Shwachman-Diamond Syndrome (SDS) is a rare autosomal recessive disorder characterized by pancreatic insufficiency, hematologic dysfunction, and skeletal abnormalities. Neutropenia is present in 98% of SDS patients, leading to frequent infections. Other organ systems are commonly involved, presenting a challenge in the perioperative period. Patients with SDS often require multiple surgical interventions, and chronic pain states are common, making the achievement of adequate pain control an ongoing concern.

Our patient, a 15 year-old male with SDS, presented with severe abdominal pain, ascites, and dyspnea. The patient had recently developed graft-versus-host disease (GVHD) following bone marrow transplantation. The patient had significant liver failure, pulmonary insufficiency, and renal dysfunction, yet his main complaint was intractable abdominal pain. He was scheduled for a flexible bronchoscopy and bone marrow aspiration biopsy.

Preoperatively, the patient rated his pain as 8/10 and physical exam included bilateral rales and severe ascites. Induction of general anesthesia was accomplished with an intravenous rapid-sequence induction using propofol and succinylcholine. After the airway was secured with an endotracheal tube, sevoflurane and dexmedetomidine were used for maintenance of anesthesia. Although postoperative pain was expected, opioids were not administered in order to optimize the setting for extubation. At the end of the procedures, the patient was extubated and transported to the recovery room.

Postoperatively, the patient was given IV morphine for pain control once fully awake. Due to a lack of adequate pain control, the patient was switched to a hydromorphone patient-controlled anesthetic (PCA). Since there was concern for his compromised pulmonary status, opioid-sparing agents such as IV ketorolac and acetaminophen were considered. In the setting of thrombocytopenia, liver failure, and immunosuppression, those modalities were deemed not suitable. An ultra-low dose ketamine infusion was started two days later as an opioid adjuvant, and this did provide modest pain relief. Due to continued pain, the decision was made to initiate IV methadone therapy on postoperative day 8 given its long duration of action and lack of active metabolites. The patient experienced adequate pain relief and was transitioned to oral methadone. He was discharged home with a plan for a slow taper, and additional PO hydromorphone was available for breakthrough pain.

Patients with SDS often have recurrent respiratory infections, decreased FRC, and in this case, profound ascites. Respiratory collapse can occur if caution is not used when administering opioids. Other pain modalities, such as acetaminophen, are not recommended as they can mask the signs and symptoms of acute infections. For these reasons, a multi-modal approach to perioperative pain control in these challenging patients is suggested.

References: