

Navigating Neuraxial Anesthesia in Fontan Physiology and Coagulopathy: A Case Report

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INTRODUCTION

Advancements in surgical techniques and perioperative care have improved long-term survival in patients with Fontan circulation, with modern 30-year survival rates of 75-87%.¹⁻³ Consequently, more women with Fontan physiology are reaching childbearing age. Over 70,000 patients worldwide are currently living with Fontan circulation and this number is projected to double in the next 20 years. Despite these improvements, the complexity of Fontan physiology remains a significant challenge, with up to 50% of patients experiencing major adverse events before reaching adulthood³.

Pregnancy introduces additional challenges due to the hemodynamic shifts of gestation, labor, and delivery, which can precipitate decompensation in these preload-dependent patients. Optimal anesthetic management requires careful risk-benefit assessment of general versus neuraxial techniques. This demands a thorough understanding of Fontan physiology and its interplay with pregnancy, particularly in the context of Fontan-associated liver disease and thrombocytopenia.

CASE PRESENTATION

A gravida 2, para 0 female in her 20s with a history of Fontan circulation was scheduled for an elective cesarean delivery. She was born with hypoplastic left heart syndrome (HLHS) and underwent staged palliation, including the Norwood procedure, Hemi-Fontan, and Fontan with fenestration, followed by fenestration closure.

Preoperative laboratory testing revealed severe thrombocytopenia (platelet count of $45 \times 10^9/L$). Bilirubin and transaminase levels were within the normal range. She was hemodynamically stable with New York Heart Association (NYHA) Class I–II functional status and denied any history of easy bruising or spontaneous bleeding. TEG from the day before surgery showed a maximum amplitude (MA) of 52 mm, which was within the normal range for our institution (threshold > 50 mm).

Given her severe thrombocytopenia and Fontan physiology, the cardiac anesthesiology team was consulted. After a thorough risk-benefit discussion with the patient and her family, the patient elected to proceed with epidural anesthesia.

After a multidisciplinary discussion involving the hematology and obstetric teams, a perioperative platelet transfusion strategy was implemented to minimize bleeding risk. The patient received one unit of platelets six hours prior to surgery. As the team prepared for epidural anesthesia, an additional unit was given three hours before epidural placement. Repeat laboratory testing and TEG after the first two units showed a platelet count of $52 \times 10^9/L$ and a MA of 55 mm.

Immediately prior to epidural placement, the patient received the third unit of platelets along with 0.3 $\mu g/kg$ of desmopressin to further reduce hematoma risk.

CASE PRESENTATION CONTINUED

An epidural catheter was placed successfully at the L3-4 interspace using an 18-gauge Tuohy needle, and an arterial line was inserted for continuous hemodynamic monitoring. Intraoperatively, the patient received another unit of platelets. The sequence of platelet transfusions, coagulation assessments, and epidural placement is summarized in **Figure 1**. Epidural anesthesia was achieved using incremental doses of 2% lidocaine, totaling 20 mL over 10 minutes, which provided a T6 sensory level without significant hemodynamic disturbance.

