Labor Pain Management of A Parturient With Friedreich Ataxia, Cardiomyopathy, Super Morbid Obesity and Challenging Lumbar Epidural Catheter Placement, A Case Report

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BACKGROUND

What is Friedreich's Ataxia?

Autosomal recessive neurodegenerative disease

Age of onset: 8–15 years

Pathophysiology:

- GAA trinucleotide repeat expansion mutation
- ↓ Frataxin protein → mitochondrial iron dysregulation → free radicals → cell damage
- Neurons, cardiomyocytes, β-cells affected [1]

Core Manifestations:

- Progressive cerebellar ataxia & gait instability
- Sensory axonal polyneuropathy
- Impaired fine motor coordination
- Hypertrophic cardiomyopathy (up to 60%)
- Diabetes mellitus
- Scoliosis

FA in Pregnancy

Physiologic Pregnancy Changes:

- ↑ Plasma volume → worsens cardiac load
- ↑ LV mass → exacerbates hypertrophy
- Ligament laxity → worsens ataxia & pain

Cardiac Risk [2,4,5]:

- Up to 60% FA patients develop cardiomyopathy.
- Pregnancy-induced cardiovascular changes can precipitate decompensation
- Progression to heart failure, arrhythmias, ST changes

Anesthetic Challenges

Cardiomyopathy

Risk of decompensation

Morbid Obesity (BMI 51.56)

Difficult neuraxial access

Neurologic Deficits

complicates epidural level assessment

GA Risks

Aspiration, hemodynamic instability, mitochondrial sensitivity to Propofol

Case Report

CASE PRESENTATION

Patient Profile

20F•G2P1•39 weeks gestation•BMI 51.56kg/m²

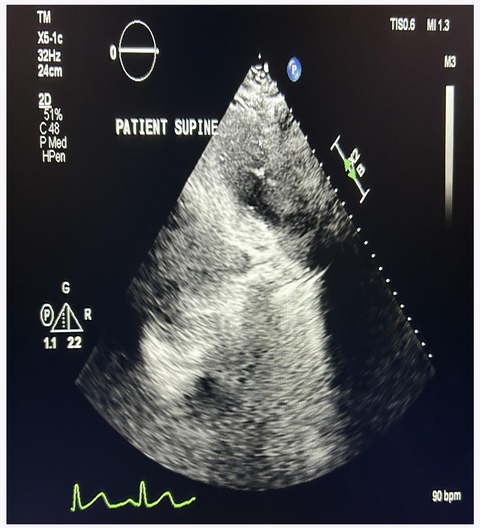
Presented for induction of labor

Diagnoses:

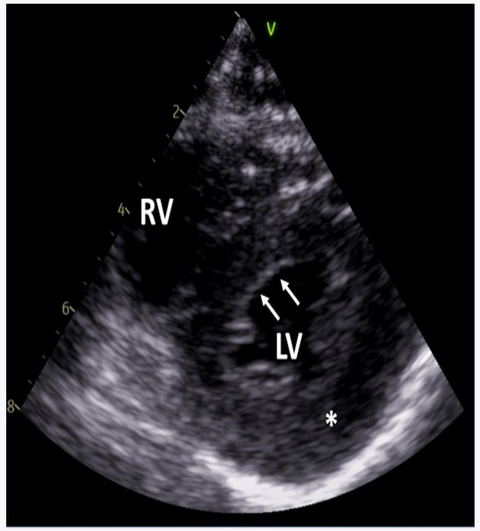
- Friedreich's Ataxia — wheelchair bound
- Hypertrophic Cardiomyopathy
- Sensory axonal polyneuropathy (bilateral)
- Concentric LV hypertrophy — EF 55–60%

Key Workup:

- Genetic testing: MT-CYB m.15182A>G (p.I146V) — variant of uncertain significance, potentially associated with cardiomyopathy
- MRI brain: mild cerebellar volume loss
- EEG: normal; Telemetry: normal
- Pre-induction TTE: inconclusive (positioning artefacts)

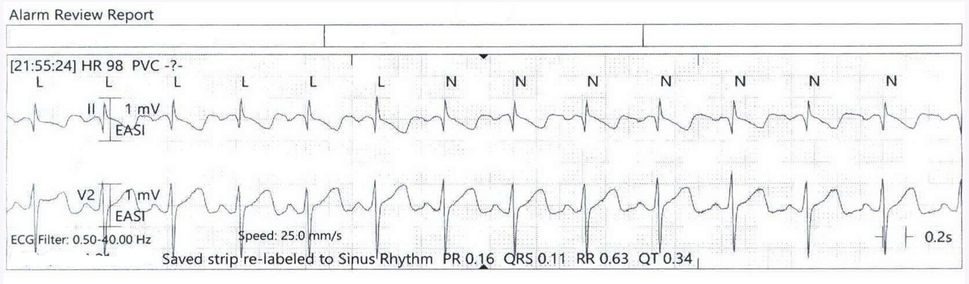


TTE unclear view



LV enlargement in Friedreich Ataxia

Source: BMJ Case Reports CP 2022;15:e248691.



Telemetry on the day of admission

Anesthesia Management

- Ultrasound used to mark epidural space location. No clear image was obtained
- Positional difficulty
- Epidural Catheter was placed after > 5 attempts. Needed replacement.
- LOR 9cm, catheter fixed at 14cm
- CEI with 0.2% Ropivacaine
- Adequate sensory level achieved.
- Uneventful vaginal Delivery
- TTE after delivery: EF 45–50% with mild global LV hypokinesis — new finding

Key Learning Points

1. Multidisciplinary planning (Cardiology, MFM, Anesthesia) is essential in FA parturients — ideally initiated in the antepartum period
2. Epidural analgesia is the preferred modality in FA with cardiomyopathy: gradual sympathectomy limits hemodynamic swings and avoids GA risks
3. Morbid obesity + FA significantly complicates neuraxial technique — ultrasound guidance and a low threshold for catheter replacement are critical
4. Pre-existing sensory deficits from FA may mask neuraxial block assessment; clinical correlation and repeat evaluation are necessary
5. Pregnancy can unmask or worsen FA cardiomyopathy — postpartum echocardiography is warranted even in previously stable patients

References

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