General anaesthesia in an adult patient with Morquio syndrome with emphasis on airway issues

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ABSTRACT

Patients with Morquio syndrome possess a number of characteristics which may complicate an anaesthetic procedure. The most important is that a deposition of mucopolysaccharides in the soft tissues of the oro-pharynx distorts the airway, making the airway management difficult, while the atlanto-axial instability puts these patients at risk of subluxation and quadriparesis. As the endotracheal intubation in Morquio syndrome patients may be difficult or even impossible, we recommend the technique of awake fiberoptic intubation to be considered. Our approach to awake fiberoptic intubation in an adult patient is described in this case report.

KEY WORDS: Morquio syndrome, mucopolysaccharidosis, neck instability, general anesthesia, fiberoptic intubation

INTRODUCTION

The mucopolysaccharidoses (MPS) are a family of lysosomal storage diseases that result in abnormal accumulation of glycosaminoglycans throughout the body. There is a broad spectrum in severity of expression of the musculoskeletal and neurological manifestations of these diseases, ranging from an early and obvious presentation, with rapid degeneration and death, to a nearly asymptomatic phenotype and normal life expectancy [1]. The Morquio–Brailsford syndrome, known as mucopolysaccharidosis type IV (MPS IV), is listed as a ‘rare disease’ by the Office of Rare Diseases of National Institutes of Health, with a prevalence of 1/200 000-250 000 people. This deficiency leads to accumulation of keratin sulfate and chondroitin-6-sulfate in connective tissue, skeletal system and teeth [2]. Infiltration of the tissues may lead to anaesthetic problems. This includes distortion of upper airway anatomy with a large tongue and dental abnormalities which makes the endotracheal intubation difficult or impossible [3, 4]. Spinal abnormalities are common. Atlantoaxial instability with resultant myelopathy and spastic quadriparesis has been described in both, MPS IV and MPS VI. Cervical fixation at the occiput–C2 level is frequently needed in these patients and in fact profilactic occipito-cevical fusion has been strongly advocated for patients with Morquio syndrome independent of neurological symptoms [1, 5]. Cardiac abnormalities may be present. Aortic regurgitation is the most commonly observed valvular abnormality in patients with Morquio syndrome. However, few reports have been published describing cardiac surgery in patients with mucopolisaccharidosis, including Morquio syndrome, because valve disease becomes usually symptomatic only late in the course of these diseases [5]. MPS IV induced thoracic cage deformity, kyphoscoliosis, may reduce the long volumes and cause ventilation-perfusion mismatch [6]. Unlike other forms of mucopolysaccharidoses, MPS IV patients

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FIGURE 1. An art adapted photo of a typical Morquio syndrome patient by Peter Romberg

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have and retain normal intelligence throughout life [1].
As the patients with mucopolysaccharidosis are being re-
ferred to hospital for surgery more frequently compared to
average population, it is important to gain basic knowledge of
the disease in spite of the low incidence of the syndrome [7].
An (art adapted) photo of a typical Morquio syndrome pa-
tient is attached as Figure 1.

CASE REPORT

A 31 year old man, with Morquio syndrome presented at our
Orthopaedic Department with the pain on the hips, limiting
his walking ability to maximally 10 minutes. Bilateral cox-
arthrosis with deformity of the joints was diagnosed. A unilater-
al, total hip replacement had to be performed on the right side
first and after the period of revalidation, on the left, as well.
His length was 170 cm and weight 79 kg. The patient had
a large head which seemed to be positioned on his tho-
rax because of extremely short neck. Inspection of the
mouth showed a large tongue with a Mallampati score III.
Past history revealed cervical stabilization by the poste-
rior spondylodesis Co-C2. Recent radiological investiga-
tion of cervical spine detected abnormal shape and insuf-
ficient height of vertebral corpora and some degenerative
changes (Figure 2. and 3.) Radiological examination of
lumbar spine showed typical scoliosis and torsion, with low
height and wide corpora. Cardiology report was normal and
the lung function test revealed slight diffusion problems.
He used no other drugs except pain medication (paracetamol
and diclofenac). During preoperative evaluation it became
clear that unsuccessful spinal anaesthesia for a recent minor
procedure precluded use of this technique again. The remain-
ing option was awake (sedated) fiberoptic intubation (AFI).
Every step of AFI was explained to the patient and empha-
sized the importance of his cooperation. Enough time was
planned for the preparations. The patient was given an infu-
sion and connected to standard monitoring. To relieve the
anxiety and to achieve some amnesia, 2 mg of midazolam
was given intravenously. As it may prove very difficult (or
impossible) to accomplish an oral AFI intubation, due to
a narrow space between the large tongue and the pharynx
wall and the sharp angle that has to be made by the scope to-
wards the glottic opening, we prepared the nasal route as well.
A vasoconstrictor, 0.5% xilometazoline nose drops were given
first, followed by 2% lidocaine gel (2 cm ) in each nostril. The
tongue and the pharynx were prepared by spraying 4-6 ml
of 2% lidocaine. After a few minutes it was possible to place
de tong depressor deep enough to anesthetize the back of
the tong and pharynx wall by spraying another 4-6 ml of 2%
lidocaine spray, asking the patient to take a deep breath si-
multaneously. We avoid higher concentration of local anes-

FIGURE 2. RTG photo of dens, preoperative.

FIGURE 3. RTG photo of cervical spine, preoperative.
The most important problems concerning the airway management are based on intracellular accumulation of mucopolysaccharides resulting in macroglossia, limited mouth opening due to involvement of temporo-mandibular joints, short neck and subglotic narrowing. These elements may lead to "cannot intubate -cannot ventilate situation" [2]. Oral fiberoptic technique might be difficult due to large, heavy, anteriorly placed, "hanging epiglotis" (sometimes resting on the tong base) making the passage of the fibrescope towards the glotic opening impossible. Nasal fiberoptic technique is straight forward, but might be difficult related to mucopolisaccharoide infiltration in the nasopharynx, with the increased risk of bleeding. Careful preoperative airway examination should be done to predict and evaluate the possible problems. When the airway risk is present, loco-regional technique should be applied if possible. However, the use of spinal anesthesia can be difficult due to progressive scoliosis. Also the optimal dose of local anesthetic is difficult to predict due to (heavy) growth retardation in some of these patients. Regardless of the technique chosen, all necessary equipment to deal with difficult airway should be readily available in case of complications or failure. There is also a controversy about the appropriate means of induction. Some anaesthesiologists prefer the use of intravenous and others of inhalational agents (especially in children), but most of the authors suggest to avoid the use of muscle-relaxants. The relaxation of the supraglottic tissue may preclude effective bag-mask ventilation [3]. Endo-tracheal intubation using fiberoptic technique, being minimally invasive and allowing neutral head position is strongly recommended in these patients [4]. Attention to maximize the respiratory function postoperative is essential to avoid the obstruction and postoperative respiratory failure.

CONCLUSION

In conclusion, we recommend the technique of awake, fiberoptic intubation to be considered in adult patients with Morquio syndrome. Oral fibreoptic approach, as less traumatic, should be the first choice, keeping in mind that nasal route might be needed, and should be prepared as well. We strongly recommend not to use muscle relaxants, as they may impair the efficacy of a bag-mask ventilation, leading to can-not -intubate can-not-ventilate situation.

DECLARATION OF INTERESTS

There are no competing interests amongst the authors and there is not going to be any forthcoming presentation.
REFERENCES


