Anesthesia Appears to be Safe in Patients with Progressive Leukodystrophies

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Leukodystrophy Information

- **Krabbe Disease**, an autosomal recessive disease, causes a reduction in galactocerebrosidase (GALC) activity
  - Onsets: Early infantile (3-6 mo.), late infantile (6 mo.-2 yr.), juvenile (2-16 yr.), adult (older than 16 yr.)
- **Metachromatic Leukodystrophy (MLD)** is caused by a deficiency in arylsulfatase A (ASA) and saposin B activity
  - Onsets: Late infantile (before 2.5 yr.), juvenile (2.5-16 yr.), adult (older than 16 yr.)
- Clinical Manifestations: Hypotonia, hypertonia, gastroesophageal reflux disease, aspiration, apneic episodes, seizure episodes

Methods

This is a retrospective review of medical records of patients who received anesthesia between 2012 and 2014. Families signed a University of Pittsburgh IRB consent for review of their medical records. The procedures were all done at the Children’s Hospital of Pittsburgh of UPMC. All of the patients were evaluated at the Program for the Study of Neurodevelopment in Rare Disorders (NDRD). Information was collected from the medical and anesthesia records. Anesthesia was provided for MRIs, central line removal and/or placement, and gastrostomy tube placement. Complications were compared to the ASA and age matched pediatric patients during the same time period. Difficult intravenous (IV) placement was defined as three or more IV attempts.

Results

- 138 anesthetics performed on 53 patients
- 30 patients with Krabbe, 23 patients with MLD
- The complication rate was 8.0% (11/138)

137 anesthetics were provided with a combination of volatile and intravenous medications. One anesthetic was a TIVA.

Conclusions

Progressive leukodystrophies appear to be associated with increased complications. The overall complication rate was 8.0% (11/138) and is higher than the general population. Although the complications were mild and self-limiting, the study is significant because this population may pose clinical challenges for the anesthesia provider and they may require an escalation of care.