

Elective caesarean delivery in a patient with osteopathia striata with cranial sclerosis (OS-CS/Horan-Beighton syndrome)

Murray L, Roberts F, Nicholas J, Douglass J, Burch A

Department of Anaesthesia, Jessop Wing, Sheffield Teaching Hospitals NHS Foundation Trust



We believe this to be the first ever published case report of neuraxial anaesthesia in a patient with OS-CS

Introduction

Osteopathia striata with cranial sclerosis (OS-CS) is a rare X-linked dominant disorder with around 100 cases globally. It is associated with bony dysplasia, craniofacial abnormalities (macrocephaly, cleft palate), hearing loss and scoliosis. [1,2] A patient (X) underwent caesarean delivery (CD) at our centre with a combined spinal-epidural anaesthetic; she had previously been advised against conceiving due to the potential risks involved in delivery.



Figure 1* (Above) Airway assessment. Note limited mouth opening, macroglossia, dental crowding and previous tracheostomy scar: difficult airway anticipated

Anaesthetic pre-assessment

- A 29-year-old nulliparous woman (X) with OS-CS was reviewed in our obstetric anaesthetic pre-assessment clinic
- She had multi-system involvement: previous tracheostomy and cleft palate repair, patent ductus arteriosus ligation, restrictive ventricular septal defect, mildly dysplastic aortic valve with mild regurgitation and mildly dilated aortic root, reflex syncope, thoracolumbar scoliosis, hearing loss and controlled asthma
- Airway assessment: prognathism, small mouth, macroglossia and dental crowding
- Specialist ENT review and nasendoscopy revealed unilateral vocal cord palsy and prolapse of both arytenoids
- After counselling that regional anaesthesia may be difficult or impossible, and predicted difficulty in airway management would pose unacceptable maternal risk in an emergency, she underwent category four CD at 38/40
- **Primary anaesthetic plan was regional anaesthesia, with awake fiberoptic intubation (AFOI) or awake tracheostomy plus general anaesthesia if this failed.**