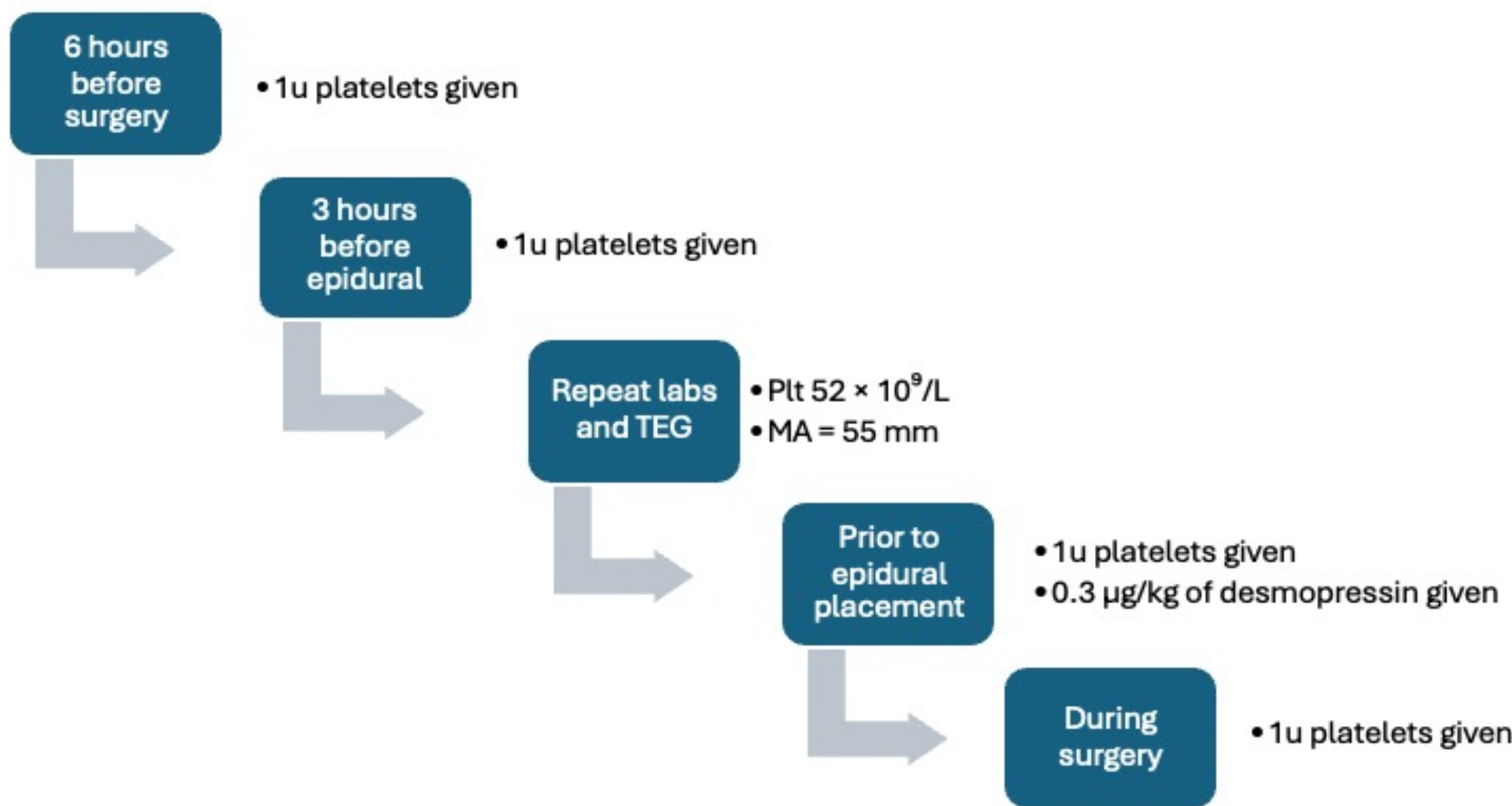


Figure 1. Timeline of perioperative transfusions, TEG results, and epidural placement in a thrombocytopenic Fontan parturient.

To prepare for possible conversion to GA, the respiratory therapy team arranged inhaled nitric oxide (NO) in the operating room to mitigate pulmonary vascular resistance (PVR) elevation in the event of positive pressure ventilation (PPV).

OUTCOMES

The cesarean delivery was uncomplicated, and both the patient and neonate had favorable outcomes. The patient remained hemodynamically stable throughout the procedure. The epidural catheter was removed at the end of surgery without incident, and no neurological complications were observed.

DISCUSSION

Key Hemodynamic Principles in Fontan Physiology

- Maintain stable pulmonary vascular resistance (PVR) to preserve passive pulmonary blood flow.
- Maintain adequate preload to ensure effective ventricular filling.
- Support ventricular contractility to maintain cardiac output.
- Avoid increases in afterload to preserve forward flow and prevent pulmonary congestion.

Anesthetic Approach in Fontan Parturients

- No definitive guidelines exist for cesarean delivery in Fontan physiology; evidence remains limited.
- Epidural anesthesia preferred over spinal due to:
 - Ability to titrate local anesthetic gradually → avoids abrupt sympathectomy.
 - Avoidance of positive pressure ventilation (PPV) required in general anesthesia, which can impair Fontan circulation.



DISCUSSION CONTINUED

Risk of Neuraxial Anesthesia in Thrombocytopenia

- Major concern: epidural hematoma risk in severe thrombocytopenia.
- SOAP guidance:
 - $>70 \times 10^9/L \rightarrow$ low risk.
 - $50-70 \times 10^9/L \rightarrow$ individualized assessment.
 - $<50 \times 10^9/L \rightarrow$ no established recommendations.
- Large multicenter study (n=1,524):
 - No epidural hematomas reported.
 - Estimated risk using rule of three
 - 3% for platelets $50-70 \times 10^9/L$.
 - 11% for platelets $<50 \times 10^9/L$.

Evidence From Lumbar Puncture Literature

- Lumbar punctures frequently performed in severe thrombocytopenia without increased risk when platelet transfusion is provided.
- Retrospective study of 83,000 lumbar punctures:
 - No hematomas in patients with platelet counts $<70 \times 10^9/L$.
 - No correlation between coagulopathy and hematoma formation.
- Suggests actual neuraxial risk may be lower than traditionally assumed, but epidural $\neq$ lumbar puncture (different risk profiles)

Comparison to Prior Literature

- A 2020 report: neuraxial anesthesia used for cesarean delivery in a patient with HELLP syndrome and pulmonary hypertension with platelets $<50 \times 10^9/L$; CSE used with perioperative platelet transfusion.
- Current case is the first known report of neuraxial anesthesia in a patient with both Fontan physiology and severe thrombocytopenia.

CONCLUSIONS

- Severe thrombocytopenia alone should not automatically preclude neuraxial anesthesia, especially when general anesthesia poses significant hemodynamic risk, as in parturients with Fontan physiology.
- The current consensus from the Society for Obstetric Anesthesia and Perinatology supports neuraxial anesthesia at platelet counts above $70 \times 10^9/L$, but lower counts may still be reasonable in asymptomatic patients with reassuring coagulation studies and multidisciplinary agreement.
- In preload-dependent Fontan patients, carefully titrated epidural dosing allows gradual hemodynamic shifts, reducing the risk of instability compared with spinal or general anesthesia.

REFERENCES (USE QR CODE TO THE LEFT)