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AIRWAY COMPLICATIONS IN PATIENTS WITH MUCCOPOLYSACCHARIDOSIS TYPE 1 (MPS1) RELATED TO SPECTRUM SEVERITY AND TREATMENT MODALITY: A 10 YEAR CASE SERIES REVIEW

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Introduction/Aim:

MPS1 is an inherited metabolic condition, associated with deposition of glycosaminoglycans due to a defect in the alpha-L iduronidase enzyme. MPS1 is a multisystem disorder affecting the neurological, skeletal, cardiac and respiratory systems, historically producing significantly challenging airways [1]. It is a spectrum disorder, including Hurlers, Hurler-Scheie or Scheie Syndrome. Bone marrow transplant (BMT) is now a treatment option, and if carried out prior to two years may significantly reduce the severity of systemic complications. Due to complications related to BMT, patients who cannot be found an early match, or if on the lower end of the severity spectrum may be treated with enzyme replacement therapy (ERT) only. The aim of this study was to assess the incidence of difficult to manage airways and airway complications in patients with MPS1 at Great Ormond Street Hospital, and assess airway difficulty according to treatment modality and spectrum severity.

Methods:

We retrospectively analysed our electronic database of all patients with MPS 1. Data was collected on difficulty with airway management, including face mask ventilation (FMV), LMA insertion, or intubation, and any airway complications peri-operatively.

Results:

We assessed the records of 38 patients. 29 children had received a BMT; 86% under 2yrs of age. The children who received BMT were all diagnosed with Hurlers, whereas the children in the ERT group had predominantly Hurler-Scheie (89%). We analysed 282 anaesthetics for children in the BMT group and 30 anaesthetics for children in the ERT group. 1.8% of anaesthetics in the BMT group documented resolvable difficulty with FMV. In 1.5% of anaesthetics a 'less than ideal fit' was documented with an LMA. Despite the BMT, 31% of intubations were still graded as difficult (requiring a boogie or conversion to videolaryngoscopy). There was one incident of a failed intubation (<0.1% of all intubation attempts). Only 4/29 patients who received a BMT did so after 24months, but we observed a higher incidence of difficult FMV, difficult LMA insertion and difficult intubation in these patients. In the ERT group there was no incidents of difficult FMV or LMA insertion, however 53% of intubations were difficult, and of the difficult intubations, 50% remained difficult with the videolarygoscope.

Discussion:

Reassuringly there is a low rate of difficult FMV/LMA use in both treatment groups, and no incidences of 'can't intubate, can't ventilate.' Children who receive a BMT are now less likely to pose

a significant airway risk, however the anaesthetist should still be prepared with airway adjuncts and videolaryngoscopy, and be particularly aware of those patients who receive a BMT over the age of 2 years. Intubation remains difficult in the ERT group despite less severe spectrum disease, and preparation for videolaryngoscopy combined with fibreoptic intubation would be our recommended practice.

Reference:

1. Walker, R. Darowski, M. Morris, P. Wraith, J. Anaesthesia and Mucopolysaccharoidoses: A review of airway problems in children. Anaesthesia. 1994;49:1078-1084.