

Learning Objective: Recognize anesthetic considerations in obstetric patients with cleidocranial dysplasia and prior spinal fusion and evaluate the feasibility and risks of neuraxial anesthesia in skeletal dysplasia with spinal instrumentation.

Introduction to Cleidocranial Dysplasia in Obstetrics

Cleidocranial dysplasia (CCD) is a rare autosomal dominant skeletal disorder affecting intramembranous ossification. It affects about 1 in 1,000,000.

Key skeletal features include:

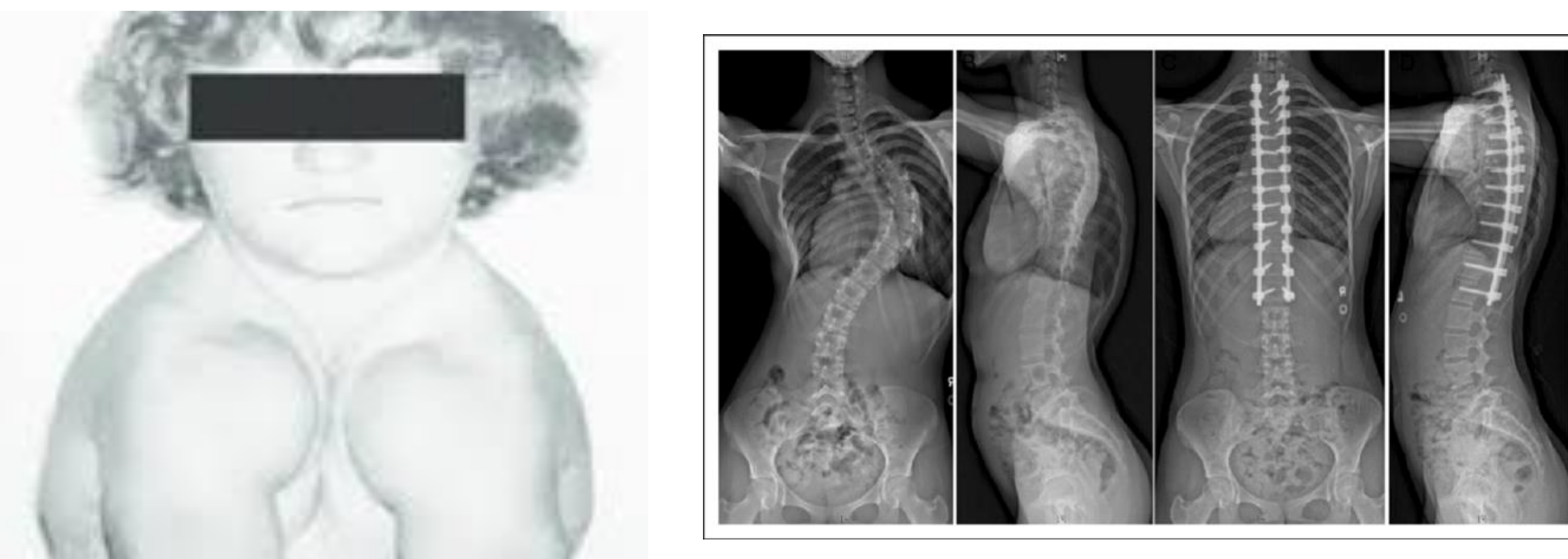
- Facial and oral abnormalities – midfacial hypoplasia, micrognathia, high arched cleft palate
- Spinal and pelvic deformities – scoliosis and anatomic variations
- Short stature
- Clavicular hypoplasia
- Delayed suture closure

Patients with CCD frequently undergo spinal fusion for scoliosis, which may make neuraxial anesthesia even more challenging

Anesthetic management may be complicated by:

- Pelvic abnormalities – increasing the risk of a cesarean delivery
- Difficult airway management

Neuraxial anesthesia is generally preferred due to reduced airway manipulation and improved postoperative outcomes, but anatomical variability in CCD introduces technical and dosing challenges

