orphanaesthesia

Anaesthesia recommendations for

Morquio syndrome

Disease name: Morquio syndrome

ICD 10: E76.219

Synonyms: Morquio-Brailsford syndrome, Mucopolysaccharidosis IV, MPS IV type IVA

Disease summary: Morquio syndrome (MS) or mucopolysaccharidosis (MPS) type IVA is a progressive lysosomal storage disorder with autosomal recessive inheritance. Morquio syndrome refers to only MPS type IVA. MPS type IVB is a different disease caused by a deficiency of lysosomal enzyme beta-D-galactosidase where as Morquio A or MPS type IVA is caused by the deficiency of the lysosomal enzyme N-Acetyl-galacosamine-6-sulphatase. Deficiency of the enzymes N-acetyl-galactosamine-6-sulphatase (16q24.3, type IV A) leads to the accumulation of glycosaminoglycans (GAGs), keratan sulphate and chondroitin-6-sulphate/chondroitin-4-sulphate. The GAGs accumulate excessively in soft tissue, cartilage, and bone causing severe skeletal dysplasia and a distinct phenotype. Skeletal features include dwarfism, pectus carinatum and kyphoscoliosis, hypoplasia of the odontoid process causing atlanto-axial instability and cervical subluxation, multilevel spinal canal stenosis, and deformity of joints. Short neck, limited mouth opening and narrow upper airway related spaces (accumulation of GAG causing deformation of the oropharyngeal and laryngeal structures, macroglossia and narrow nasal cavities) all lead to sleep apnoea or disordered breathing. AGs have a predisposition for the trachea and cornea, especially keratan sulfate (the predominant GAG in patients with MS) which favours the hyaline cartilage of the anterior tracheal rings causing tracheal narrowing and tracheomalacia. Trachea also has a propensity to buckle and twist resulting in ‘kinking’ of trachea when patient’s head and neck is flexed. Therefore patients with MS prefers a position of head and neck extension, also known as ‘look up-the-sky’ position.

Comorbidity is common in MS. Thoracic deformities result in severe restrictive pulmonary disease, reduced alveolar capacity, and recurrent infections. Accumulation of GAG material in the coronary arteries, heart valves, and myocardium leads to cardiomyopathy, myocardial ischaemia, and valve dysfunction. Other comorbidity is neurological due to spinal canal stenosis and cord compression. In general, MS patients are wheelchair-bound as teenagers, but have a normal intelligence. In most cases, life span is limited to the second or third decade of life, pulmonary and cardiac complications being responsible for the early demise.

For MPS IVA, enzyme treatment therapy (weekly IV elosulfase alpha) and or human haematopoietic stem cells transplantation improve the outcome and quality of life of the patients but do not affect airway difficulty nor the progression of cardiac valvular disease. Patients with MS undergo multiple anaesthetics for surgical procedures, the majority of which are orthopaedic, and ear-nose-throat, cardiac and medical imaging studies (young children). Intraoperative neurophysiologic monitoring (somatosensory and motor evoked potentials) is recommended not only for spine surgeries but also for non-spinal surgical procedures that

may be longer than 90 minutes in duration, especially if the patients have a kyphoscoliosis present.
Airway management is challenging, and general anaesthesia and sedation are high-risk procedures involving the risk of major morbidity including death.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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Typical surgery

Fusion and decompression of the cervical spine, correction of kyphoscoliosis with growing rod instrumentation which requires periodic lengthening, lower extremity surgical procedures involving femur, acetabulum and other long bones are some of the commonest orthopaedic procedures. In older patients, cardiac surgery (primarily for valve repair or replacement) and also more recently tracheal resection and anastomosis are to be expected. Access for enzyme infusion, often called a port placement, is another procedure that brings MS patients to the operating rooms (Hack).

Type of anaesthesia

In general, neuraxial anaesthetics are to be avoided in patients with MS, even though a previous report by Theroux et al. (2012) reported 6 successful epidural placement in 8 patients. The later reports of spinal cord injury (irreversible spinal cord ischaemia/infarction) reported in MS patients preclude the use of neuraxial anaesthesia in MPS IVA patients. (Drummond et al 2015., Tong 2012). More specifically, the case reports describe the spinal cord infarction that occurred in a teenager with MS who had an epidural placed for lower extremity procedure, but was found to have sustained a high thoracic level infarction of the spinal cord at the T1-T7 levels. Note that the epidural had been placed in the lumbar level yet the ischaemia/infarction occurred at the vulnerable part of the spinal cord. In the second case report by Tong et al., the same level of spinal cord infarction occurred in a teenage patient with MS who was undergoing cervical spine surgery. Again, notably, the spinal cord infarction occurred in the T1-T7 area even though the surgical site was well away from such site. Given the devastating nature of the paralytic complication in association with neuraxial anaesthetics in MS patients, the current recommendation is to avoid neuraxial anaesthetics.

