

# Cesarean Delivery for a Patient with Von Hippel-Lindau Syndrome

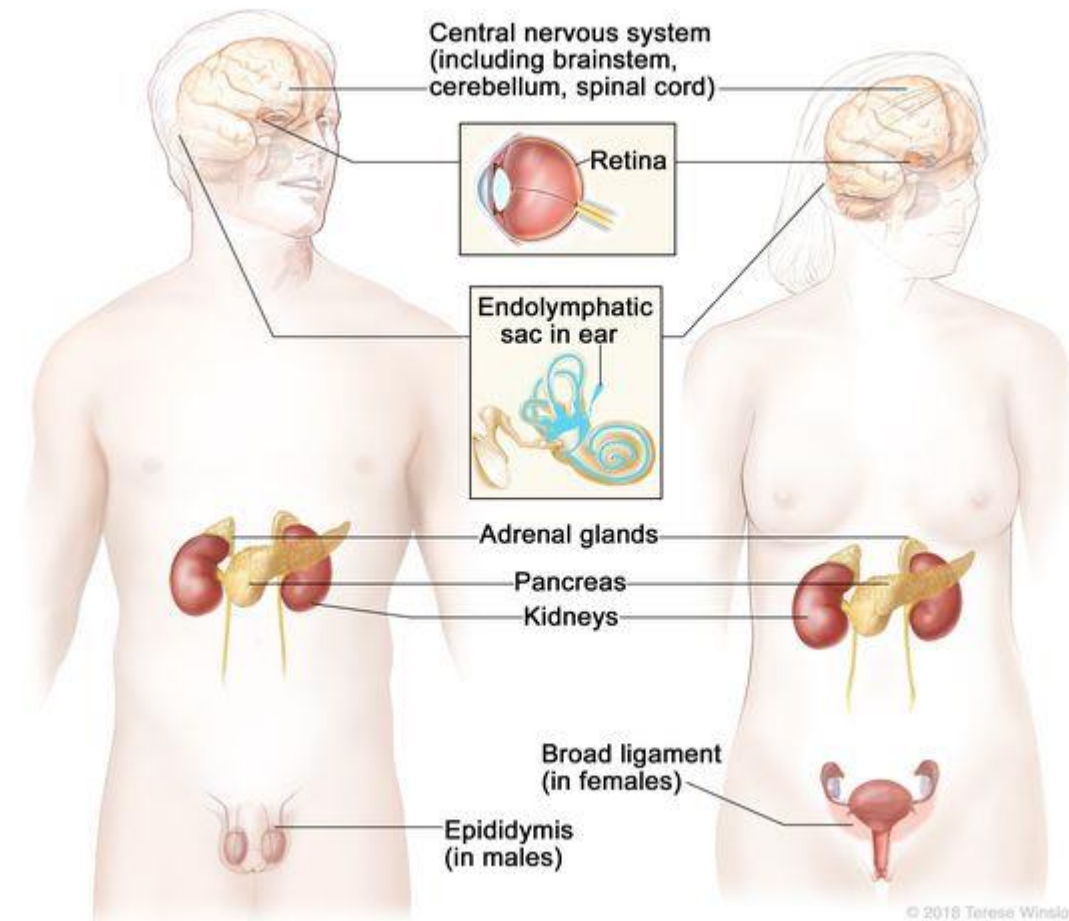
Margaret Lund MD, Blake Caracci DO, James Damron MD, Neva Lemoine MD



## Background:

- Von Hippel-Lindau syndrome is an **autosomal dominant** disease caused by mutation of the **VHL tumor suppressor gene**, thereby leading to growth of numerous tumors and cysts.
- Characteristic lesions include retinal hemangioblastomas, renal cell carcinomas, pheochromocytomas, **CNS hemangioblastomas**, pancreatic tumors, and endolymphatic sac tumors<sup>1</sup>.
- Management of a pregnant patient with Von Hippel-Lindau syndrome requires careful monitoring (due to **risk of hemangioblastoma development and progression** with increased estrogen and total blood volume) and delivery planning<sup>2</sup>.

