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Introduction

Creutzfeldt-Jakob Disease (CJD) is a rare, progressive neurodegenerative disorder characterized by rapid cognitive decline and neurological impairment, typically leading to death within months. CJD is classified into four forms: **sporadic CJD**, **genetic CJD**, **variant CJD (vCJD)**, and **iatrogenic CJD**.^[1,2]

- Sporadic CJD**, the most common form, typically affects individuals over 60 but can also present in younger patients, as seen in this case.
- Genetic CJD** is inherited and typically presents earlier in life.
- vCJD**, associated with prion transmission through contaminated food, primarily affects younger individuals.
- Iatrogenic CJD** results from medical procedures involving prion-contaminated equipment.

Anesthetic management for CJD patients is complex due to risks of **neurotoxicity** from anesthetics and **prion transmission** during medical procedures. Given the rapid progression of the disease, anesthetic techniques must be carefully tailored to minimize complications. This case report discusses a **33-year-old female** with **sporadic CJD** undergoing **dilation and evacuation** following intrauterine fetal demise, highlighting key anesthetic considerations and protocols.

Case Description

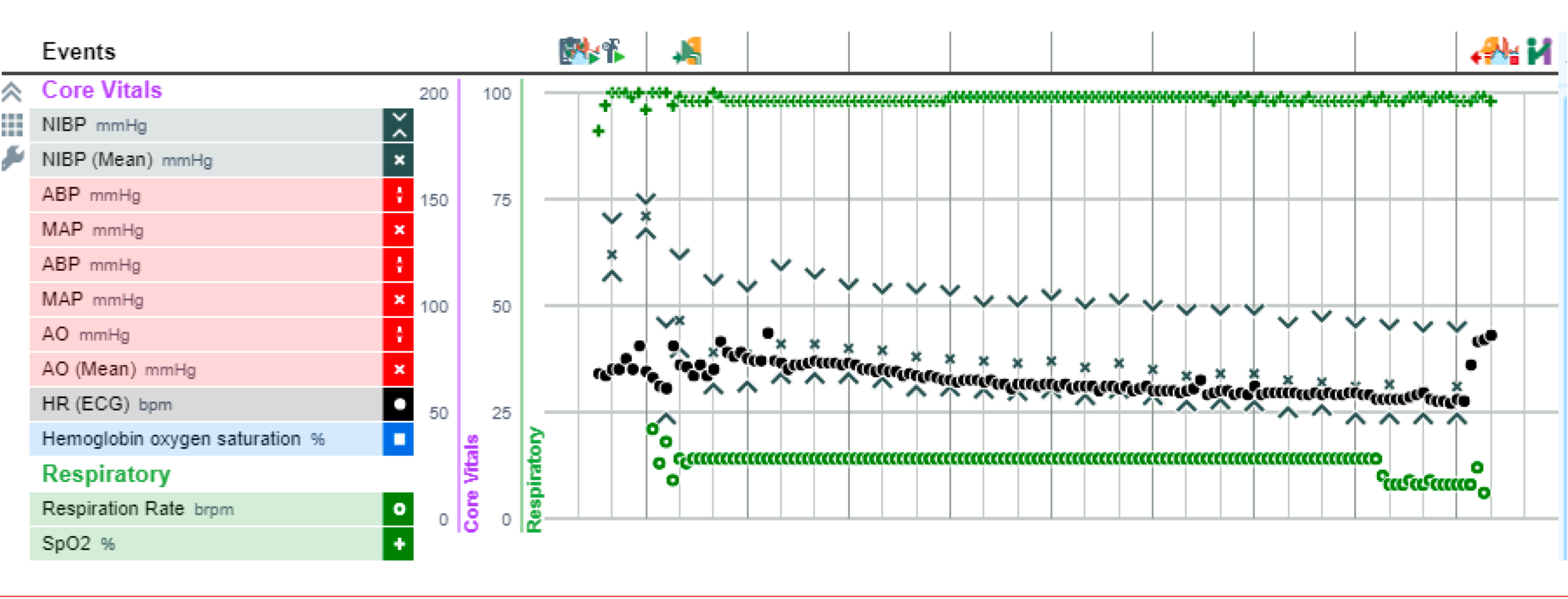
A 33-year-old female with **diagnosed sporadic CJD** was scheduled for a **dilation and evacuation (D&E)** following intrauterine fetal demise at 26 weeks gestation.

