Antihypertensive Management Recommendations for Scoliosis Surgery in Paediatric Patients with Loeys-Dietz Syndrome.

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Description

We describe our experience, complications, and recommendations for the perioperative management of a 2-stage kypho-scoliosis repair, in a patient with Loeys-Dietz syndrome (LDS). Significant haemodynamic instability was observed and managed during stage 1 of the repair.

Case

A 16 year old female, 41kg, with LDS, presented for elective repair for significant thoracolumbar kyphoscoliosis (Cobb angle of 84°). Co-morbidities included severe pectus excavatum, micrognathia, moderative restrictive pulmonary function tests (FEV_1 and FVC in low 60s, preserved ratio), and aortic root dilatation of 3.8cm (z-score +6.6). Resting ECG revealed frequent unifocal PVCs which suppressed on exercise testing. Transthoracic echo revealed left ventricular shortening fraction of 28% with preserved valvular integrity, and she had been previously started on atenolol. Additional preoperative assessment included full multidisciplinary risk assessment including a comprehensive review of all available anaesthesia literature.
The procedure consisted of combined general surgical and orthopaedic access via a left thoracolumbar incision with anterior vertebral release and trans diaphragmatic access of the lower thoracic vertebra.

Following uneventful IV induction, intubation, and placement of arterial, central, and peripheral vascular access, maintenance anaesthesia was delivered via target controlled infusions of propofol (3-5mcg/ml), remifentanil (2-3 ug/ml), ketamine (4mcg/Kg/hr).
She demonstrated significant hemodynamic lability approximately 10 minutes post induction characterised by both atrial (PACs, junctional rhythm) and PVCs (burden approximately 10/min). Following prone positioning, noradrenaline and adrenaline infusions were required for low mean arterial pressure. External vascular compression during thoracic dissection required bolus fluids, vasopressors, and ongoing communication between the anaesthesia and surgical teams to facilitate haemodynamic recovery. Over the 5 ½ hour surgical time, there was a total blood loss of 25ml/kg, and she received nearly 100ml/kg of crystalloid and 220mls of cell saver blood.

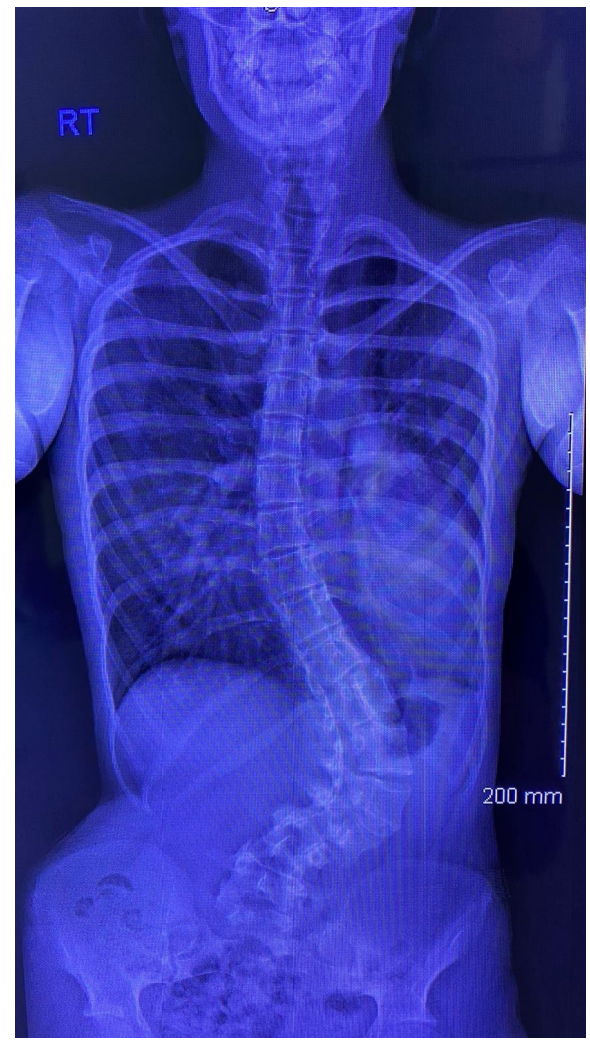

Figure 1
X-ray of case patient demonstrating thoracolumbar kyphoscoliosis

Acknowledgements
The patient's parents kindly consented to the use of this data.

Manifestations of Loeys-Dietz Syndrome[5]

Cardiovascular	Craniofacial	Skeletal	Skin	Hands	Ocular	Other
- Aneurysms - Arterial tortuosity - Congenital heart defects	- Hypertelorism - Bifid uvula - Malar hypoplasia - Craniosynostosis - Cleft palate - Micrognathia	- Scoliosis - Joint laxity - C-Spine instability - Pectus excavatum - Osteoarthritis	- Easy bruising - Abnormal scaring - Translucency	- Arachnodactyly - Contractures	- Myopia - Retinal Detachment - Eye Muscle disorders	- Food allergies - Herniae

Discussion

LDS is a rare autosomal dominant syndrome first described by Loeys et al in 2005[1]. Characteristics are summarised in the above table, of note there is generalised arterial tortuosity and ascending aortic aneurysms with a disproportionately high propensity for dissection[2].
Haemodynamic instability for patients with LDS are described due to underlying comorbidities and abnormal anatomical relationship between the heart, great vessels, and thoracic vertebrae.
Previous case reports of scoliosis surgery in patients with LDS describe an inability to tolerate rotational correction[3] and perioperative cardiac arrest[4], both in patients who were on antihypertensives preoperatively.

For our patient's subsequent two surgeries, a revision of the first stage and completion second stage procedure, atenolol was held, and she demonstrated no significant perioperative hypotensive episodes.

The above case may further suggest that patients with LDS demonstrate disproportionate sensitivity to antihypertensives perioperatively, and that strong consideration should be given to their discontinuation for major physiologically disruptive surgery.

References

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