Review Article

Anaesthesia management in craniovertebral junctional anomalies

ABSTRACT

Craniovertebral Junctional (CVJ) anomalies are developmental disorders that affect the skeleton and enclosed neuraxis at the junction of cranium and cervical spine. The high prevalence of airway obstruction and restrictive pulmonary disease in combination with cardiovascular manifestations poses a high anaesthetic risk to these patients. This article provides a discussion of management of anaesthesia in patients with craniovertebral anomalies, the evaluation of risk factors in these patients and their management, including emergency airway issues.

Key words: Airway; anesthesia; anomalies; craniovertebral.

Introduction

The delicate balancing of the skull and its contents on the first and second cervical vertebrae evokes admiration. The arrangement of bones, ligaments, and muscles permits a wide range of movements that include nodding, turning, and tilting the head from side to side, looking up or down. The passage of the medullospinal junction from the relatively large volume of the posterior fossa into the narrow confines of the spinal canal is also catered to marvelously.[12]

Craniovertebral junction (CVJ) anomalies are developmental disorders that affect the skeleton and enclosed neuraxis at the junction of cranium and cervical spine. The clinical syndromes associated with these anomalies are attributable to the following: (i) pressure on the neuraxis by the bony abnormalities, (ii) intrinsic malformations of the nervous system, and (iii) disturbance of the cerebrospinal fluid circulation and blood supply. The bony abnormalities usually encountered in CVJ anomalies are basilar invagination, occipitalization, and congenital atlantoaxial dislocation.[4] Congenital dislocations of the atlas form an interesting category of craniovertebral anomalies.[8] They may produce no pain and at times, no symptoms whatsoever, being discovered accidentally on x-ray films of the upper neck. Atlantoaxial dislocations in patients with the mucopolysaccharidosis (MPS) such as Morquio's disease[2,13] are often overshadowed by the crippling physical deformities in chest and limbs and malfunctioning hearts. Trauma of varying severity can also be the cause.

The patients may present with respiratory dysfunction (restrictive lung disease) owing to compression of the brainstem by odontoid process of the second cervical vertebra affecting the respiratory center and weakening of the muscles of respiration including diaphragm.

When surgical treatment is indicated for the patient with a craniovertebral anomaly, he needs special reassurance as an

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operation in the vicinity of the medullospinal junction, as with an operation in the neighborhood of the brainstem, carries risks to life, consciousness, breathing, limbs, and control over the passage of urine/stools. The situation is especially worrisome when the craniovertebral anomaly has been accidentally discovered, the patient being free from symptoms from it. In countries, such as India, where the poor, deprived, and illiterate abound, the responsibilities of the doctor are even greater.[12]

Five Important Concerns that an Anesthetist Needs to Pay Attention

a. Patient positioning
b. Difficult airway
c. Neurological monitoring
d. Blood loss
e. Associated medical ailments including chronic pulmonary disease and cardiac disease.

Patient positioning

Sitting position

Patient head secured in a three pin head holder. Infiltration of the scalp and periosteum at pin site reduces hypertensive response. Bony prominences are padded. Legs placed in thigh high compression stockings to limit pooling of blood. Elbows supported by pillows or pads to avoid contact with table or stretch on brachial plexus and the legs freed of pressure at the level of common peroneal nerve just distal and lateral to the head of fibula. Maintain an inch space between the chin and chest to prevent cervical cord stretching and obstruction of venous drainage from the face and tongue. Avoid large airway and bite block placements. Avoid excessive neck rotation. Avoid excessive flexion of knees toward the chest to prevent abdominal compression, lower extremity ischemia, and sciatic nerve injury.

Prone position

Patient's head elevated to decrease venous bleeding. Face compression is prevented by keeping the head elevated and shoulders at or above the edge of the operating table. However, surgical field is not as clear as in sitting position; eye compression can produce blindness from retinal artery thrombosis, conjunctival edema. Venous pooling in the lower extremities sufficient to impair venous return and hypotension especially in elderly debilitated patients.

