

CASE REPORT

WALKING THE TIGHTROPE: NAVIGATING ANESTHETIC CHALLENGES IN A PEDIATRIC CARDIAC SURGERY CASE WITH PORTAL HYPERTENSION AND SEVERE THROMBOCYTOPENIA

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Abstract

Background: Anesthetic management of pediatric patients for corrective cardiac surgery is highly complex when portal hypertension, severe thrombocytopenia and inherited thrombophilia coexist. The interplay between bleeding and thrombotic risks requires meticulous planning.

Case report: We present the case of a 5-year-old child with congenital heart disease and severe thrombocytopenia who was scheduled for corrective cardiac surgery. The child was hospitalized for the first time 6 months earlier because of a sudden abdominal pain and hematemesis, leading to the diagnosis of portal hypertension and inherited thrombophilia. At the time he presented for surgery, the child had very low platelets, $36 \times 10^9/L$ with no other abnormalities in the laboratory results. Anesthetic management focused on balancing hemorrhage and thrombosis, optimizing coagulation with viscoelastic testing, goal-directed transfusion therapy and maintaining stable hemodynamics. Surgery was completed successfully, with close postoperative monitoring in the pediatric cardiac ICU.

Conclusion: This case highlights the challenges of managing a child with combined bleeding and thrombotic risks. Individualized anesthetic strategies and multidisciplinary approach are essential to achieve safe outcomes in such high-risk background.

Key words: *congenital cardiac surgery, portal hypertension, thrombocytopenia, thrombophilia*

Introduction

Portal vein thrombosis (PVT) is a rare but serious condition leading to impaired hepatic blood flow, specifically obstruction in the blood flow of the splenic and superior mesenteric veins. To date, the pathogenesis of PVT in children still remains unexplained, yet it is major cause of portal hypertension in children and adolescents. (1) In pediatric patients, portal vein thrombosis can result from underlying prothrombotic disorders, abdominal infections or surgical complications, and may be manifested by abdominal pain, vomiting or gastrointestinal bleeding. Hereditary thrombophilias that are known to predispose to venous thrombosis and PVT include certain mutations of the prothrombin, factor V Leiden or methylenetetrahydrofolate reductase genes or deficiency of one of the natural anticoagulant proteins C and S. (1) In our patient, there were mutations in several thrombophilic genes and a deficiency of anticoagulant protein C, but this result was obtained less than 3 months after

the diagnosis of portal vein obstruction, therefore, falsely low levels related to active thrombosis are not excluded. Another test in further time was not done.

PVT often manifests with symptoms like splenomegaly, hepatomegaly or ascites, that result from ensuing portal hypertension (PH). Due to the obstruction in the portal vein and the increased hepatic venous pressure, a net of portosystemic collaterals is formed, resulting in increased flow in splenorenal, paraumbilical or gastric and esophageal veins. (2)

Anesthesia in patients with portal hypertension is really challenging. The anesthetist must carefully balance hemodynamics, choose drugs with minimal hepatic metabolism and be prepared for significant bleeding risk. The patients with portal hypertension often have low systemic vascular resistance and high cardiac output. (3) This makes them very sensitive to anesthetic agents, which can further lower blood pressure. In our patient with coarctation of the aorta, there is a severe afterload mismatch: the upper body is hypertensive, while the lower body is hypoperfused. It is therefore essential to maintain adequate proximal perfusion pressure without compromising distal circulation (kidney, gut, liver).

Sometimes, there are respiratory considerations too. For instance, if there is an ascites, it can cause reduced lung volumes, increase risk of hypoxemia and impaired ventilation under anesthesia. Massive ascites increases the intra-abdominal pressure, predisposing the patient to reflux and aspiration during induction. (4)

Also, sudden increases in venous pressure (e.g. from intubation, coughing or fluid overload) may precipitate variceal bleeding. (5) Often there is thrombocytopenia due to splenic sequestration and hypersplenism. Impaired liver function reduces synthesis of coagulation factors. Anesthesia must minimize fluctuations in venous pressure and be prepared for massive transfusion (6).

