Anesthesia and Perioperative Care for Patients with Metachromatic Leukodystrophy

Author(s): JJ Mooney, CB Berde

Affiliation(s): Children's Hospital, Boston and Harvard Medical School

Introduction: Metachromatic Leukodystrophy (MLD) is a rare (estimated at 1 in 40,000 births) lysosomal storage disease that results in the deposition of cerebroside sulfate in tissues, particularly the brain. It is most commonly due to a defect in the enzyme arylsulfatase-A, a cerebroside sulfatase. Three variants have been described, according to the age of onset and severity of progression. Infantile and juvenile forms progress to profound cognitive and motor deterioration, severe spasticity, and mortality during childhood. No effective treatments are currently available, though stem cell transplantation has been explored. Two previous reports on anesthetic management (1,2) are limited to single cases.

Methods: After IRB approval and waiver of informed consent, electronic medical records and hospital databases were searched for the diagnosis of metachromatic leukodystrophy. Charts were reviewed, and diagnoses were confirmed. Anesthetic records and postoperative records were reviewed for management practices, outcomes, and morbidity and mortality.

Results: 26 anesthetics were performed among 10 patients, as summarized in table 1. There were no cases of unexpected intraoperative hemodynamic instability or unusual responses to specific drug classes, including depolarizing (3 cases) or non-depolarizing (16 cases) neuromuscular blocking drugs (1). Among 3 patients who underwent implantation of baclofen pumps, all subsequently responded to intrathecal baclofen with a beneficial effect on their spasticity.
Major complications occurred in 3 patients, leading to 2 postoperative deaths. A 7 year old had a seizure in the PACU without sequelae. A 3 year old developed stridor and pneumonia on day 3 postoperatively, which progressed to respiratory failure and death on hospital day 27. A 14 year old underwent posterior spinal fusion, complicated by 7 liter blood loss. Following prolonged postoperative ventilation with two failed extubations prior to successful extubation, she was discharged to home in palliative care, with frequent desaturation during sleep and difficulty with pulmonary toilet. She developed pneumonia and died at home 3 months following her surgery.

Discussion:
MLD produces cognitive deterioration, severe spasticity, and profound weakness, which can predispose to respiratory difficulties postoperatively. Available information does not lead to firm recommendations regarding preferred choice of anesthetic agents or techniques.

Table 1 Operative Procedures for Patients with MLD

Procedures	Frequency
Endoscopic Procedures	7
Biopsies	7
Baclofen Trial/ Pump Placement	5
Scoliosis Surgery	2
Intra-thoracic Procedures	1
Intra-Abdominal Procedures	1

Miscellaneous	10

References

1. Anaesthesia 1977; 32: 644-6.
2. Paediatric Anaesthesia 2003; 13:733-4.