Excessive pressure on the abdomen impedes ventilation, compresses vena cava, and increases epidural venous pressure and bleeding. There may be congestion of face and tongue, and pressure sore on malar prominences owing to horseshoe headrest.[3] Extreme flexion of the neck may cause endobronchial intubation because of short trachea and intraoral kinking of endotracheal tube (ETT); hence, armoured ETTs are preferred. Excessive flexion or extension of head may cause brainstem compression in patients with Arnold–Chiari malformation.

Difficult airway

It is advisable to discuss the anesthesia plan with the team before the start of the procedure, which often results in a Plan A and a backup Plan B.

Mouth opening or temporomandibular joint, the range of neck movement should be measured, as some patients can only open their mouth a few centimeters (which may complicate intubation). In addition, the tissues may be very thickened, stiff, and immobile, especially in patients with MPS I and II.[15] The anesthetist should be surrounded by an experienced team and have access to all equipment and support that may be required (e.g., ENT specialist in case of a difficult airway, and intensive care backup).

Patients with potentially unstable necks require induction of anesthesia with minimal or no neck movement to prevent spinal cord damage.[1] This may complicate conventional direct laryngoscopy. The patient is anesthetized using a short-acting anesthetic, succinylcholine,[8] so that following intubation and positioning, a recovery of the respiratory functions determines that no cord injury has occurred during these procedures. Movements at the atlantoaxial joint may be prevented by placing the cervical collar in a position both during intubation and subsequent positioning and by the use of fibreoptic, awake intubation. During the transoral procedures, intubation is done by a reinforced ETT.[8] An alternative intubation technique is preferred, a video laryngoscope,[14] (angled video intubation laryngoscope); the distal blade has a 25° angle to guarantee a greater visualization or fibreoptic intubation. Good preparation of the nose using a vasoconstrictor is essential to avoid bleeding into the airway (which could make the intubation more difficult). Insertion of an intubating laryngeal mask airway will often improve ventilation and facilitate sufficient time to do a bronchoscopy-guided intubation. Another option is represented by supraglottis devices (I-gel) that allow ventilation and oxygenation during endotracheal intubation attempts.

In a very difficult airway, the Combitube[1,11] is a double lumen tube inserted blindly into the esophagus or trachea. The position of the tube is confirmed by the presence of breath sounds or capnography. By inflating one of the two cuffs present, the lungs may then be ventilated. Problems arise after positioning with definitive securing of a tracheal tube, and again with the protection of the airway from aspiration, although stomach suctioning is possible through the gastric port.

Preparation for extubation should include the use of intraoperative steroids, full reversal of the muscle relaxant and placement of a nasopharyngeal airway to reduce upper airway obstruction after extubation. Extubation should be performed in an area where the patient can be reintubated as necessary and where all essential fibreoptic equipment and specialized personnel are available. In the high-risk patient, ENT support for emergency tracheostomy. A carefully positioned tube changer introduced through the ETT can be used to allow reintubation. This serves as a bridge device when successful extubation is uncertain, however, its use may promote obstruction and airway irritation in an already severely narrowed airway.

Some institutions believe that leaving an ETT in place for a prolonged period may lead to additional postintubation changes in the airway, which will exaggerate existing airway obstruction, significantly limiting the potential success of attempted extubation. Patients are best extubated when the patient is fully awake, breathing adequately and moving deliberately, early after surgery. This allows early assessment of neurological status and reduces airway swelling from intubation. If postoperative intubation is required for several days, fibreoptic bronchoscopy can be used to assess the extent of any swelling of the laryngeal area or obstruction from blood clots or other debris. As before, an adequate respiratory effort, a leak around the ETT and other measures necessary to ensure safe extubation should be followed.

Neurological monitoring

Spinal surgery at any level is associated with a higher risk of spinal cord injury. Therefore, neurophysiological monitoring using somatosensory or motor evoked potentials during surgery is required to monitor spinal cord function. This allows identification of surgery or anesthesia-induced neurophysiological changes, suggesting changes to the perfusion or direct damage to the spinal cord. Early recognition may prevent permanent damage.

