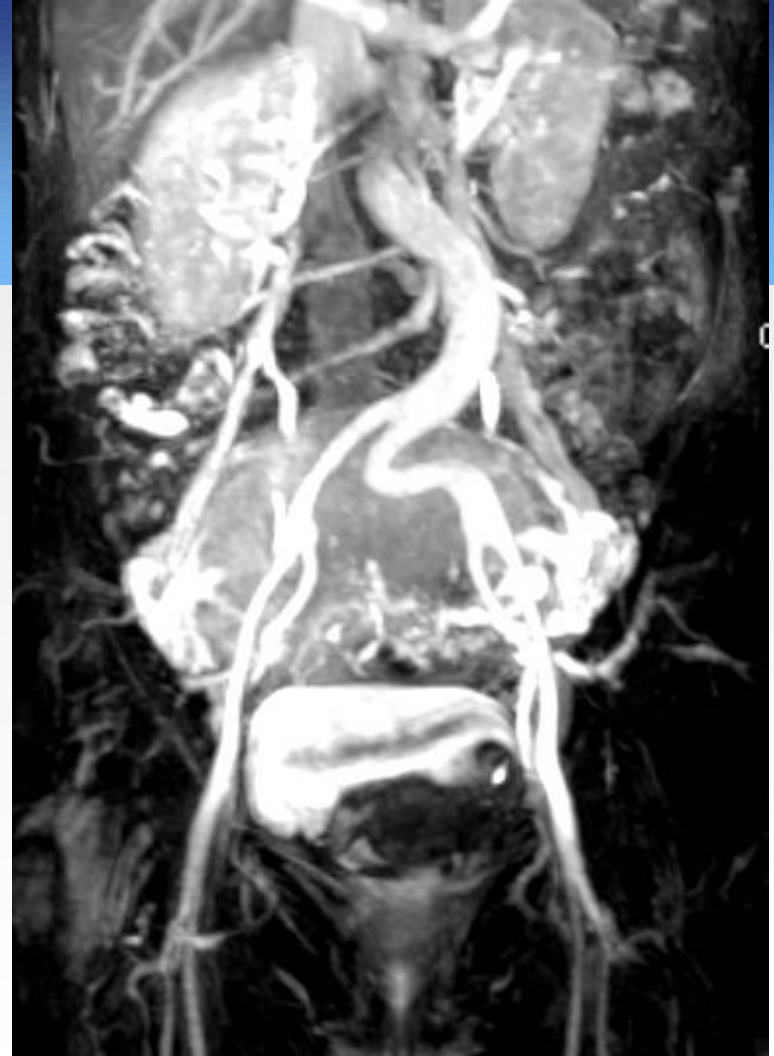
CS: Main theatre- Haemorrhage 1.8L  
BP stable post delivery.

Imaging: CMR and MRA.

Plan for cardiac surgery approx 6months.

Contraception discussed.

Offered start Losartan (ARB)





# Summary

LDS is rare but aggressive aortopathy

Data suggest risk of dissection is significant (>1%).

Preconception Counselling **MANDATORY** for this group.

Managed tertiary cardiac/obstetric service