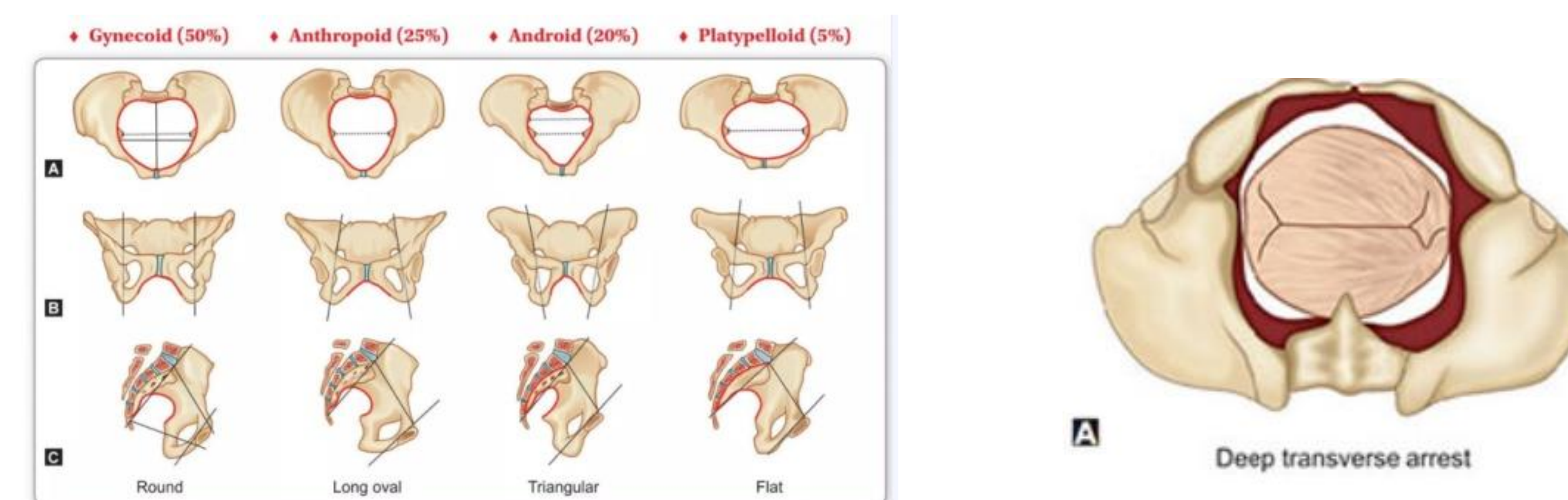


Case Description

A 24-year-old G1P0 at 33 weeks 4 days presented in preterm labor with 9 cm dilation and bulging membranes. She had a known diagnosis of cleidocranial dysplasia with short stature (4'10") and scoliosis. At age 17, she underwent T2–L2 spinal fusion for scoliosis correction. Prenatal pelvic exam was consistent with an android pelvis, raising concern for obstructed labor. Although a trial of labor is not contraindicated in CCD, the patient elected cesarean delivery due to suspected pelvic inadequacy and her prior spinal fusion.

Airway assessment:

Mallampati III
Thyromental distance > 3 FB
Full neck ROM
Normal dentition



She presented in advanced labor with 9 cm dilation, prompting urgent cesarean section. Laboratory values were normal.

A single-shot spinal was performed at L3–L4. Intrathecal medications included:

1.4 mL of 0.75% bupivacaine
15 mcg fentanyl
100 mcg morphine

Case Discussion

- CCD creates anesthetic challenges due to skeletal abnormalities involving the spine, pelvis, and craniofacial structures.
- Prior T2–L2 spinal fusion can make neuraxial access difficult
- Despite these limitations, neuraxial anesthesia is often preferred given potential airway difficulty and the risks associated with general anesthesia in pregnancy.
- Literature notes that neuraxial spread may be unpredictable in CCD due to short stature and spinal deformity.
- In this case, L3–L4 access remained feasible, and spinal anesthesia was successful on the first attempt.
- A lower intrathecal dose was utilized given her short stature.

Successful emergent cesarean delivery highlights the importance of:

- Early and thorough pre-delivery airway and neuraxial evaluation
- Anticipation of anatomic variability
- Multidisciplinary collaboration
- Patient-centered delivery planning and shared decision-making

Conclusion

This case demonstrates a case in which neuraxial anesthesia was performed for cesarean delivery in patients with cleidocranial dysplasia and prior spinal fusion. Given the rarity and anatomical variability of CCD, individualized pre-delivery planning and multidisciplinary coordination are essential to maintain maternal safety and autonomy during obstetric emergencies.

Sources

DORE, D. D., MACEWEN, G. D. & BOULOS, M. I. (1987). Cleidocranial Dysostosis and Syringomyelia Review of the Literature and Case Report. *Clinical Orthopaedics and Related Research*, 214, 229-234.

Baloglu MB, Kargin D, Albayrak A, Atici Y. The Treatment of Cleidocranial Dysostosis (Scheutauher-Marie-Sainton Syndrome), a Rare Form of Skeletal Dysplasia, Accompanied by Spinal Deformities: A Review of the Literature and Two Case Reports. *Case Rep Orthop*. 2018 Jul 9;2018:4635761. doi: 10.1155/2018/4635761. PMID: 30123598; PMCID: PMC6079611.

K. Rozdars, J. Riordan, Anaesthetic management of an obstetric patient with cleidocranial dysplasia. *International Journal of Obstetric Anesthesia*. Volume 33, 2018, Pages 94-96, ISSN 0959-289X, <https://doi.org/10.1016/j.ijoa.2017.07.003>. (<https://www.sciencedirect.com/science/article/pii/S0959289X1730242X>)