Cardiothoracic Anesthesia, Respiration and Airway

Tracheal intubation in children with Morquio syndrome using the angulated video-intubation laryngoscope

[L'intubation endotrachéale chez des enfants, atteints de la maladie de Morquio, à l'aide du laryngoscope angulaire pour intubation vidéo]

Alexander Dullenkopf MD,* David Holzmann MD,† Rita Feurer RN,* Andreas Gerber MD,* Markus Weiss MD*

Purpose: There are a number of syndromes with proven or suspected instability of the cervical spine especially in pediatric patients. It is a challenge for the anesthesiologist to intubate these patients with as little movement of the cervical spine as possible. A new device to facilitate this task is the angulated video-intubation laryngoscope (AVIL).

Clinical features: The AVIL is a curved endoscopic intubation laryngoscope with angulated distal tip. The video-view from the distal blade tip improves glottic visualization during difficult direct laryngoscopy. We report three sisters with Morquio syndrome scheduled for otorhinolaryngology surgery the same day. Two of them had radiologically suspected cervical spine instability. Tracheal intubation was planned with careful direct laryngoscopy under manual in-line stabilization of the neck and head by an assistant. Direct visualization of the larynx using a Miller blade No. 2 was impaired in two of the three children in whom the cervical spine was immobilized. They were both successfully intubated under endoscopic control using the AVIL.

Conclusion: The AVIL may become a helpful device to aid endotracheal intubation in patients when cervical spine immobilization impairs direct laryngoscopy.

bougeant la colonne cervicale le moins possible. Un nouveau dispositif, le laryngoscope angulaire pour intubation vidéo (LAIV), peut faciliter cette manœuvre.

Éléments cliniques : Le LAIV est un laryngoscope d'intubation endoscopique courbé dont la pointe distale est angulée. L'image vidéo provenant de cette pointe de la lame permet de mieux visualiser la glotte pendant la laryngoscopie directe. Nous présentons le cas de trois sœurs atteintes de la maladie de Morquio devant subir une intervention oto-rhino-laryngologique le même jour. L'examen radiologique de deux patientes laissait présager une instabilité de la colonne cervicale. L'intubation endotrachéale, planifiée avec une laryngoscopie directe prudente, a été réalisée sous stabilisation manuelle synchronisée du cou et de la tête par un assistant. La visualisation laryngoscopique directe, réalisée avec une lame Miller no 2, a été difficile chez deux des enfants dont la colonne cervicale avait été immobilisée. L'intubation des deux enfants a été réussie sous contrôle endoscopique en utilisant le LAIV.

Conclusion : Le LAIV peut devenir un instrument utile à l'intubation endotrachéale chez des patients dont l'immobilisation de la colonne cervicale affecte la laryngoscopie directe.

Objectif : De nombreux syndromes comportent une instabilité reconnue ou présumée de la colonne cervicale, surtout chez les enfants. C'est un défi pour l'anesthésiologiste de procéder à l'intubation en

MORQUIO'S syndrome (mucopolysaccharidosis Type IVa-c) is an uncommon autosomal recessive disorder. It was described independently in 1929 by

From the Department of Anaesthesia,* University Children's Hospital, and the Department of Otorhinolaryngology,† University Hospital, Zurich, Switzerland.

Address correspondence to: Dr. Markus Weiss, Department of Anaesthesia, University Children's Hospital, Steinwiesstrasse 75, 8032 Zurich, Switzerland. Phone: +41 1 266 77 53; Fax: +41 1 266 71 68; E-mail: markus.weiss@kispi.unizh.ch

Disclosure statement: Dr. Weiss is the inventor of the angulated video-intubation laryngoscope. Dr. Weiss neither does maintain any patent rights on the device nor any agreements with the fiberoptic manufacturer.

Accepted for publication August 28, 2001.

Revision accepted November 5, 2001.

