Best Practices in Peri-operative Management of Patients with Skeletal Dysplasia

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Skeletal dysplasias are a heterogeneous group of inherited disorders of development, growth, and maintenance of the human skeleton.[1] Patients with skeletal dysplasias have an increased risk for peri-operative complications related to the anatomy of their upper airway, abnormalities of tracheal-bronchial morphology and function; deformity of their chest wall; abnormal mobility of their upper cervical spine; and associated issues with general health and body habitus. A panel of 13 multidisciplinary international experts participated in a Delphi process that included a thorough literature review; a list of 22 possible care recommendations; two rounds of anonymous voting; and a face-to-face meeting. Recommendations with more than 80% agreement are included below.

General considerations

The mortality and morbidity risk for a patient with skeletal dysplasia undergoing surgery are greater than for the general population because of disproportionate body habitus and subsequent unreliable body measurements, smaller chest and narrow airway, jaw malformations and limited mouth opening, obesity, difficult intravenous access, and associated affection of non-skeletal organ system in some diseases. Hence, patients with skeletal dysplasia should undergo surgery at facilities that can anticipate and manage life-threatening complications.

I. Preoperative assessment

1. Spinal issues

Patients with skeletal dysplasia are at risk for neural compression at both the central and peripheral levels, most significantly at the cervical spine. Prior to general or regional anesthesia, they should have a comprehensive neurological examination as well as an evaluation of neuroaxial integrity and stability with appropriate imaging including cervical, thoracic, and lumbar spine. Flexion/extension MRI is a useful and safe imaging adjunct for preoperative assessment of patients with skeletal dysplasia in whom there is a concern for cervical spinal cord compromise.[2]

2. Airway issues

Patients with skeletal dysplasia are more likely than the general population to have decreased mobility of the cervical spine, which will contribute to difficulty with securing the airway. They are more likely than the general population to have abnormal upper airway as well as trachea-bronchial morphology and function, which can contribute to increased morbidity and mortality.

Patients with respiratory signs or symptoms or thoracic cage structural abnormalities should have preoperative pulmonary evaluation. Polysomnography should be considered in the

preoperative assessment of patients with skeletal dysplasia.[3] Cardiac evaluation should be considered prior to surgery particularly for adults.[4]

II. Preoperative sedation

Preoperative sedation may be given safely to patients with skeletal dysplasia, but there is no evidence that it will decrease the risk of spinal cord damage if the patient struggles during induction. In the event that severe airway abnormalities are suspected, sedation may be initiated in the operating room itself under full monitoring and control of the airway personnel.

III. Anesthesiologic practice during surgery

1. Airway issues

Abnormal anatomy of the head and neck in patients with skeletal dysplasia may preclude emergency tracheostomy. Therefore, preoperative assessment of the position of the cricothyroid membrane relative to the sternal notch is essential. Equipment for intubation of a difficult airway, such as a video laryngoscope and fiber-optic bronchoscope, must be immediately available in the operating room. Post-operative extubation is preferably performed in the operating room by an anesthesiologist,[5] and if it is to be performed outside of the operating room, qualified personnel and equipment must be immediately available because of the higher risk of life-threatening airway complications following anesthesia.

2. Spinal issues

Anecdotal reports suggest that epidural anesthesia should be used with caution in children with skeletal dysplasia due to risk of neurological injury.[6] In the presence of spinal cord compromise or “spine at risk” findings, neuromonitoring is recommended for all surgical procedures exceeding one hour.

Recommendations

1. The mortality and morbidity risks for patients with skeletal dysplasia undergoing surgery are greater than for the general population.
2. Because of the higher risk of anesthesia complications in patients with skeletal dysplasia, such patients should undergo surgery at facilities that can anticipate and manage life-threatening complications.
3. Patients with skeletal dysplasias should have a comprehensive neurological examination prior to general or regional anesthesia.
4. Patients with skeletal dysplasia should have evaluation of neuroaxial integrity and stability with appropriate imaging, including cervical, thoracic, and lumbar spine, prior to anesthesia.
5. Flexion/extension MRI is a useful and safe imaging adjunct for the preoperative assessment of patients with skeletal dysplasia in whom there is a concern for cervical spinal cord compromise.

6. Patients with skeletal dysplasia are more likely than the general population to have decreased mobility of the cervical spine, which will contribute to difficulty with securing the airway.
7. Patients with skeletal dysplasia are more likely than the general population to have abnormal upper airway morphology and function, which can contribute to increased morbidity and mortality.
8. Patients with skeletal dysplasia are more likely than the general population to have abnormal tracheal bronchial morphology and function that contributes to increased morbidity and mortality.
9. Patients with skeletal dysplasia with respiratory signs or symptoms or thoracic cage structural abnormalities should have a preoperative pulmonary evaluation.
10. Polysomnography should be considered in the preoperative assessment of patients with skeletal dysplasia.
11. Cardiac evaluation should be considered prior to surgery for patients with skeletal dysplasia, particularly for adults.
12. Preoperative sedation can be given safely to patients with skeletal dysplasia.
13. Preoperative sedation is NOT indicated in patients with skeletal dysplasia to reduce risk of spinal cord damage if the patient struggles during induction.
14. Abnormal anatomy of the head and neck in patients with skeletal dysplasia may preclude emergency tracheostomy. Therefore, preoperative assessment of the position of the cricothyroid membrane is essential.
15. For patients with skeletal dysplasia, equipment for intubation of a difficult airway, such as a video laryngoscope and fiber-optic bronchoscope, must be immediately available in the operating room.
16. Post-operative extubation of a skeletal dysplasia patient is preferably performed in the operating room by an anesthesiologist.
17. Because of the higher risk of life-threatening airway complications following anesthesia in patients with skeletal dysplasia, if extubation is to be performed outside of the operating room, qualified personnel and equipment must be immediately available.
18. Anecdotal reports suggest that epidural anesthesia be used with caution in children with skeletal dysplasia due to risk of neurological injury.
19. In patients with skeletal dysplasia and evidence of spinal cord compromise or “spine at risk” findings, neuromonitoring is recommended for all surgical procedures.

References

1. Savarirayan R, Rimoin DL. Skeletal dysplasias. *Adv Pediatr.* 2004;51:209-29.
2. Mackenzie WG, Dhawale AA, Demczko MM, et al. Flexion-extension cervical spine MRI in children with skeletal dysplasia: is it safe and effective? *J Pediatr Orthop.* 2013;33(1):91-8.
3. Theroux MC, Nerker T, Ditro C, Mackenzie WG. Anesthetic care and perioperative complications of children with Morquio syndrome. *Paediatr Anaesth.* 2012;22(9):901-7.
4. Braunlin EA, Harmatz PR, Scarpa M, et al. Cardiac disease in patients with mucopolysaccharidosis: presentation, diagnosis and management. *J Inherit Metab Dis.* 2011;34(6):1183-97.
5. Mashour GA, Stallmer ML, Kheterpal S, Shanks A. Predictors of difficult intubation in patients with cervical spine limitations. *J Neurosurg Anesthesiol.* 2008;20(2):110-5.
6. 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 Anaesth.* 2015;62(1):45-9.