Author’s response to reviews

Title: Anesthetic considerations of Adults with Morquio's Syndrome-A Case Report

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Version: 3 Date: 23 October 2009

Author's response to reviews: see over
Dear Editor,

Please find enclosed our manuscript entitled:

“Anesthetic considerations of Adults with Morquio’s Syndrome-A Case Report”

which we wish to resubmit for publication to BMC Anesthesiology under the heading of case reports. We thank the reviewers for their helpful comments, which have greatly assisted in the redraft of the manuscript.

Reviewer one
1. I was disappointed by the fact that the authors focus their attention mainly on the fiberoptic tracheal intubation, generically describing the fiberoptic technique without any report i.e. concerning the endotracheal tube: these patients may require an endotracheal tube that is smaller than predicted.

*Thank you for this comment. The main thrust of the paper is to draw the readers attention to the anaesthetic complexities posed by a patient with Morquio syndrome who presents for surgery. We include in the paper previously published reports on the use of neuroaxial and general anaesthesia in this cohort. However, due to the complexity of the airway in Morquio syndrome both from cervical instability and deposition of mucopolysaccharides we used an awake fibroptic technique. To our knowledge this is the first time this technique has been reported in adults with Morquio syndrome.*

2. These patients must be carefully positioned intraoperatively secondary to joint laxity and instability. Therefore, I think it would be more valuable to discuss about the problems related to the orthopaedic surgery,

*Thank you for this comment the positioning of the patient intraoperatively is now included in the text. The patient was positioned on his left lateral side with lateral supports at the pelvis padded with absorbent cotton with gauze.*

Reviewer Two
1. The authors describe their management of a patient with Morquio’s syndrome undergoing orthopedic surgery under general anaesthesia. They should be congratulated for their successful care of this difficult patient. The authors briefly review some of the aesthetic implications of Morquio syndrome. However, their review misses many of the important papers about this form of mucopolysaccharidosis

*Thank you for this comment, papers discussing the anesthetic management are now included.*

“The complexity of anesthesia associated with Morquio syndrome has been previously published, particularly in paediatric literature. In a series reporting on occipito-cervical fusion in 17 patients with Morquio syndrome (age 3-22) intubation under GA is described. Video assisted intubation following gas induction is described in a case series of three pediatric patients undergoing GA for otorhinolaryngology surgery. Furthermore, a case of spinal anaesthesia and a GA with inhalation induction in two children undergoing orthopaedic procedures, and an inhalation induction for a child undergoing stabilisation of cervical spine are described. A specifically designed plaster bed which was used to fix the neck during intubation and surgery has been reported. To avoid cervical cord damage in patients with cervical instability, Walker et al. described manual in-line stabilisation during intubation. Furthermore, 2 cardiac valve surgery cases under GA is described. Nott describes an awake intubation in a patient with unstable neck after spraying the pharynx with topical lignocaine 4% and applying fentanyl and ketamine intravenously. Awake fibroptic intubations have been previously described in two pediatric cases of Morquio syndrome, a 23 month-old-girl and a 9 year old boy however our report highlights the use of fibrooptic anesthesia in adults with Morquio syndrome.”
2. Page 3: Please label the major radiological findings depicted in figures 1A and 1B. The figure legends should also be expanded to include descriptions of these findings.

The important features on the images are now labelled and the legends expanded.

3. Page 4: The authors performed a standard fiberoptic intubation. Does the disease mandate any specific changes in technique compared to a patient with a difficult airway who does not have Morquio’s syndrome?

Due to concerns regarding cervical stability, the head should be assisted to be held in the neutral position. This is now included in the text.

The head was held in the neutral position during laryngoscopy.

4. Page 4: How did the authors assess cervical stability and tracheal integrity?

Thank you for this comment. Cervical stability and tracheal integrity was assessed in so far as is possible by diagnostic imaging. However we have added the following text to the report.

Furthermore, cervical spine instability in these patients is often not confirmed or excluded by adequate radiographic examination and functional clinical testing and cervical spine stability may not be preserved in the deeply anesthetized patient with neuromuscular blockade.

5. Page 4: What information did the authors get from the arterial catheter that influenced their intraoperative management?

Thank you for this comment, this is now included in the text.

An arterial line was inserted into the right radial artery for continuous invasive blood pressure monitoring and blood gas analysis as a means of monitoring the adequacy of ventilation or the development of pulmonary compromise.

6. Page 4: Would the authors recommend neuraxial anesthesia in these patients? Why or why not?

Thank you for this comment. There have been a number of cases of successful use of neuroaxial anaesthesia in Morquio syndrome which are now commented on, the text. However, one must consider the type of surgery and the individual patient factors. In this case the patient had an intramedullary syrinx and this out ruled use of spinal anaesthesia. We have drawn the attention of the reader to the importance of a thorough preoperative evaluation in the conclusion of the case report.

There is limited data on anaesthesia in adults with Morquio syndrome, general anaesthesia has been reported in paediatric and to a lesser extent adult patients. This is the first report to our knowledge in which fiberoptic laryngoscopy in an adult is used to assist intubation. Paramount in the anaesthetic care of such patients is a thorough preoperative evaluation of airway in addition to cardiac, respiratory, neurological function.

We are very grateful to the reviewers for their assessment of the paper, we hope that you will now consider it suitable for publication in BMC Anaesthesiology.

Yours sincerely,

Dr Anne Marie McLaughlin