Providing Sedation for Medical Imaging Studies

Sedation is often provided for radiological procedures for these children, most commonly for Magnetic Resonance Imaging (MRI). Sedation, which often exceeds ‘conscious sedation’, should be considered a high-risk procedure in MS patients. Due to a high degree of known upper airway difficulties including difficulty in intubating the trachea, the provider of the sedation or general anaesthesia should be an anaesthetist with experience in MS patients.

Intraoperative Neuromonitoring

A group of experts published detailed guidelines on the pre-operative testing and peri-operative management of patients with skeletal dysplasias, among which MS (White KK,). A care algorithm based on these guidelines and focused on determining when using intraoperative neurophysiologic monitoring (somatosensory and motor evoked potentials) in children with mucopolysaccharidosis undergoing non-spinal surgery has recently been published (Kandil AI). The cornerstones of this algorithm are the degree of kyphoscoliosis and the duration of surgery. As a rule of thumb, intraoperative neurophysiologic monitoring is used when:

- the kyphotic angle is > 60°
- the procedure is expected to last longer than 90 min. and or haemodynamic changes are anticipated

- the kyphotic angle is < 60° and the procedure will last longer than 90 min. and or haemodynamic changes are anticipated.

Its application resulted in significant neurophysiologic changes being observed in 3/21 patients in whom intraoperative neurophysiologic monitoring was used. These changes resulted from surgical positioning or hypotension and were resolved within 5–10 minutes after correcting their cause.

Necessary additional pre-operative testing (beside standard care)

Preoperative Testing

Preoperative testing should include airway examination including a consultation from otolaryngology (ENT) to examine the upper airways. A sleep study to estimate obstructive and central sleep apnoea, pulmonary evaluation to include spirometry (if the patient is old enough to cooperate) and cardiac evaluation (echocardiography) are mandatory. CT or CT/Angiogram of the chest focusing on large airways and blood vessels, is very helpful to assess central airway obstruction caused by buckling and kinking of the trachea. Tracheal narrowing may begin as early as 8 years of age and therefore this aspect of pre-operative evaluation is increasingly being recognised.

In addition to echocardiography, cardiac evaluation may need to include an MRI to evaluate aortic root dilatation especially in older MS patients (>20 years). Bear in mind that silent cardiac abnormalities are common in MS patients with predominantly left-sided valve involvement, especially in children. Reported valvular abnormalities include aortic valve insufficiency, which is the most common valve disease present in MS and is a result of GAG deposits that thicken and deform heart valves. Factor et al. studied the coronary arteries by light and electron microscopy in a 15-year-old boy (post mortem) with MS and showed that intimal sclerosis was a prominent feature of MS. Rarely, deposits in the myocardium itself can cause a decrease in myocardial compliance. Currently, mortality and morbidity from MS continue to be of respiratory and neurological origin. However, the advent of enzyme therapy for MS may increase life expectancy, and other organ failures may become more evident.

Induction of anaesthesia

Parameters to consider when planning the induction of anaesthesia are:

- the severity of airway involvement such as history of obstructive sleep apnoea or snoring, macroglossia, obstructed nostrils
- history of cervical fusion: most patients have cervical spine surgery when they are children.
- prior history of anaesthetic difficulty.

It is prudent to obtain an intravenous line before starting an inhalation induction. Nitrous oxide mixed with oxygen may be used with or without topical skin anaesthesia to obtain analgesia to start an intravenous line. A combination of slow inhalational anaesthesia (sevoflurane) combined with small doses of intravenous agent (propofol, ketamine or similar agents) can be used to progressively deepen the level of anaesthesia while assessing airway patency. Dexmedetomidine or ketamine could be used, too. Whether to maintain spontaneous ventilation or not is a matter of clinical judgement by the anaesthesiologist and

is based on the criteria that the patient will be ventilable using a face mask and positive pressure ventilation.

Use of muscle relaxants is a step that should take into consideration the risk of losing the airway if ventilation is not possible via mask and if intubation is not achieved in a timely fashion. This is a crucial step in the anaesthetic management and requires careful consideration. There are indeed patients who may be easier to intubate once relaxed using neuromuscular blockers (such as rocuronium). Videolaryngoscopy should be used from the start in order to limit the number of laryngoscopy attempts (Dohrmann).

Using a laryngeal mask airway (LMA) in case of failed intubation is generally possible but is not guaranteed to provide adequate ventilation: it may be an option for a short period of time (e.g., while preparing a fibreoptic scope or waiting to awaken the patient in the event that intubation failed and surgery is cancelled). Placement of LMA will be easier if the tongue is displaced by retracting the tongue forward by a gauze piece or a tongue retractor. If LMA does not provide air entry after placement it is almost always due to the posterior tongue blocking the LMA aperture. It is also possible to intubate via a well positioned LMA.

It is reasonable to consider an awake fibreoptic nasotracheal intubation in adolescence if the patient agrees, or in the event that all other methods of intubation have previously failed. The nostrils should be carefully prepared using decongestants to minimise the chance of bleeding from the nostril and use a smaller endotracheal tube for easier passage. An ENT surgeon should be immediately available in situations where considerable difficulty is anticipated and tracheostomy may be a consideration. Tracheostomy placement is more difficult in MS patients due to tracheal wall abnormalities (malacia, narrowing, kinking) and their very short neck.

Particular preparation for airway management

The most important aspect of the anaesthetic management of the MS patients is airway management. Anticipation of difficulties and preparation are of paramount importance. Difficult mask ventilation is common, and 'two person' technique may be necessary. Airway tools necessary to manage difficult airway should be readily available. Equipment that displaces soft tissue in a definitive and rigid manner is preferable in children with MS. This includes an oral or nasopharyngeal airway, a video laryngoscope (with either a McIntosh or Miller blade, or an angulated blade for indirect laryngoscopy), a laryngeal mask airway (LMA) and an endoscopy mask (used to provide mask ventilation while a fibreoptic intubation is being performed). A fibreoptic bronchoscope should always be available due to its ability to add additional modes of intubation if other quicker modes such as video laryngoscopy fail.

The fibreoptic scope can be used using a LMA as a conduit when video laryngoscopye has failed. It can also be used in association with a videolaryngoscope to guide the tip of the fibreoptic scope toward the glottis opening. Last but not least, the fibreoptic scope can be useful to control the position of the tip of the tracheal tube in case of tracheal narrowing or kinking.

In a patient with very difficult airway, one may fail to visualise the larynx in spite of best efforts. An ENT surgeon should be available to perform a tracheostomy in the event that all else fail and airway control becomes an emergency. Placement of tracheostomy, electively, should only be after considerable discussion with the parents and patient (if old enough) due to difficulty and risk of the surgical technique (short neck, tracheomalacia, narrow trachea, prominent innominate vessels) but also of maintaining a tracheostomy in such patients. In case of multilevel tracheal obstruction, a Montgomery tracheal tube can be inserted into the

tracheostoma to stent the malacic tracheal wall (Soni-Jaiswal). Another important aspect of the Morquio airway is that they have smaller calibre airway and therefore anticipate using smaller endotracheal tubes (2 or 3 sizes smaller) than predicted for age.

Particular preparation for transfusion or administration of blood products

Not a particular concern in MS patients.

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transportation and mobilisation

Transportation of MS patients requires knowledge of the stability of the individual patient's cervical spine. If in doubt, one may assume the possibility of an unstable cervical spine due to a high prevalence and known risk of cervical subluxation (such situation may arise if a MS patient is being transferred from the site of a trauma or motor vehicle accident where the patient's history may not be readily available. Thus all measures should be taken to stabilise cervical spine while transporting MS patients following an incident such as a 'fall' or major trauma).

Interactions of chronic disease and anaesthesia medications

Not a significant reported problem.

Anaesthetic procedure

Concerns related to induction of anaesthesia are related to upper airway management and intubation of trachea. Ventilating with mask may be fraught with obstruction due to macroglossia and GAG deposits in the naso and orophangeal area. A second anaesthesia provider should be present to help. Two person mask ventilation may be necessary; where one individual manages the mask with anterior displacement of the tongue while the second individual applies positive pressure breaths. Displacing the tongue with a piece of gauze (or a ring forceps) may be necessary to relieve oropharyngeal obstruction and adequately oxygenate the patient. When nasal fibreoptic intubation is attempted, bleeding from the nostrils can obliterate the view. Prior application of a vasoconstrictor should thus be considered (i.e. an adrenalin infused piece of gauze). Inline stabilisation is required during airway manipulation in a patient who may have C1-C2 instability. Neurophysiological monitoring may also be provided, if significant concern of spinal cord compromise exists during airway manipulation for intubation.