Blood loss

Remaining alert throughout the surgery with adequate fluid resuscitation and management of massive blood loss with adequate blood transfusions is very essential to maintaining hemodynamic stability throughout surgery and good recovery. The incidence of pulmonary complications was significantly more in patients who received blood transfusion during the intraoperative period.[10] Blood transfusion is considered to be an independent risk factor for the development of postoperative pulmonary complications (PPCs). Blood transfusion is known to cause immunotolerance and immunosuppression, which in turn predispose to nosocomial and postoperative infections.[6]

Associated medical ailments

Cardiac manifestations are common in all types of MPS it is important to assess cardiac risk before general anesthesia. Results of the cardiac consultation should be communicated to both surgeon and anesthesiologist.[13-15] The cardiac consultation should include assessment of current hemodynamic stability, provide recommendations on the need for additional medications or tests, suggest the level of postoperative care and may even uncover a need to defer the procedure. Preoperative cardiac assessment should identify any cardiac disease that would place the patient in the surgical high-risk category.

An anxious combative patient may have a detrimental rise in intracranial pressure (ICP) during induction. Gaseous induction is an acceptable alternative with a nonirritant volatile anesthetic agent such as sevoflurane. A rapid gaseous induction is preferable to prolonged attempts at cannulation in a distressed and unwell child. Hypercarbia should be avoided as it causes cerebral vasodilatation and may worsen raised ICP especially in combination with laryngoscopy and airway manipulation. Hypotension should be avoided because of the risk of decreasing cerebral perfusion pressure in the face of raised ICP. Invasive blood pressure monitoring may be required to maintain the mean arterial pressure and hence the cerebral perfusion.[5]

Respiratory manifestations include restrictive/obstructive lung disease, obstructive sleep apnea (OSA). The etiology of respiratory dysfunctions in CVJ anomaly patients is multifactorial.[7,9,13] There may be associated weakness and dysfunction of respiratory muscles, including the diaphragm. Direct compression of medulla due to bony anomalies results in lower cranial nerve dysfunction or damage to the respiratory center. Cranial nerve dysfunction may cause poor gag and cough reflex resulting in frequent aspiration and pulmonary infection. Afferent respiratory dysfunction has also been described. At times the respiratory involvement may be concealed.[10]

However, respiratory dysfunction may also result from compression at the craniocervical junction due to atlantoaxial disease. This may result in an acute or progressive respiratory deterioration. Furthermore, lesions at this site have been shown to produce disordered breathing during sleep which may occur in the absence of daytime respiratory symptoms, focal brainstem signs or myelopathy.[10]

Aging can be associated with severe narrowing of the larynx or trachea and often with severe OSA, which poses significant challenges to the anesthetist. Dyspnea in the patient with rheumatoid disease is commonly the result of pulmonary, pleural or cardiac involvement by the disease or its treatment.

Rosomoff *et al*. have described the reduced vital capacity, maximum breathing capacity, and lung compliance, despite the absence of clinical evidence of pulmonary diseases in patients of CVJ anomalies.[13]

Postoperative Care

Postoperative complications of awakening after anesthesia are apnea, bronchospasm, cyanosis, and respiratory failure promoted by abundant oral secretions, thoracic cage stiffness, and heart and lung failure, which is why, particularly in MPS and other high-risk patients, intensive therapy, is necessary for the following 24–48 h. Early extubation, immediately after the procedure, reduces the risk of urgent tracheotomy. Postoperative treatment includes steroid prophylaxis to reduce edema, standard treatment for patients with upper airway obstruction (bi-level positive airway pressure, continuous positive airway pressure) and continuous monitoring of cardiac and respiratory function with physiotherapy for several days after the definitive surgery.

Factors significantly associated with PPCs were American Society of Anesthesiologists grade higher than II, preoperative lower cranial nerves palsy and respiratory involvement, duration of surgery, and intraoperative blood transfusion. The most common cause for reintubation and delayed extubation in their study was pharyngeal swelling in transoral odontoidectomy and occipitocervical fixation. Postoperatively, the tracheal intubation is continued for at least 12–24 h.

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Conflicts of interest
There are no conflicts of interest.

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