



Prepared Before the Push: Peripartum Management in a Parturient with Gorlin and Sotos Syndrome and a Known Difficult Airway

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Background

Traditionally, pregnancy has been deemed a cause of "difficult airway" due to increased airway edema, reduced functional residual capacity, and full-stomach precautions. Furthermore, the physical and hormonal changes during pregnancy lead to higher risk of aspiration. Gorlin and Sotos Syndromes each pose challenges to airway and neuraxial management, adding to the changes with pregnancy.

Gorlin Syndrome:

- Facial dysmorphism, odontogenic keratocysts, macrocephaly, meningioma, spina bifida, fused vertebrae

Sotos Syndrome:

- Intellectual disability, hypotonia, pre-eclampsia, macroglossia, macrocephaly/frontal bossing, mandibular hypoplasia, high-arched palate, pointy chin, scoliosis

Clinical Findings

- 40-year-old G6P3 at 36+5 weeks gestation
- Induction of labor for history of pre-eclampsia with severe features
- Prominent facial features from Gorlin and Sotos Syndromes
- History of meningioma resection and ventricular drain for hydrocephalus prior to pregnancy
- Unanticipated difficult airway during meningioma resection two years prior (failed direct laryngoscopy, required bougie and indirect laryngoscopy for intubation)
- Two prior vaginal deliveries at outside hospitals, epidural records unknown
- Stable intracranial pressures throughout pregnancy

Key Considerations:

- Concern for contraindications to neuraxial with increased intracranial pressure, possible mass effect, vertebral anomalies
- Complex PMH and advanced maternal age increase risk of cesarian section
- Known difficult airway and concern for fiberoptic intubation in STAT cesarean section



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Case Presentation

Clinical History

Meningioma resection two years prior, stable intracranial pressure

Two prior NSVD's at outside hospitals, epidural records unknown

Documented difficult airway

Presentation

Induction at 36+5 weeks GA for pre-eclampsia with severe features

No symptoms of elevated intracranial pressure or mass effect

Mallampati III, large tongue, small mouth opening, mandibular hypoplasia

Work-Up and Consults

Neurosurgery: MRI lumbar spine to rule out tethered cord/spina bifida, cleared for neuraxial anesthesia with vaginal delivery

ENT: consulted for flexible laryngoscopy confirming hypertrophied tongue

Trauma surgery: on standby for possible emergent surgical airway

Delivery Plan

Multidisciplinary discussion advocating for early labor epidural

Indirect laryngoscopy and flexible bronchoscopy available for awake intubation

Delivery and Post-Partum Care

Successful placement of dural puncture epidural

Induction of labor with uncomplicated vaginal delivery

Postpartum follow-up with neurosurgery and ENT



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Discussion

- This case highlights the need for meticulous planning in parturients with known difficult airways and complex comorbidities
- Early multidisciplinary planning is essential with thorough work-up if time permits. Image based risk assessment allowed for safe neuraxial anesthesia
- Without an early functioning neuraxial, an emergent cesarean delivery may have required an awake fiberoptic intubation, possibly delaying delivery of the fetus and increasing maternal and fetal risk

Take Away Points

A functioning labor epidural reduced risk of intubation in case of an emergent cesarean section in a patient with a known difficult airway and complex medical history