When intubating the trachea, regardless of the choice of instrument being used, a simple but effective manoeuvre to facilitate visualising the larynx is to displace the tongue anteriorly with a piece of gauze (or a ring forceps). Whatever the airway management strategy used,

optimal head and neck positioning is critical in children with MS: the head and neck should be in a neutral position with a small roll under the shoulders and no headrest in order to align the clavicles with the external auditory meatus and to avoid any flexion of the neck. The airways of patients with MS easily obstruct when the neck is flexed, as demonstrated by flow–volume loop, tracheal tomography, and fibreoptic tracheography (Pritzker), anterior buckling of the posterior tracheal wall during flexion of the head, causes a slit-like narrowing of the tracheal lumen. While the causative mechanism is unknown, it might be attributed to a loss of tensile integrity of the tracheal walls due to a combination of abnormal hyaline cartilage composition and GAG deposits in the sub mucosal tissue. Other structural and morphological airway abnormalities result from sub-mucosal GAG deposits in the upper airways (tongue, floor of mouth, epiglottis, ary-epiglottis folds, and tracheal wall), all of which combined impart a rigid anatomy. Although rare, vocal cord paralysis has also been described in children with MS.

Based on their study and experience with perioperative anaesthetic care of patients with MS, Theroux et al. recommend the following: (1) airway evaluation including CTA in patients older than 8 years; (2) otolaryngology consult to document abnormalities of airway and pulmonary consult to evaluate respiratory function; (3) videolaryngoscopy to intubate trachea when difficulty with direct laryngoscopy is anticipated; (4) awareness that some patients with MS will be very difficult to intubate, regardless of choice of equipment; (5) evoked potentials monitoring during intubation in the sub-population where spinal cord compromise is a concern; (6) manual displacement of the tongue anteriorly during intubation attempts; (7) expectation of difficulty with nasal FB because of narrow nasopharyngeal path; (8) cardiac consult to evaluate structural and functional cardiac abnormalities.

Particular or additional monitoring

Monitoring spinal cord integrity during spinal fusion procedures and non-spinal procedures (in case of severe kyphoscoliosis or procedures with expected significant haemodynamic changes) using somato sensory (SSEP) and motor evoked potentials (MEP) is recommended, because spinal cord compromise is an ever present threat to MS patients during anaesthesia. There are reports of ischaemic spinal cord injury not adjacent to the surgical site in MS children (see neuromonitoring).

Possible complications

Complications related to airway management:

- Failure to adequately ventilate and oxygenate the patient, hypoxaemic cardiac arrest
- Failure to intubate trachea resulting in cancellation of the surgery or worse hypoxic injury to the patient
- Trauma to the larynx and trachea from repeated and traumatic attempts to secure airway
- Regional neuraxial anaesthesia has been described but may lead to late recognition of spinal cord injury; currently not recommended
- Profuse epistaxis from attempts at nasal intubation
- Cervical flexion causing airway obstruction or spinal cord injury

- Airway obstruction following extubation of trachea.

Complications related to cardiac anomalies

- Myocardial ischaemia
- Heart failure.

Complications related to spinal canal multilevel stenosis

- Postoperative paraplegia.

Post-operative care

Extubation of the patient requires careful consideration, especially following a difficult intubation and long/extensive surgical procedures. If intubation was difficult prior to cervical fusion, greater difficulty should be anticipated following cervical spine fusion. An awake patient able to obey commands (take a deep breath; put your tongue out) is re-assuring when preparing for extubation. Watch patient's respiratory pattern to ensure that no diaphragmatic or other respiratory muscle weakness has ensued from the surgical manipulation of the spine during surgery. All difficult airway equipment should be readily available along with the same skilled personnel who were present during intubation. Extubation over a tube exchanger is recommended in select cases. An otolaryngologist present at induction of anaesthesia and extubation of the trachea is desirable; however, this practice varies between institutions and countries. If the anaesthesiologist chooses to transfer the patient to intensive care with endotracheal tube in place to be extubated at a later period, such extubation should receive the same cautious considerations as described above.