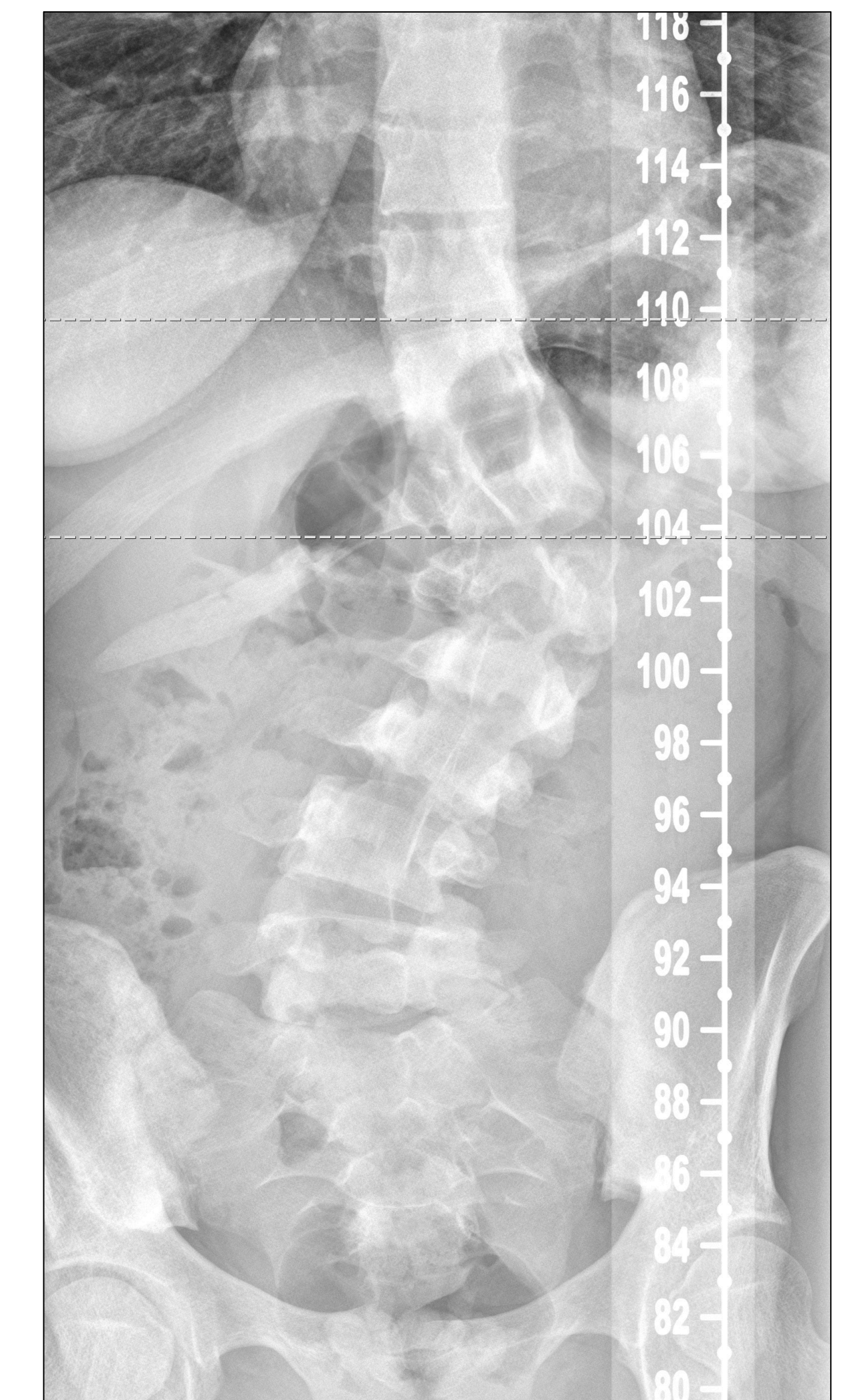
Intra-operative management

- We sited two wide-bore cannulae, an arterial line and used nasal high flow oxygen throughout
- We prepared a fiberoptic scope and nasal tube in case AFOI was required, and obtained the difficult airway trolley
- We also prepared infusions of remifentanyl and clonidine in case anxiolysis was required
- A separate-space combined spinal epidural was sited using ultrasound to map anatomical targets. We administered a total of 2 mL 0.5% bupivacaine with 1:200000 adrenaline via the epidural, and 2.6 mL 0.5% heavy bupivacaine with 300 µg diamorphine intrathecally. Bilateral sensory block to T3/T4 and dense motor block were achieved
- As per our usual practice, we ran an infusion of phenylephrine 100 µg/ml at 10-20ml/h, giving boluses as required to manage hypotension
- Post-delivery, 100 µg dilute carbetocin was given via infusion in view of a history of cardiac disease
- We gave 1g of tranexamic acid 20 minutes post delivery at the request of the obstetrician. Blood loss totalled 800 mL and the epidural catheter was removed in recovery
- For venous thromboembolism prophylaxis, dalteparin was given 6 hours post epidural catheter removal, along with use of compression stockings

Post-operative management

The patient was monitored in our obstetric high dependency area overnight prior to ward transfer. We followed her up on day one post-partum and both X and her partner expressed gratitude for the care received.

Figure 2* (below): Patient's thoracolumbar radiograph demonstrating significant scoliosis: difficult regional anaesthesia anticipated



Discussion

Patients with a combination of difficult airway and abnormal spinal anatomy pose a particular challenge to the obstetric anaesthetist. Multi-disciplinary planning is vital in these cases, with pre-operative counselling, preparation of theatre teams and equipment, and availability of specialist ENT support. We have demonstrated that successful neuraxial anaesthesia is achievable in patients with OS-CS, using pre-procedure ultrasound and separate space combined spinal epidural technique.

References

- 1.Orphan Anesthesia: Anaesthesia recommendations for Osteopathia Striata (with cranial sclerosis) (March 2025) <https://www.orphananesthesia.eu/docman-de/rare-diseases/eng/published-recommendations/osteopathia-striata/1794-osteopathia-striata-final/file.html> (accessed 10/12/2025)
- 2.Gear R, Savarirayan R. Osteopathia Striata with Cranial Sclerosis. *GeneReviews [Internet]* 2021 (Revised 2023) (accessed 10/12/2025)

***Written patient consent for publication/presentation and permission for use of clinical images and patient photographs has been obtained. Publication/presentation of this abstract has been approved by the Trust Caldicott guardian.**