Morquio in Uruguay and Brailsford in England.^{1,2} The basic defect is an abnormality in mucopolysaccharide metabolism, which leads to accumulation of keratin sulfate in connective tissues, mainly in cartilage and bone.³

Beside chest wall deformities, children with Morquio's syndrome have hypoplasia of the dens (odontoid process) as a consistent finding, putting them at considerable risk of anterior dislocation of the vertebral axis with resultant spinal cord compression.⁴ This can occur during head positioning for tracheal intubation. Thus, head extension should be avoided by having an assistant hold the head in the neutral position during laryngoscopy.⁵ However, manual in-line stabilization of the head and neck (MAIS) can make direct laryngoscopic intubation difficult.⁶⁻⁸ A new device to facilitate this task is the angulated video-intubation laryngoscope (AVIL).

The AVIL has been originally developed from a cast plastic Macintosh video-laryngoscope blade No. 4⁹ in which the distal blade (3 cm) is angulated to about 25° to give an increased viewing angle of the integrated fibreoptic endoscope, which otherwise is partially obstructed by the distal horizontal flange of the

Macintosh blade (Figure 1). In addition, the vertical flange is flattened to allow to use the device in children. Actually, only one size of cast plastic prototypes is available.

The AVIL with the angulated distal blade tip resembles an activated McCoy blade.¹⁰ In contrast to the McCoy blade, which can improve direct glottic visualization, the AVIL has been particularly designed to give an improved glottic view on a video-monitor during difficult tracheal intubation, similar to the angulated prism laryngoscope.^{6,10,11}

The AVIL is used to elevate the tongue or, if required, to lift up the epiglottis to provide a full view of the vocal cords. Once the cords are visualized on the bedside video screen, the endotracheal tube (ET), styletted and bent to form a hockey stick, is guided under direct vision along the vertical flange of the blade to the distal blade tip and then guided through the vocal cords under monitor control. The AVIL has been used successfully in children aged from 0.25–17.3 yr with simulated difficult intubations by residents and nurse anesthetists not experienced with fibreoptic intubation.⁶

We report the successful use of the AVIL for tracheal intubation in two of three sisters with Morquio's syndrome scheduled for adenotomy the same day in whom MAIS impaired direct laryngoscopy.

Case report

Case 1

The nine-year-old, 16.2 kg, 101 cm, girl presented for re-adenotomy and myringotomy. During two previous anesthetics, tracheal intubation had once been accomplished by direct laryngoscopy and once by fibreoptic bronchoscopy. Pre-anesthetic neutral and extension lateral *x-ray* view revealed hypoplasia of the dens and cervical spine instability. Induction of anesthesia was performed by inhalation of sevoflurane in nitrous oxide and oxygen. Subsequently, the patient's head was placed carefully in a soft head ring and secured in the neutral position with tapes. After establishing venous access, morphine 0.05 mg·kg⁻¹ and mivacurium 0.2 mg·kg⁻¹ were given to facilitate tracheal intubation. Direct laryngoscopy was performed under the MAIS procedure. Best possible direct laryngoscopy using a Miller laryngoscope blade No. 2 revealed only the tip of the epiglottis (Cormack and Lehane¹² Grade 3 view). Adequate external outer laryngeal pressure did not improve laryngeal visibility. The patient was ventilated again by mask. Then, the AVIL was inserted into the oral cavity and the base of the tongue was carefully elevated until the vocal cords became visible on the monitor. A stilette RAE ET

FIGURE 1 The angulated video-intubating laryngoscope consists of a cast plastic laryngoscope with an integrated fibreoptic endoscope (1.8 m long, OD 2.8 mm, 70° viewing angle, VOLPI AG Schlieren, Switzerland). The distal blade tip is angulated at about 25° to provide an unrestricted viewing angle for the fibreoptic endoscope lens, positioned at the site of angulation (arrow). The endoscope carries optic fibres for image transmission and light transmitting fibres for airway illumination. The proximal viewing ocular is attached to a conventional video-endoscope camera and the standard Storz light adaptor is connected to a light source using a light cable.

FIGURE 2 Video-assisted tracheal intubation using the angulated video-intubation laryngoscope in a patient with impaired direct laryngoscopic visibility during manual in-line stabilization of the head and neck (case 2). Left: The angulated video-intubation laryngoscope was inserted into the oral cavity and the base of the tongue carefully elevated until the vocal cords became visible on the monitor. Right: A stiletted RAE endotracheal tube (ID 5.0) with cuff, bent to form a hockey stick, was guided under direct vision along the vertical flange of the laryngoscope blade into the hypopharynx. Finally, it was directed under video-endoscopic control through the vocal cords, into the trachea.

(ID 5.0) with cuff, bent to form a hockey stick, was guided along the vertical flange of the laryngoscope blade to the larynx and finally through the cords under video-endoscopic control. After the tracheal tube had passed the cords, the stylet was removed and the tracheal tube was further advanced. Final tube position was adjusted to place the tracheal cuff below the vocal cords as displayed on the monitor. Intubation was successful within 30 sec and without arterial desaturation.

Case 2

The ten-year-old sister, 17.2 kg, 102 cm, presented for re-adenotomy and myringotomy. Her past history included anesthesia five years ago with uneventful tracheal intubation. Pre-anesthetic *x-ray* of the lateral spine revealed instability of the first and second cervical

vertebrae and hypoplasia of the dens. Anesthesia was induced with sevoflurane in nitrous oxide and oxygen, supplemented by morphine $0.05 \text{ mg}\cdot\text{kg}^{-1} \text{ iv}$. Direct laryngoscopy was performed with MAIS after administration of mivacurium $0.2 \text{ mg}\cdot\text{kg}^{-1} \text{ iv}$. Best direct laryngoscopic view was only a Grade 3. With additional external laryngeal pressure the arytenoids became visible. To avoid forced airway manoeuvres, tracheal intubation was interrupted and the patient ventilated by mask. In a second attempt, the larynx was visualized using the AVIL (Figure 2 left). Tracheal intubation using the video-laryngoscopic view was performed easily by the nurse anesthetist with a stiletted ID 5.0 cuffed RAE ET without oxygen desaturation (Figure 2 right). Tracheal intubation lasted about 45 sec.

Case 3

The third patient was a nine-year-old girl, 16.5 kg, 102 cm, who was scheduled for adenotomy and myringotomy. Pre-anesthetic radiological examination could not exclude atlanto-occipital instability. The patient was intubated uneventfully with conventional laryngoscopy performed with a Miller laryngoscope blade (No. 2) while MAIS was provided by an assistant.

Neutral position of the head was maintained with tapes during the entire surgical procedure in all three patients. After excluding any residual neuromuscular blockade, the patients were extubated while breathing spontaneously and under deep inhalational anesthesia to reduce the risk of straining and uncontrolled movements of the head during emergence. The postoperative course was uneventful in all three sisters.

Discussion

In pediatric patients there are a number of syndromes and diseases requiring special attention with endotracheal intubation either because of anatomical abnormalities or because of known or suspected immobility or instability of the cervical spine.^{13–17} One example is Morquio's syndrome,^{1,2} one of seven mucopolysaccharidoses.⁴ These patients present various degrees of hypoplasia of the odontoid process of the vertebral axis. Vertebral abnormalities like these bear the risk of causing atlanto-axial-subluxation and spinal cord damage by forced flexion and extension of the neck.^{4,14} In addition, there is sometimes generalized soft tissue infiltration and swelling including macroglossia, tonsillar enlargement, thickening of the nasal mucosa and adenoidal hypertrophy, further impairing direct laryngoscopy.¹⁶

The two patients in cases 1 and 2 represent situations in which direct laryngoscopy was made difficult by the MAIS procedure. Our previous experience with the AVIL in children, prompted us to use the AVIL as a standby device for rapid endoscopic intubation assistance if the MAIS procedure should impair visibility of the vocal cords during direct laryngoscopy.⁶ Both patients were rapidly and uneventfully intubated within the following intubation attempt.

Case 3 represents a patient who was intubated successfully without problems under direct laryngoscopy and MAIS. Cervical spine instability in these patients is often not confirmed or excluded by adequate radiographic examination¹⁸ and functional clinical testing. Cervical spine stability may not be preserved in the deeply anesthetized patient with neuromuscular blockade. Thus, these patients require cautious tracheal intubation. Particularly in smaller children, direct laryngeal visibility may be adequate in spite of

cervical spine immobilization manoeuvres. Direct laryngoscopy with MAIS is an often successful and therefore a suitable approach in these circumstances.⁶

In conditions of known or suspected instability of the cervical spine, intubation should be accomplished with as little movement of the cervical spine as possible. Flexible fiberoptic tracheal intubation or another technique that minimizes cervical spine movements should be the standard of care. Most of these pediatric patients are managed in specialized centres by experienced anesthesiologists in well equipped anesthesia units. However, any practicing anesthesiologist is likely to encounter these patients, possibly requiring immediate endotracheal intubation. Although fiberoptic tracheal intubation should now be part of every anesthesiologist's armamentarium, not all anesthesia units and hospitals are equipped with adequately sized pediatric fiberoptic equipment. Direct laryngoscopic intubation under manual in-line neck stabilization of the head and neck and cricoid pressure is an often performed and a well accepted approach in such situations.^{5,19,20} However, direct laryngoscopy can be more difficult under MAIS and forced laryngeal or cricoid pressure can cause movements of the cervical spine during intubation.^{6–8} If difficulties with direct visualization of the vocal cords occur, anesthesiologists often rely on blind tracheal intubation using a styletted ET or a gum-elastic bougie.²⁰ Other possibilities include the Bullard Laryngoscope²¹ and fiberoptic intubation using the laryngeal mask airway,²² among many others.^{23–25}

The AVIL technique combines conventional laryngoscopy and steering of a styletted ET, using the monitor view from the laryngoscope blade tip. Therefore, it is a technique familiar to anesthesiologists. It can be used as a primary intubation technique or in case of emergency since it does not require extensive patient preparation or personnel assistance when urgent endoscopic intubation is required. The use of an endoscopic device after failure of conventional laryngoscopy allows the anesthesiologist to gain information on direct laryngoscopy for subsequent anesthetic procedures. The delicate fiberoptic bronchoscope is used only if needed.

Beside limitations inherent to endoscopic intubation devices such as fogging and soiling of the lens, the AVIL technique is not suitable in patients with reduced mouth opening further restricting manoeuvrability of the styletted ET. The usefulness of the AVIL for both direct laryngoscopy (similar to the McCoy blade) and endoscopic laryngoscopy (if difficulties occur) needs further investigations.

In conclusion, the AVIL described in this case report was used successfully to facilitate tracheal intubation.

bation in two pediatric patients with difficult intubation during manual cervical spine immobilization. The technique is simple and may become a helpful adjunct in the management of these children, particularly for the anesthesiologist not equipped or experienced with pediatric fiberoptic bronchoscopes.

