Impact of Contemporary Treatment of Mucopolysaccharidoses on Airway Management in Children

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Introduction: Patients with mucopolysaccharidosis (MPS) present significant perioperative challenges and have been described as the “worst airway problem in pediatric anesthesia”¹. The natural history of MPS may be altered by new therapies.² We sought to describe the impact of bone marrow transplantation (BMT) and recombinant alpha-L-iduronidase enzyme replacement therapy (ERT) on airway management in the pediatric MPS population.

Methods: Research ethics board approval was obtained for a retrospective review of the database maintained by the metabolic genetics department from 2000 to 2010. Charts were reviewed to identify patients with MPS, determine classification of MPS, type of therapy used and airway management for anesthetic encounters.

Results: 61 children with MPS required 294 anesthetics. The overall rate of difficult airway was 33%. Of 27 patients with Hurler’s syndrome, 63% were treated with BMT and 22% with ERT. In those treated with BMT, 12% had difficult airways, compared with 75% in patients who received neither treatment. 50% of patients with Hurler’s syndrome who had received ERT had a difficult airway. In most of these patients, ERT was commenced over 5 years of age. 89% of patients with Hunter’s syndrome had difficult airways. ERT did not improve airway difficulty. 36% of all anesthetics were conducted without airway instrumentation. Direct laryngoscopy, with or without difficulty, was used in 29% of anesthetics, the laryngeal mask airway in 22%, fiberoptic bronchoscope in 3.5%, and video laryngoscope in 3.8%.

Discussion: Patients with MPS have a 25% reported rate of difficult airway, and up to 54% and 71% in patients with Hurler’s and Hunter’s syndromes, respectively.¹ BMT and ERT appear to slow the progression of MPS, although the benefits of BMT in patients over 18 months of age are reported to be minimal.² We report an 84% reduction of difficult airways in patients with Hurler’s syndrome treated with BMT. Compared to BMT, ERT remains relatively novel and did not improve airway difficulty to the same extent, however the impact of ERT administered in infancy remains unknown. Use of newer equipment has altered the airway management of the pediatric MPS patient. Total intravenous anesthesia techniques may have facilitated minimal airway instrumentation for non-invasive procedures.

References: 1. Anaesthesia 1994; 49(12): 1078-84