

NEUROFIBROMATOSIS TYPE 1 IN PREGNANCY: IMPLICATIONS FOR DELIVERY PLANNING

MARGARET LUND MD, STEPHEN FORD DO, NEVA LEMOINE MD, JAMES DAMRON MD

Clinical Manifestations:



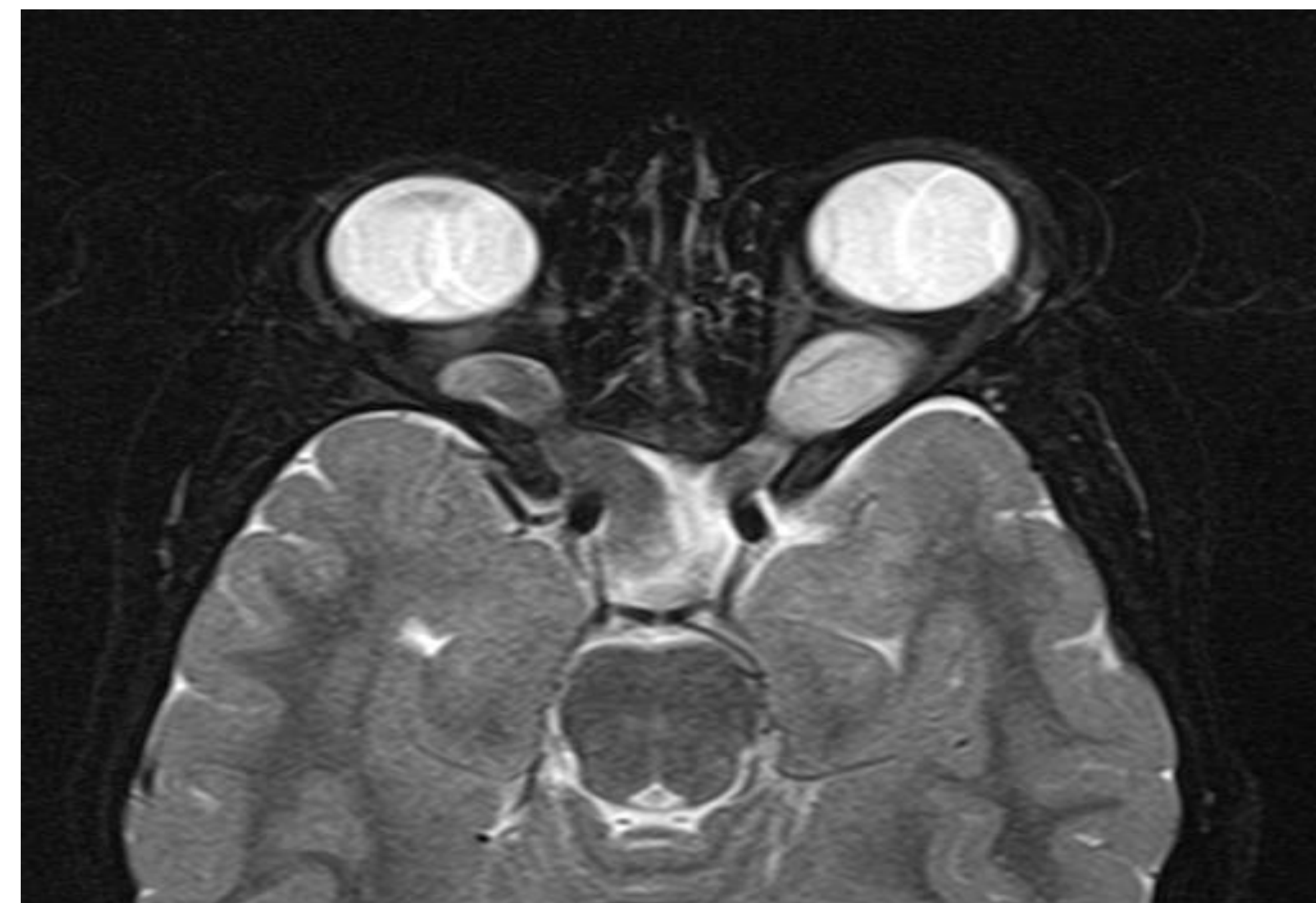
Neurofibromas (WebMD)



Lisch nodules (Physiopedia)



Café au lait spots (MD Searchlight)



Optic pathway glioma (Radiopaedia)

Background

- Neurofibromatosis Type 1 (NF-1), or von Recklinghausen disease, is an **autosomal dominant** syndrome characterized by a **loss-of-function mutation** of the gene encoding neurofibromin, a **tumor-suppressing protein**