Case Presentation

A 5-year-old child, weighting 17kg, with coarctation of the aorta was admitted for elective surgery, reconstruction of the aorta with end-to-end anastomosis. Six months prior, the patient had experienced sudden abdominal pain and hematemesis, prompting investigations that revealed extrahepatic portal vein thrombosis due to inherited mutations of several thrombophilic genes. Given that the obstruction was not complete, i.e., tunnels, so-called cavernomas, were created in the thrombus itself, surgical creation of a shunt to bypass the portal vein was not necessary. The patient underwent antegrade transvenous embolization of the varicose fundal gastric veins 2 months prior to surgery in Turkey, but on his last checkup, he had another enlarged gastric varices on top of splenomegaly and severe thrombocytopenia, which are associated with continued persistence of portal hypertension.

On admission, laboratory evaluation showed severe thrombocytopenia (platelets: $36 \times 10^9/L$), coagulation parameters were otherwise normal, and unremarkable renal and hepatic function except for evidence of portal hypertension confirmed by ultrasound.

Preoperatively, the anesthetic team conducted a detailed risk assessment, focusing on balancing the risk of bleeding and thrombotic complications and multidisciplinary collaboration with hematology, cardiology and surgical teams to formulate a perioperative plan.

In consultation with the hematology and transfusion specialists, we treated the thrombocytopenia with steroids and immunoglobuline, but the platelet count did not improve during treatment. Following bone marrow biopsy, a normal count of blood elements was found with no pathological cells, so it was confirmed that the thrombocytopenia was the result of increased sequestration of platelets in the portal circulation and spleen. The same conclusion was confirmed by the resistance to an increased platelet count following platelet transfusion.

We suggested that the surgery should be delayed until splenectomy or embolization of the splenic artery is performed, but due to the severity of the coarctation, impossibility to implant a stent to the stenotic area and the unsafe use of antiplatelets drugs, the surgical team insisted that there is an urgent need for open surgery.

On the day of surgery, the patient was transfused with a unit of platelets (six pooled platelets following apheresis of donor whole blood, approximately 10ml/kg) two hours prior to surgery. The rechecked platelet count immediately before surgery was $62 \times 10^9/L$. He was also anemic with a haemoglobin level of 87g/L. The preoperative international normalized ratio (INR) was 1.14 and activated partial thromboplastin time (aPTT) was 24 seconds (21-36 sec).

The induction of anesthesia was done with titrating doses of propofol and rocuronium. In patients with coarctation of the aorta, propofol provides rapid and reliable induction; however, its vasodilatory and negative inotropic effects may precipitate hypotension, thereby reducing coronary, cerebral and systemic perfusion distal to the coarctation site. Therefore, careful titration is essential to avoid abrupt decreases in systemic vascular resistance, and adequate intravascular volume status should be ensured (7). Although rocuronium is mainly eliminated by the liver, it can be safely used in patients with portal hypertension if the hepatic function is preserved (8).

After induction of anesthesia, the central venous access and arterial line were established carefully to minimize trauma, with ultrasound guidance to avoid inadvertent vascular injury and bleeding. Standard ASA monitors were applied, including invasive arterial pressure, central venous pressure and continuous ECG. Regarding hemodynamic management, we inserted two arterial lines - one pre-ductal (right arm) to demonstrate perfusion to brain/heart and one post-ductal (femoral) to demonstrate distal perfusion. Therefore, blood pressure was closely monitored (beat-to-beat) and maintained around 85-95 mmHg systolic, to optimize perfusion without increasing portal pressures and reduce the risk of variceal bleeding. We also did not insert nasogastric tube to avoid trauma of the gastric mucosa.

Following initiation of surgery and during the chest opening, another unit of platelets was transfused and one unit of 350ml packed red blood cells as well. Intraoperatively, we administered 15mg/kg (~250mg) bolus dose of tranexamic acid followed by continuous infusion 5mg/kg/h. We also monitored the blood gases every 30 min for signs of bleeding and used ROTEM for real-time guidance and to maintain hemostatic balance while avoiding overcorrection that could exacerbate thrombosis risk. ROTEM analysis had not shown abnormal results preoperatively or postoperatively. During clamping of the aorta we did not use heparin because of the risk of bleeding, while the total cross clamping time was less than 15 minutes.

Surgery was completed successful and the patient was transferred to the pediatric cardiac ICU with ongoing hemodynamic and coagulation monitoring. Postoperative platelet count was $67 \times 10^9/L$, thus no further platelet transfusions was administered. The tranexamic acid infusion was continued in the first 24 hours after surgery. Early extubation in the first six hours after surgery was done with careful monitoring of hypertension and bleeding.

A single thoracic drain was placed intraoperatively, with postoperative drainage amounting to 20ml of blood within the first 24 hours.

