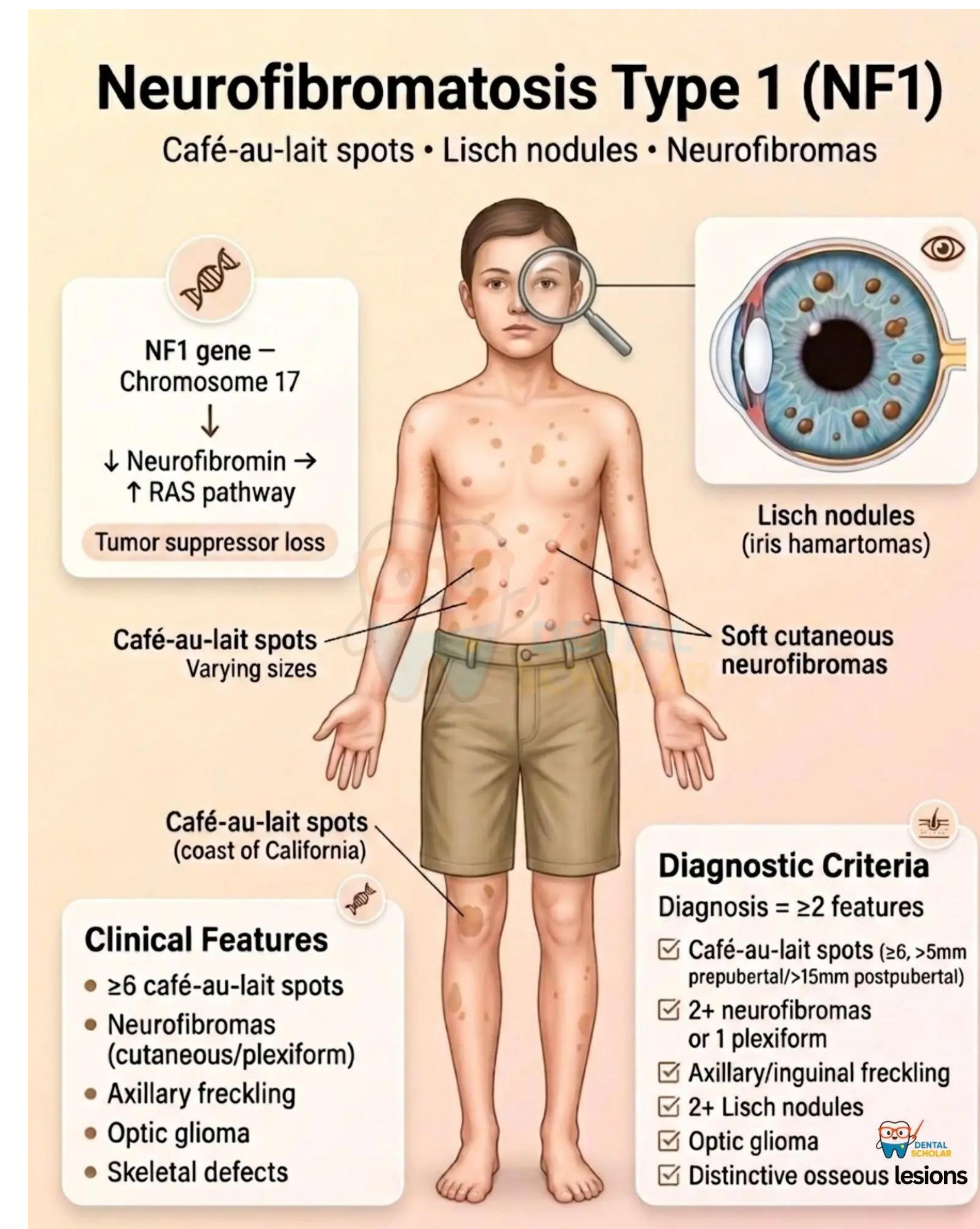


Background

■ **Neurofibromatosis Type 1 (NF1):**

- Autosomal dominant
- Multiorgan neurocutaneous disease
- Characterized by widespread neurofibroma formation that can involve the spinal, intracranial, and airway regions



■ **Anesthetic Considerations:**

- The use of neuraxial anesthesia in obstetric patients with NF1 may be concerning when spinal imaging is unavailable, because of possible spinal or intracranial tumor involvement
- Additional challenges: airway neurofibromas, which may pose a difficult airway risk
- These patients may have other comorbid conditions that may affect anesthetic decision-making (such as cardiac conditions and ICD placement in this patient)

Case Presentation

31-year-old G2P1 at 31 weeks presented with elevated blood pressures concerning for **severe preeclampsia** and non-reassuring fetal heart tones

PMHx: NF1, long QT complicated by ventricular tachycardia/fibrillation arrest s/p ICD placement, chronic hypertension

PSHx: Pancreatic cancer s/p Whipple procedure, prior cesarean delivery

Exam: Reassuring airway & numerous cutaneous neurofibromas and café au lait macules

Plan: Decision to repeat C-section due to resistant hypertension and non-reassuring FHT

Neuraxial anesthesia was deferred due to lack of spinal imaging and potential for unknown spinal or intracranial tumor involvement

Cardiology was consulted and recommended placement of magnet over the ICD to mitigate the risk of inappropriate shocks during electrocautery use

Patient underwent RSI with video laryngoscopy to assess airway for neurofibroma involvement

Proceeded without complication; both maternal/neonatal outcomes were favorable



Example image of café au lait spots and neurofibroma in a patient with NF1



Example image of an intraspinal neurofibroma in a patient with NF1

Discussion / Teaching Points

- Possible tumor involvement: spinal cord, nerves, and soft tissues increases risk for nerve damage and hematomas during neuraxial placement in patients with NF1
- Although there are no formal guidelines for managing parturients with NF1, assessment of prior CT/MRI spine imaging should be made or acquired if time permits
- Airway management may be complicated by neurofibromas: tongue, pharynx, or larynx, as well as by cervical spine abnormalities and scoliosis.
- Video laryngoscopy, fiberoptic scope, and emergency airway equipment should be accessible and prepared
- For those with pacemakers and ICDs, a cardiac consult may be warranted, and care should be taken to interrogate the device to determine risk for electrocautery interference
- When standard obstetric anesthetic approaches are not feasible, prioritize individualized anesthetic planning, careful risk–benefit assessment, and early multidisciplinary collaboration are necessary

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