Disease-related acute problems and effect on anaesthesia and recovery

Not reported.

Ambulatory anaesthesia

In MS patients, there are no minor anaesthetics even when surgery may be "minor". There are case reports of airway difficulties during minor surgery such as myringotomy and tube placement. The airway difficulties usually progress and worsen with age along with progression of cardiac and respiratory mechanics. Utmost caution should always be exercised during all anaesthetics including ambulatory anaesthesia and the skilled anaesthesia providers experienced in the care of MS patients should provide care. In the United States, care of MS patients in ambulatory centres is not recommended.

Obstetrical anaesthesia

There are no publications specifically addressing the anaesthetic care of MS patients during labour and delivery. As the life expectancy of MS patients increases, such reports may be forthcoming. Regarding epidural anaesthesia, one should bear in mind that neuraxial spinal stenosis is to be expected especially in older patients. In addition, we no longer think epidural anaesthesia is appropriate for MS patients. For explanation, please see "types of

anaesthesia". Regarding spinal anaesthesia, scoliosis or prior vertebral instrumentation makes it difficult to perform and the spread of the local anaesthetic solution is unreliable. Moreover, systemic hypotension should be avoided. Successful labour analgesia can probably be achieved with remifentanil PCA, and caesarean section requires general anaesthesia following fibreoptic intubation.

References

1. Averill LW, Kecskemethy HH, Theroux MC, et al. Tracheal narrowing in children and adults with mucopolysaccharidosis type IVA: evaluation with computed tomography angiography. Pediatr Radiol 2021;51:1202–1213
2. Aziz MF, Healy D, Kheterpal S, et al. Routine clinical practice effectiveness of the Glidescope in difficult airway management: An analysis of 2,004 Glidescope intubations, complications, and failures from two institutions. Anesth 2011;114: 34–41
3. Dohrmann T, Muschol NM, Sehner S, Punke MA, Haas SA et al. Airway management and perioperative adverse events in children with mucopolysaccharidoses and mucolipidoses: a retrospective cohort study. Paediatr Anaesth 2020;30:181–190
4. Drummond JC, Krane EJ, Tomatsu S, Theroux MC, Lee RR. Paraplegia after epidural-general anesthesia in a Morquio patient with moderate thoracic spinal stenosis. Can J Anesth 2015; 62:45–49
5. Dullenkopf A, Holzmann D, Feurer R, et al. Tracheal intubation in children with Morquio syndrome using the angulated video-intubation laryngoscope. Can J Anaesth 2002;49:198–202
6. Hack H, Chapman I, Finucane K, Barber C. Anaesthesia and orphan disease: Tracheal reconstruction in two children with Morquio diseae. Eur J Anaesthesiol 2020;37:132–137
7. Harmatz P, et al. The Morquio A Clinical Assessment Program: Baseline results illustrating progressive, multisystemic clinical impairments in Morquio A subjects. Mol Genet Metab 2013;109:54–61
8. Hendriksz CJ, et al. Review of clinical presentation and diagnosis of mucopolysaccharidosis IVA. Mol Genet Metab 2013;110:54–64
9. Hendriksz CJ, Berger KI, Giugliani R, Harmatz P, Kampmann C, Mackenzie WG, Raiman J, Villareal MS, Savarirayan R. International guidelines for the management and treatment of Morquio A syndrome. Am J Med Genet A 2015;167A:11–25
10. John RM, Hunter D, Swanton RH. Echocardiographic abnormalities in type IV mucopolysaccharidosis. Arch Dis Child 1990;65:746–749
11. Kandil AI, Pettit CS, Berry LN, Busso VO, Careskey M et al. Tertiary pediatric academic institution's experience with intraoperative neuromonitoring for nonspinal surgery in children with mucopolysaccharidosis, based on a novel evidence-based care algorithm. Anesth Analg 2020;130:1678–1684
12. Karsli C, Armstrong J, John J. A comparison between the GlideScope® Video Laryngoscope and direct laryngoscope in paediatric patients with difficult airways - a pilot study. Anaesthesia 2010;65:353–357
13. Madoff LU, Kordun A, Cravero JP. Airway management in patients with mucopolysaccharidoses: the progression toward difficult intubation. Paediatr Anaesth 2019; 29:620–627
14. Megens JHAM, de Wit M, van Hasselt PM, Boelens J, van der Werff DBM, de Graaff JC. Perioperative complications in patients diagnosed with mucopolysaccharidosis and the impact of enzyme replacement therapy followed by hematopoietic stem cell transplantation at early age. Paediatr Anaesth 2014;24:521–527
15. Mollmann C, et al. Development of a Scoring System to Evaluate the Severity of Craniocervical Spinal Cord Compression in Patients with Mucopolysaccharidosis IVA (Morquio A Syndrome). JIMD Rep 2013
16. Montaño AM, Tomatsu S, Gottesman GS, et al. International Morquio A Registry: Clinical manifestation and natural course of Morquio A disease. J Inherit Metab Dis 2007;30:165–174
17. Morgan KA, Rehman MA, Schwartz RE. Morquio's syndrome and its anaesthetic considerations. Paediatr Anaesth 2002;12:641–644
18. Nielsen, Pedersen, Olsen. Airway management in a patient with Morquio-Brailsford syndrome. Eur J Anaesthesiol 2013;30:133–134
19. Pritzker MR, King RA, Kronenberg RS. Upper airway obstruction during head flexion in Morquio's disease. Am J Med 1980;69:467–470
20. Ransford AO, Crockard HA, Stevens JM, et al. Occipito-atlanto-axial fusion in Morquio-Brailsford syndrome. A ten-year experience. J Bone Joint Surg Br 1996;78:307–313
21. Rodriguez ME, Mackenzie WG, Ditro C, et al. Skeletal dysplasias: evaluation with impulse oscillometry and thoracoabdominal motion analysis. Pediatr Pulmonol 2010;45:679–686

