

An Interdisciplinary Team Approach to Monthly Sedations for Patients with Hunter Syndrome

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Introduction: Mucopolysaccharidosis (MPS) II (Hunter Syndrome) is a genetic lysosomal storage disorder. Progressive symptoms including developmental delays, joint, bone, and airway involvement begin early in childhood. Promising clinical trials for monthly intrathecal enzyme replacement in pediatric patients with Hunter syndrome are in progress at UNC. These children require monthly sedations for intrathecal enzyme injections. We describe our interdisciplinary team's methods for sedating these patients. The sedation service is composed of a child life specialist, pediatric sedation nurses, nurse practitioners, and a pediatric anesthesiologist.

Discussion: Historically, the MPS II phenotype resulted in a progressively worsening airway due to mucopolysaccharide deposits and gradual cervical spine immobility. Pediatric anesthesiologists provided all sedations due to these airway challenges. Our service has been excited to witness the phenotype changes effected by enzyme replacement therapies. Due to a decrease in airway deposits and an increase in cervical flexibility while on enzyme treatment, the sedation nurse practitioners and nurses now safely care for some of these patients. All patients require monthly triage to identify the appropriate staff and sedation prescription. We discuss the patient's individual phenotype and sedation history. Patients who have an implanted IDDD (intrathecal drug delivery device) may receive anxiolysis with midazolam or a moderate sedation with dexmedetomidine and midazolam in the hands of the nurse practitioner and nurses. Those needing a lumbar puncture for enzyme injection require a general anesthetic by a pediatric anesthesiologist. Many patients begin their journey with us receiving GA, and "graduate" to anxiolysis after several months.

References:

1. Muenzer, J et al., *Rheumatology*, 2011
2. Muenzer, J et al., *Genetics in Medicine*, 2016
3. Kamata, M et al., *Pediatric Anesthesia*, 2017