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Clinical Course and Treatment

37y/o G5P0 with NF-1 and IUGR with elevated UAD presented with **persistent Category 2 tracing** at 33w4d

- Innumerable neurofibromas
- **Oral cavity neurofibroma**
- MRI without epidural or intrathecal lesions
- No signs/symptoms of elevated ICP
- Appropriate for neuraxial

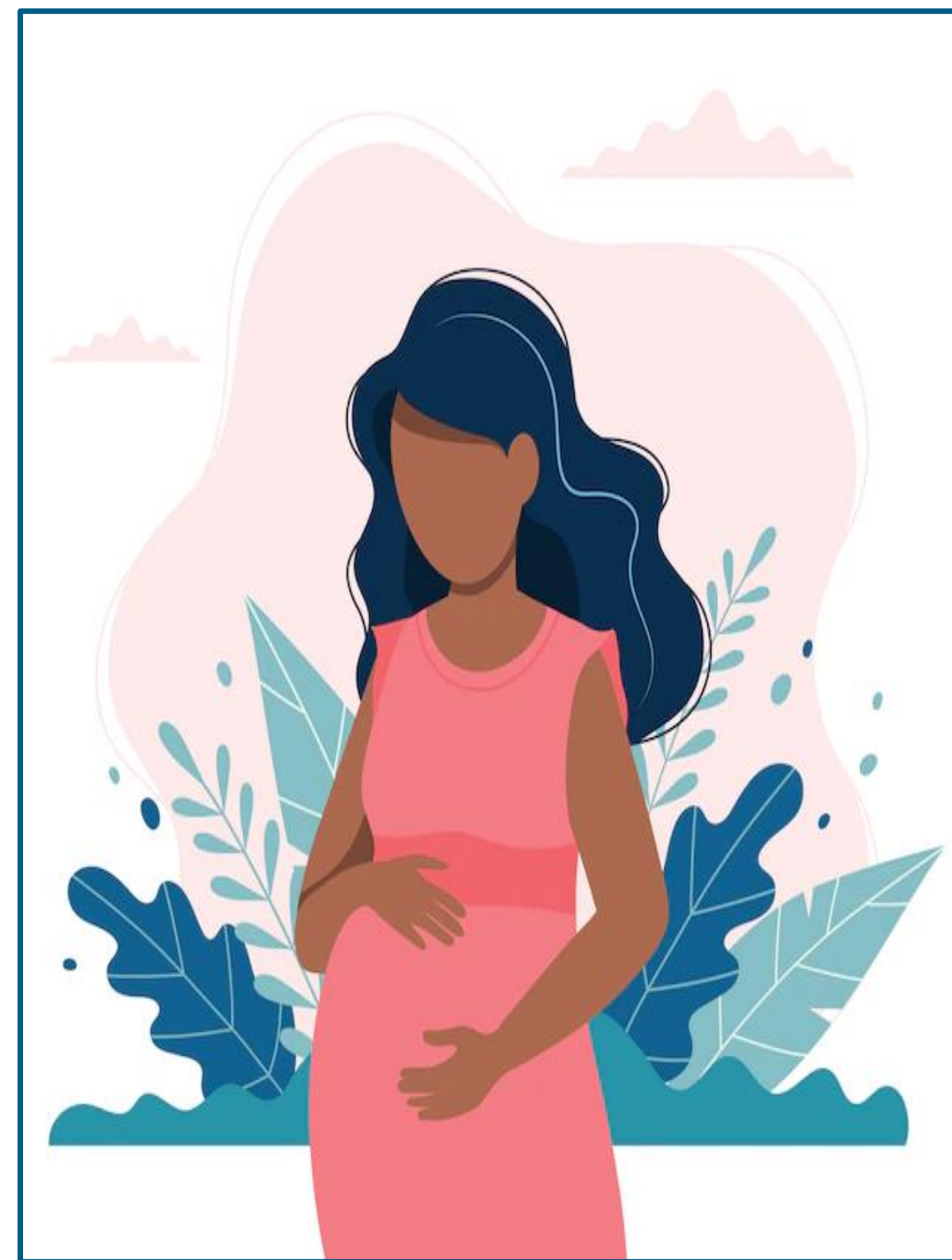
Crash Cesarean section under general anesthesia.

Video laryngoscopy to navigate around oral cavity lesion.

Neonatal Apgars 8/9, both mother and baby did well postoperatively.