Two months after surgery, the patient platelet count was moderately low, $58 \times 10^9/L$, so despite the thrombophilic genetic background he was not recommended any antithrombotic drugs.

Discussion

Preoperative thrombocytopenia could be associated with an increased risk of bleeding. Excessive bleeding leads to increased mortality, morbidity, transfusion requirements and reintervention. (9) Platelet thresholds in pediatric cardiac surgery are not always uniform, but there are widely accepted practice ranges. The general principle is that platelet count should be $\geq 100 \times 10^9/L$ before major surgery involving cardiopulmonary bypass (CPB). If there is a moderate thrombocytopenia ($50-100 \times 10^9/L$), surgery can still be performed with availability of platelet transfusion and point-of-care coagulation monitoring (ROTEM/TEG). In case of severe thrombocytopenia ($<50 \times 10^9/L$), elective cardiac surgery is considered unsafe and correction with platelet transfusion or addressing the underlying cause is generally recommended before proceeding (10).

Children tolerate thrombocytopenia somewhat differently than adults, but CPB amplifies platelet dysfunction, making higher counts desirable. Fortunately, the corrective cardiac surgery in our patient did not require CPB. The platelet function (qualitative) can be just as important as the absolute count, so viscoelastic testing (ROTEM/TEG) is increasingly used to guide transfusions. (11)

There have been some reports where splenectomy was performed in an attempt to improve platelet counts pre-cardiac surgery and also the use of off-pump cardiac surgery to reduce post-operative bleeding. There is one case report that suggests that safe and successful cardiac surgery is feasible in a group of patients despite a very low platelet count ($< 20 \times 10^9/L$). (12) In this group of patients, coagulopathy should be promptly treated in the immediate postoperative phase and management could be guided by point-of-care TEG analysis and this may be possible in the future with a more reliable point-of-care platelet function assessment kit.

Genetic thrombophilia predisposes to arterial and venous thrombosis. In cardiac surgery, the risk is heightened due to altered hemostasis and endothelial activation during CPB, surgical manipulation of the aorta which brings risk of clot formation at repair site and postoperative reduced mobility and central lines. (13) These patients often require carefully titrated anticoagulation, both intraoperatively and postoperatively. Standard heparinization $400 IU/kg$ is used for CPB; ACT monitoring is essential. If surgery is done without CPB (such as ours) heparin bolus is still usually given to reduce clot risk. We did not proceed with this because the surgeons were convinced that they will be finished with the aortic repair in less than 15 minutes. A blessing in disguise was that our patient had low platelet count, so he did not

require long- term postoperative coagulation or antithrombotic prophylaxis. Thrombosis involving the aortic arch or stenotic coarctation segment is rare in children; most published reports are isolated neonatal/infant cases or thrombotic complications after stent/graft placement. (14-17)

Two months after surgery the child is recovering well, with no evidence of rethrombosis and stable hemodynamic status. Importantly, although thrombocytopenia persists (platelet count $53 \times 10^9/L$ on last checkup), there has been no clinically significant bleeding or deterioration. The absence of new thrombotic events in the early postoperative period is reassuring, considering the prothrombotic background associated with portal hypertension.

Long-term follow up will be essential to monitor vascular patency, control of portal hypertension and the evolution of thrombocytopenia in this patient.

Overall, this case demonstrates that with meticulous perioperative management, excellent outcomes are achievable, even in high risk pediatric patients with coexisting hematologic abnormalities and congenital cardiovascular malformations.

Conclusion

This case underscores the complexity of anesthetic management in children with combined bleeding and thrombotic risks. Portal hypertension and splenomegaly contribute to severe thrombocytopenia and increased risk of bleeding during surgery, while thrombophilia predisposes to intraoperative and postoperative clot formation. Multidisciplinary coordination, goal-directed transfusion strategies and vigilant intra and postoperative monitoring are essential to ensure safe outcomes.