References

- 1 *Morquio L.* Sur une forme de dystrophie osseuse familiale. *Bull Soc Pediat de Paris* 1929; 27: 145.
- 2 *Brailsford JF.* Chondro-osteo-dystrophy. Roentgenographic and clinical features of a child with dislocation of vertebrae. *Amer J Surg* 1929; 7: 404.
- 3 *Mikles M, Stanton RP.* A review of Morquio syndrome. *Am J Orthop* 1997; 26: 533–40.
- 4 *Walker RWM, Darowski M, Morris P, Wraith JE.* Anaesthesia and mucopolysaccharidoses. A review of airway problems in children. *Anaesthesia* 1994; 49: 1078–84.
- 5 *MacIntyre PA, McLeod ADM, Hurley R, Peacock C.* Cervical spine movements during laryngoscopy. Comparison of the Macintosh and McCoy laryngoscope blades. *Anaesthesia* 1999; 54: 413–8.
- 6 *Weiss M, Hartmann K, Fischer JE, Gerber AC.* Use of the angulated video-intubation laryngoscope in children undergoing manual in line neck stabilization. *Br J Anaesth* 2001; 87: 453–8.
- 7 *Heath KJ.* The effect of laryngoscopy of different cervical spine immobilisation techniques. *Anaesthesia* 1994; 49: 843–5.
- 8 *Hastings RH, Wood PR.* Head extension and laryngeal view during laryngoscopy with cervical spine stabilization maneuvers. *Anesthesiology* 1994; 80: 825–31.
- 9 *Weiss M.* The video-intubating laryngoscope. *The Internet Journal of Anesthesiology* 1998; Vol3N1: <http://www.icaap.org/iuicode?81.3.1.11>.
- 10 *McCoy EP, Mirakhur RK.* The levering laryngoscope. *Anaesthesia* 1993; 48: 516–9.
- 11 *Bellhouse CP.* An angulated laryngoscope for routine and difficult tracheal intubation. *Anesthesiology* 1988; 69: 126–9.
- 12 *Cormack RS, Lehane J.* Difficult tracheal intubation in obstetrics. *Anaesthesia* 1984; 39: 1105–11.
- 13 *Lynn A, Sasaki S.* Unusual conditions in paediatric anaesthesia. In: Sumner E, Hatch DJ (Eds). *Paediatric Anaesthesia*, 2nd ed. London: Arnold, 2000: 335–564.
- 14 *Dasen KR.* Atlantoaxial rotatory subluxation after a pediatric tonsillectomy. *Anesth Analg* 1999; 89: 917–19.
- 15 *Herzka A, Sponseller PD, Pyeritz RE.* Atlantoaxial rotatory subluxation in patients with Marfan syndrome. A report of three cases. *Spine* 2000; 25: 524–6.
- 16 *Berkowitz ID, Raja SN, Bender KS, Kopits SE.* Dwarfs: pathophysiology and anesthetic implications. *Anesthesiology* 1990; 73: 739–59.
- 17 *Birkinshaw KJ.* Anaesthesia in a patient with an unstable neck. Morquio's syndrome. *Anaesthesia* 1975; 30: 46–9.
- 18 *Kulkarni MV, Williams JC, Yeakley JW, et al.* Magnetic resonance imaging in the diagnosis of the cranio-cervical manifestations of the mucopolysaccharidoses. *Magn Reson Imaging* 1987; 5: 317–23.
- 19 *McLeod ADM, Calder I.* Spinal cord injury and direct laryngoscopy – the legend lives on (Editorial). *Br J Anaesth* 2000; 84: 705–9.
- 20 *Nolan JP, Wilson ME.* Orotracheal intubation in patients with potential cervical spine injuries. An indication for the gum elastic bougie. *Anaesthesia* 1993; 48: 630–3.
- 21 *Bjoraker DG.* The Bullard intubating laryngoscopes. *Anesthesiology* 1990; 17: 64–70.
- 22 *Walker RWM, Allen DL, Rothera MR.* A fiberoptic intubation technique for children with mucopolysaccharidoses using the laryngeal mask airway. *Paediatr Anaesth* 1997; 7: 421–6.
- 23 *Osses H, Poblete M, Asenjo F.* Laryngeal mask for difficult intubation in children. *Paediatr Anaesth* 1999; 9: 399–401.
- 24 *Rehmann MA, Schreiner MS.* Oral and nasotracheal light wand guided intubation after failed fiberoptic bronchoscopy. *Paediatr Anaesth* 1997; 7: 349–51.
- 25 *Cook-Sather SD, Schreiner MS.* A simple homemade lighted stylet for neonates and infants: a description and case report of its use in an infant with the Pierre Robin anomalad. *Paediatr Anaesth* 1997; 7: 233–5.