Preoperative Assessment:

- Vital Signs:** Stable
- Neurological Status:** The patient was oriented to name and place but could not answer complex questions, responding only with "ok" or "yes." She was unaware of her pregnancy status and the intrauterine demise, relying on family members for daily activities.
- Laboratory Findings:** Macrocytic anemia detected on preoperative testing.

Anesthesia Plan: Given the patient's cognitive decline and potential for neurotoxic reactions to certain anesthetic agents, a **total intravenous anesthesia (TIVA)** technique was chosen. The anesthetic agents included **propofol** for induction, with **lidocaine** and **glycopyrrolate** for airway management. Maintenance was with **propofol** and **remifentanyl**. A **disposable blade** was used for intubation to minimize infection risks.^[3]

Anesthesia Intraoperative Events



Analysis and Recommendations

Anesthesia management in patients with CJD requires special attention to several key factors:

- Neurotoxicity:** Inhalational anesthetics may worsen neurological decline in CJD patients. To minimize central nervous system impact, **total intravenous anesthesia (TIVA)** with **propofol** and **remifentanyl** was used, avoiding neurotoxic effects from volatile agents.^[3]
- Risk of Prion Transmission:** Prions are highly resistant to sterilization and can be transmitted through contaminated medical equipment. To mitigate this risk, we utilized **single-use disposable equipment** and installed a **charcoal filter** on the anesthesia machine to capture any prion-contaminated aerosols. This aligns with current guidelines for handling prion diseases and helps prevent cross-contamination between patients.
- Avoid Muscle Relaxants:** Given the lack of definitive safety data on the use of muscle relaxants in CJD patients, these were avoided to reduce the risk of prolonged neuromuscular blockade and other potential complications.
- Avoid Invasive Procedures:** Neuraxial anesthesia (e.g., epidural or spinal) carries an elevated risk of transmitting prions through neural tissues, thus it was **avoided** in this case.^[3]

MRI Impression

- Symmetric FLAIR and diffusion abnormality involving bilateral caudate heads and anterior putamen is associated with extensive cortical ribboning within bilateral cerebral hemispheres and prominent generalized parenchymal volume loss for age. The imaging findings are consistent with the stated history of CJD.
- Whole brain and cortical gray matter volume are greater than 2 standard deviations below the mean compared to age matched controls (1 percentile).

Conclusion

Anesthetic management of patients with CJD requires careful consideration of the risks associated with neurotoxicity and prion transmission. In this case, the use of **TIVA** with **propofol** and **remifentanyl**, avoidance of muscle relaxants or neuraxial anesthesia, and the use of **single-use equipment** and a **charcoal filter** on the anesthesia machine were critical in minimizing potential complications. These precautions aligned with current guidelines for handling prion diseases and aimed to reduce the risk of prion contamination during medical procedures. The patient had a smooth recovery with stable vital signs, and no adverse neurological effects were noted, highlighting the effectiveness of a tailored anesthetic approach. This case highlights the importance of thorough planning and specialized protocols in optimizing anesthesia care for patients with neurodegenerative diseases like CJD. Given the rarity and complexity of CJD, **further research** is needed to refine anesthetic practices and establish clearer guidelines for managing these high-risk patients. Continued investigation into prion transmission and the neurotoxic effects of anesthesia agents will be essential in improving outcomes for patients with CJD.

References

1. Sikorska B, Knight R, Ironside JW, Liberski PP. Creutzfeldt-Jakob disease. Adv Exp Med Biol. 2012;724:76-90. doi: 10.1007/978-1-4614-0653-2_6. PMID: 22411235.
2. In CB, Choi YS, Park EY, et al. Anesthetic management in patients suspected of Creutzfeldt-Jakob disease -A case report-. Korean J Anesthesiol. 2011;61(3):262-264. doi:10.4097/kjae.2011.61.3.262
3. Thangavel K, Sawhney C, Bandyopadhyay A. Perioperative concerns in a patient with suspected Creutzfeldt - Jakob disease: A case report and review of literature. Saudi J Anaesth. 2024 Jan-Mar;18(1):120-122. doi: 10.4103/sja.sja_503_23. Epub 2024 Jan 2. PMID: 38313721; PMCID: PMC10833012.