References:

1. Pietrobattista A, Luciani M, Abraldes JG, et al. Extrahepatic portal vein thrombosis in children and adolescents: Influence of genetic thrombophilic disorders. *World J Gastroenterol.* 2010 Dec 28;16(48):6123-7. doi: 10.3748/wjg.v16.i48.6123. PMID: 21182228; PMCID: PMC3012577.
2. Vrijburg M, Sari S, Koot BGP, Fijnvandraat K, Klaassen ILM. A high rate of post thrombotic complication in pediatric portal vein thrombosis. *Thrombosis Research,* 2023 Sep 30; Vol 231, 44 – 49. doi: 10.1016/j.thromres.2023.09.015
3. Gilbert-Kawai N, Hogan B, Milan Z. Perioperative management of patients with liver disease. *BJA Educ.* 2022 Mar;22(3):111-117. doi: 10.1016/j.bjae.2021.11.006. Epub 2022 Jan 31. PMID: 35211328; PMCID: PMC8847800.
4. Friedman LS. Surgery in the patient with liver disease. *Trans Am Clin Climatol Assoc.* 2010;121:192-204; discussion 205. PMID: 20697561; PMCID: PMC2917124.
5. Garcia-Tsao G., Bosch J. and Groszmann R.J. (2008), Portal hypertension and variceal bleeding—Unresolved issues. Summary of an American Association for the study of liver diseases and European Association for the study of the liver single-topic conference[‡]. *Hepatology,* 47: 1764-1772.
6. Ramsay M. Anesthesia for patients with liver disease. In: Butterworth JF, Mackey DC, Wasnick JD, editors. *Morgan & Mikhail's Clinical Anesthesiology.* 5th ed. New York: McGraw-Hill Education; [accessed 2025 Sep 26].

7. Williams GD, Jones TK, Hanson KA, Morray JP. The hemodynamic effects of propofol in children with congenital heart disease. *Anesth Analg*. 1999 Dec;89(6):1411-6. doi: 10.1097/00000539-199912000-00016. PMID: 10589618.
8. Magorian T, Wood P, Caldwell J, et al. The pharmacokinetics and neuromuscular effects of rocuronium bromide in patients with liver disease. *Anesth Analg*. 1995 Apr;80(4):754-9. doi: 10.1097/00000539-199504000-00018. PMID: 7893030.
9. Salenger R, Arora RC, Bracey A, et al. Cardiac surgical bleeding, transfusion, and quality metrics: Joint consensus statement by the Enhanced Recovery After Surgery Cardiac Society and Society for the Advancement of Patient Blood Management. *Ann Thorac Surg*. 2025;119(2):280–95.
10. Kaufman RM, Djulbegovic B, Gernsheimer T, et al. Platelet transfusion: a clinical practice guideline from the AABB. *Ann Intern Med*. 2015;162(3):205–13. doi:10.7326/M14-1589.
11. Machovec KA, Jooste EH. Pediatric transfusion algorithms: coming to a cardiac operating room near you. *J Cardiothorac Vasc Anesth*. 2019 Jul;33(7):2017–29. doi:10.1053/j.jvca.2018.12.008. PMID: 30686658. Ashoub A, Lakshmanan S, Luckraz H. Cardiac surgery in a patient with severe thrombocytopenia: How low is too low?. *Ann Card Anaesth* 2013;16:215-7.
12. Massoudy P, Thielmann M, Müller-Beissenhertz H, Görlinger K, Dietrich W, Herget-Rosenthal S, Jakob H. Thrombophilia in cardiac surgery-patients with symptomatic factor V Leiden. *J Card Surg*. 2009 Jul-Aug;24(4):379-82. doi: 10.1111/j.1540-8191.2008.00761.x. PMID: 19040405.
13. Wieland I, Jack T, Seidemann K, et al. Neonatal aortic arch thrombosis: analysis of thrombophilic risk factors and prognosis. *Cardiology in the Young*. 2014;24(1):33-39. doi:10.1017/S1047951112002077
14. Giacaman N, Tos SM, Ibdah MG, Ismail MKM, Ellahham NH. A child diagnosed with midaortic syndrome and inherited thrombophilia after presenting with a stroke: A case report. *Ann Med Surg (Lond)*. 2022 Aug 19;81:104455. doi: 10.1016/j.amsu.2022.104455. PMID: 36147148; PMCID: PMC9486722.
15. Williams FZ, Clampitt-Holsenbeck A, Lopilato A, et al. Successful surgical management of aortic arch thrombosis in the neonate. *Ann Thorac Surg*. 2021 Feb;111(2):e105–8. doi:10.1016/j.athoracsur.2020.05.091. PMID: 32673659.
16. Gürsu HA, Varan B, Oktay A, Özkan M. A case of neonatal arterial thrombosis mimicking interrupted aortic arch. *Turk Pediatri Ars*. 2015 Jun 1;50(2):118-22. doi: 10.5152/tpa.2015.869. PMID: 26265897; PMCID: PMC4523985.