22. Shih SL, Lee YJ, Lin SP, et al. Airway changes in children with mucopolysaccharidoses. Acta Radiol 2002;43:40-43
23. Soni-Jaiswal A, Penney SE, Jones SA, Walker R, Rothera MP, Bruce IA. Montgomery® T-tubes in the management of multilevel airway obstruction in mucopolysaccharidosis. Int J Pediatr Otorhinolaryngol 2014;78:1763–1768
24. Solanki GA, et al. Spinal involvement in mucopolysaccharidosis IVA (Morquio-Brailsford or Morquio A syndrome): Presentation, diagnosis and management. J Inherit Metab Dis 2013; 36:339–355
25. Stevens JM, Kendall BE, Crockard HA, et al. The odontoid process in Morquio-Brailsford's disease. The effects of occipitocervical fusion. J Bone Joint Surg Br 1991;73:851–858
26. Theroux MC, Nerker T, Ditro C, Mackenzie WG. Anaesthetic Care and Perioperative Complications of Children with Morquio Syndrome. Paediatr Anaesth 2012;22:901–907
27. Tomatsu S, Montaño AM, Oikawa H, et al. Mucopolysaccharidosis type IVA (Morquio A disease): Clinical review and current treatment. Curr Pharm Biotechnol 2011;12:931–945
28. Tong CK, Chen JC, Cochrane DD. Spinal cord infarction remote from maximal compression in a patient with Morquio syndrome. J Neurosurg Pediatrics 2012;9:608–612
29. Walker R, Belani KG, Braunlin EA, Bruce IA, Hack H et al. Anaesthesia and airway management in mucopolysaccharidosis. J Inherit Metab Dis 2013;36:211–219
30. Weiss M, Hartmann K, Fischer JE, et al. Use of angulated video-intubation laryngoscope in children undergoing manual in-line neck stabilization. Br J Anaesth 2001;87:453–458
31. White KK, Bompadre V, Goldberg MJ, Bober MB, Cho T-J et al. Best practices in perioperative management of patients with skeletal dysplasias. Am J Med Genet 2017;173A:2584–2595
32. Yasuda E, et al. Pathogenesis of Morquio A syndrome: An autopsied case reveals systemic storage disorder. Mol Genet Metab 2013;109:301–311.

Date last modified: **August 2022**

This recommendation was prepared by:

Mary Theroux, Paediatric anaesthesiologist, Thomas Jefferson University, Philadelphia, USA
Mary.Theroux@nemours.org

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This recommendation was reviewed by:

Rikke Malte Nielsen, Anaesthesiologist, Glostrup Hospital, University of Copenhagen, Denmark
rikkemaltenielsen@gmail.com

Eugen Mengel, Paediatrician, University Hospital Mainz, Germany
mengel@kinder.klinik.uni-mainz.de

Editorial Review 2022:

Francis Veyckemans, retired Paediatric Anaesthesiologist, Belgium
veyckemansf@gmail.com

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