Areas of the Body Affected by von Hippel-Lindau Disease



# Cesarean Delivery for a Patient with Von Hippel-Lindau Syndrome

Margaret Lund MD, Blake Caracci DO, James Damron MD, Neva Lemoine MD



24-year-old G2P0010 with known type 1 von-Hippel Lindau. Lesions included posterior fossa and brainstem hemangioblastomas s/p prior resection, bilateral ocular vascular lesions (right eye blindness), pancreatic cysts, and spinal cysts.

Worsening headaches and new dysphagia prompted repeat scan → significant increase in size of known lesion.

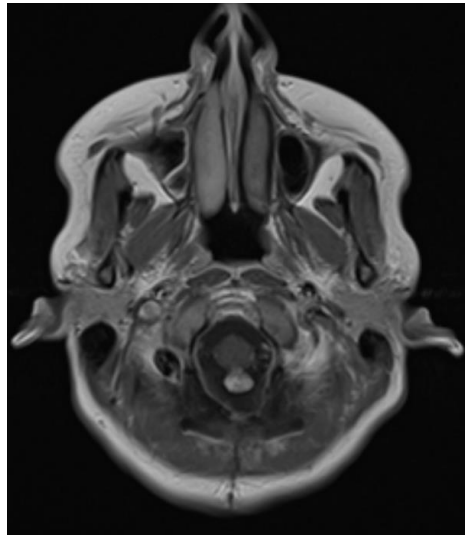
Referred to neurosurgery, who recommended delivery at 37 weeks to permit prompt surgical intervention.

## Anesthesia Plan:

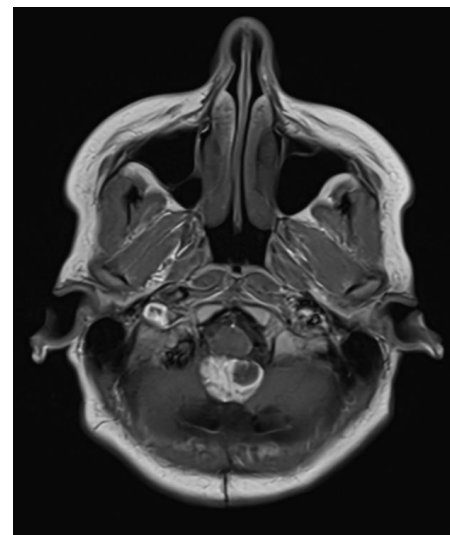
- General anesthesia in favor of neuraxial anesthesia
- Preinduction arterial line
- Videoscope intubation
- Multimodal pain control including pre-emergence TAP blocks

Underwent lesion resection postpartum.

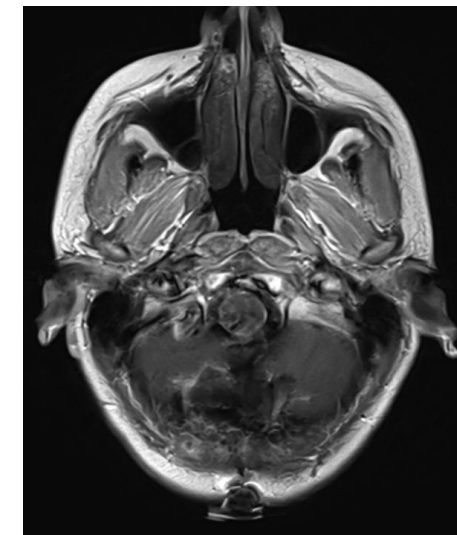
## Case Report:



MRI from ~1 year prior to delivery



MRI near time of delivery



MRI post-resection

# Cesarean Delivery for a Patient with Von Hippel-Lindau Syndrome

Margaret Lund MD, Blake Caracci DO, James Damron MD, Neva Lemoine MD



## Conclusions:

### Anesthetic management

Broad range of clinical presentations

Monitor existing CNS tumors

Evaluate for elevated ICP

Frequent MRI imaging may be warranted due to growth during pregnancy<sup>2</sup>

TABLE 1. Contraindications to spinal anesthesia.

#### Absolute Contraindications

- Patient refusal
- Infection at the site of injection
- Uncorrected hypovolemia
- Allergy
- Increased intracranial pressure

#### Relative Contraindications

- Coagulopathy
- Sepsis
- Fixed cardiac output states
- Indeterminate neurological disease

Spinal Anesthesia - NYSORA



References