Developed severe abdominal pain and prolonged fetal decel concerning for **placental abruption** at 34w0d

Uneventful Cesarean Section
Confirmed Abruption



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Teaching Points

Special delivery planning of parturients with NF-1



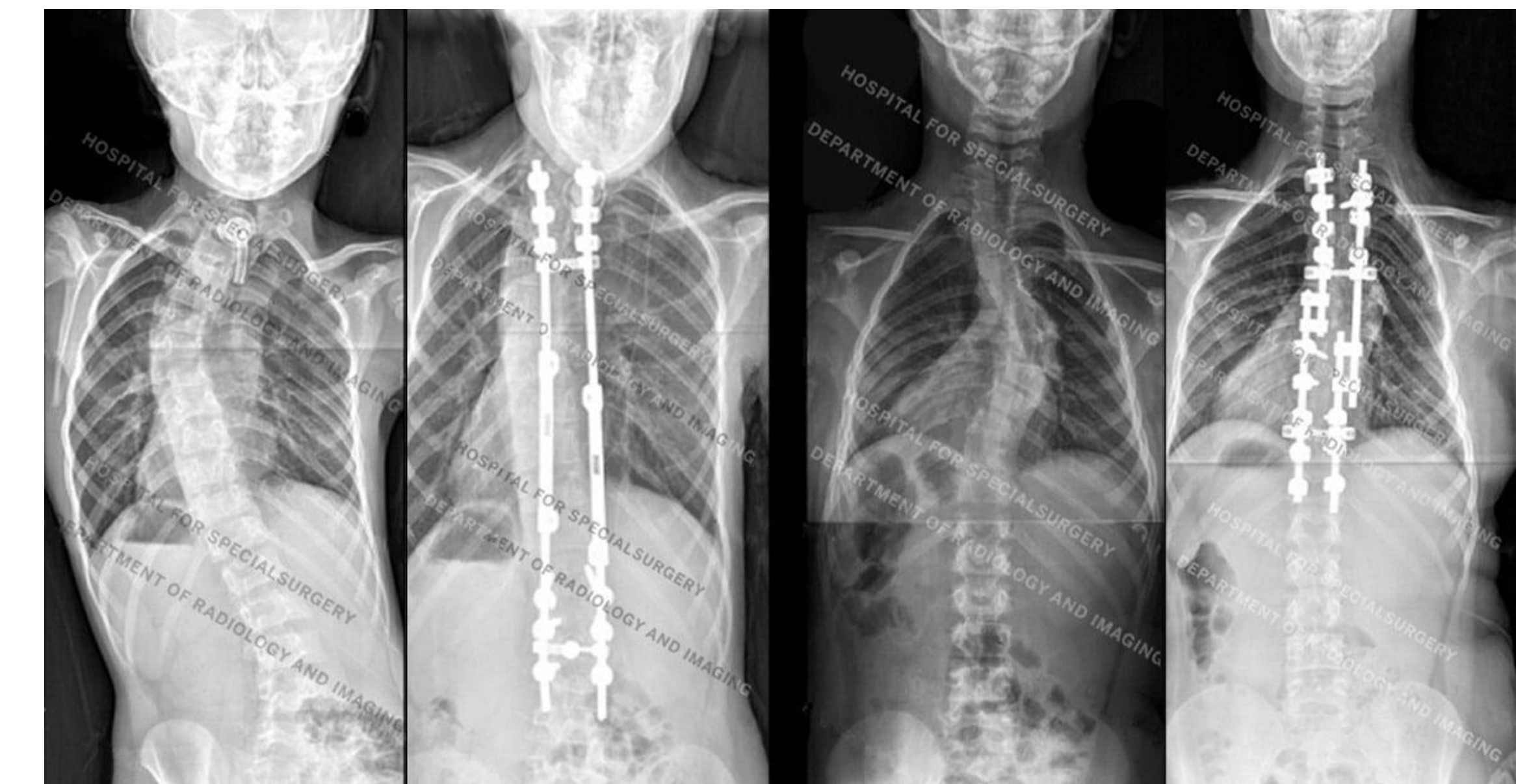
Dural ectasia (Radiopaedia)

Are there Neuraxial Neurofibromas?

- Given the propensity for new neurofibroma formation and existing neurofibroma growth in pregnancy, imaging should be timed as close to delivery as possible.

Other complicating aspects of NF-1 must be considered:

- Dystrophic scoliosis – could make neuraxial placement technically challenging
- Dural ectasia – could limit neuraxial anesthetic efficacy
- Pharyngeal/laryngeal neurofibromas – could further complicate airway management



Dystrophic scoliosis (HSS)

References

