**Title**: Anesthetic care of a patient with Bernard-Soulier syndrome for posterior spinal fusion

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**Introduction:** Bernard-Soulier syndrome (BSS) is characterized by excessive and prolonged bleeding due to thrombocytopenia and platelet dysfunction with increased platelet size and deformability.The primary defect in BSS involves the platelet glycoprotein (GP) Ib-IX-V complex, which is important in initiating platelet aggregation and thrombosis after vascular injury by facilitating the adhesion of platelets to von Willebrand factor.The lack of or defect in the functioning of GPIb-IX-V results in a compromised ability to initiate platelet adhesion and aggregation resulting in a bleeding diathesis. We present a patient with BSS who presented for posterior spinal fusion for idiopathic scoliosis.

**Case Report:** The patient was a 17-year-old, 70.9 kg girl with macrothrombocytopenia and a baseline platelet count between 50,000-70,000/mm3. Due to previous limited responses to platelet transfusions, it was decided to monitor *in vivo* coagulation function using the rotational thromboelastogram (ROTEM®). The values were normal except for a prolongation of clot formation time (CFT). Anesthetic induction involved propofol (200 mg), lidocaine (100 mg), and sufentanil (20 μg). Maintenance anesthesia included desflurane and a sufentanil infusion, and was supplemented by continuous infusions of lidocaine and esmolol. She received intraoperative recombinant factor VIIa and tranexamic acid. In total, she received 3 units of pheresed platelets, 2 pre-operatively and 1 post-operatively. Up to 10 days postoperatively, ε-amino caproic acid (Amicar) was administered. Although she lost 900mL of blood during surgery and her platelet count decreased to a 39,000/mm3, there was no clinically concerning bleeding and her ROTEM® values remained stable. Her postoperative course was unremarkable and she discharged home on postoperative day 5.

**Discussion**: Our case was unique in two respects including our patient’s refractory state to the administration of allogeneic platelets with minimal increase in the platelet count. Despite this, work-up did not reveal platelet antibodies or other issues that might be amenable to corticosteroid therapy or plasmapheresis. To date, there are a limited number of reports in the literature regarding the perioperative care of pediatric patients with BSS. While the mainstay of therapy includes platelet transfusions, adjunctive therapies have included desmopressin, anti-fibrinolytic agents (tranexamic acid or ε-aminocaproic acid), or recombinant factor VIIa. Given our patient’s inadequate response to platelet transfusions, we chose tranexamic acid and recombinant VIIa to augment platelet. Our anecdotal experience adds to the literature suggesting the demonstration of utility of bedside, point-of-care coagulation function monitoring using the ROTEM®. With such care, even invasive surgical procedures can be accomplished with minimal sequelae related to the primary bleeding disorder.

**Conflict of Interest:** Authors have no conflict of interest to declare.