orphanaesthesia

Anesthesia recommendations for

Crouzon syndrome

Disease name: Crouzon syndrome

ICD 10: Q75.1

ORPHAcode: 207

Synonyms: Craniofacial dysostosis, first branchial arch syndrome

Disease summary: Crouzon syndrome is a congenital disorder characterized by premature closure (synostosis) of the coronal sutures, and less frequently of the sagittal or lambdoidal skull sutures. This results in a dysmorphic appearance of the skull and face, with a high forehead, flattened occiput and brachycephaly. In addition to the craniosynostosis, affected children may also have abnormal fusion of the bones of the skull base and midface, resulting in maxillary hypoplasia, high arched palate and shallow orbits, causing pronounced exophthalmos [1].

Severity is variable. The combination of a reduced intracranial volume and a growing brain may result in raised intracranial pressure (ICP), optic atrophy, hearing loss, seizures, and rarely, cognitive impairment.

Extended dysostosis of the facial and cervical bones and subsequent soft tissue abnormalities may compromise the upper airway. Obstructive sleep apnea (OSA) is common in Crouzon syndrome. Spine abnormalities may also be present and can reduce cervical mobility.

Crouzon syndrome may also be associated with a patent ductus arteriosus (PDA) and aortic coarctation (AoC). Crouzon, Apert, and Pfeiffer syndrome are among the most recognizable of the syndromic craniosynostoses [2,3].

Crouzon occurs in approximately 1 in 25,000 births and is due to a mutation in the fibroblast growth factor receptor (FGFR) 2 gene on chromosome 10. It may be inherited in an autosomal dominant fashion or occur sporadically as a spontaneous mutation. It has a male-to-female ratio of 3:1.

Diagnosis may be incorrect; if uncertainty exists, the diagnosis should be re-evaluated.

Every patient is unique; individual circumstances must always guide clinical care.

Medicine is in progress; new clinical knowledge may not be yet reflected in this guideline Perhaps new knowledge.

Recommendations are not rules or laws; they provide a framework to support clinical decision-making. Although this recommendation has passed a structured review process, it does not meet the formal criteria of a guideline.

Translations may not always reflect the most recent updates of the English version.

Find more information on the disease, its centers of reference and patient organizations on Orphanet: www.orpha.net

Emergency information

A	**AIRWAY / ANESTHETIC TECHNIQUE**	Anticipate difficult mask ventilation and upper airway obstruction (OSA). Have a supraglottic airway available. Video laryngoscopy is the preferred option. Intubation can be easier than mask ventilation. However, be fully prepared for difficulty! Both general and regional anesthesia are feasible.
B	**BLOOD PRODUCTS (COAGULATION)**	No intrinsic coagulopathy! Major craniofacial surgeries = potential massive blood loss!
C	**CIRCULATION**	Crouzon syndrome can be associated with PDA and aortic coarctation. OSA and hypoventilation can lead to pulmonary hypertension.
D	**DRUGS**	Avoid strong opioids and benzodiazepines in OSA patients. Consider short acting agents and opioid sparing strategies.
E	**EQUIPMENT**	Difficult airway cart: video laryngoscope! Prepare for postoperative ventilation needs.

Typical surgery and procedures

Cranial vault expansion: Infants often require surgery to relieve raised ICP and allow brain growth. Posterior cranial vault expansion (with or without spring-assisted techniques) may be done in early infancy (as early as 3-6 months) if ICP is elevated. Fronto-orbital advancement (FOA) is usually performed in late infancy (app. 9-12 months) to correct skull shape and protect the eyes by enlarging the orbital volume [4].

Midface advancement: Severe midface hypoplasia causing exorbitism or airway obstruction (OSA) is addressed with a Le Fort III osteotomy, often with distraction osteogenesis using a rigid external distraction (RED) frame. This is typically done in childhood (around 5-12 years old, depending on severity).

Tracheostomy and Ear-Nose-Throat (ENT) procedures: Neonates with severe upper airway obstruction (e.g., severe choanal stenosis or micro airway) might need tracheostomy. Adenoidectomy or adenotonsillectomy is commonly performed in early childhood to palliate OSA symptoms.

Orthognathic and cosmetic surgeries: In adolescence or adulthood, patients may have corrective jaw surgery (Le Fort I maxillary advancement, mandibular surgery) or cosmetic revisions.

Multistage approach: Definitive management is staged across childhood.

Neurosurgical shunts: Hydrocephalus occurs in a subset of patients.

Other: For patients with a PDA or AoC, pediatric cardiac surgery may be necessary.

Type of anesthesia

There are no known contraindications to specific anesthetic drugs, and surgery can proceed with regional or general anesthesia [5].

Both inhalational and total intravenous techniques are feasible. In the case of increased intracranial pressure, the use of nitrous oxide is not advised; any further increase in ICP should be avoided.

Avoiding general anesthesia and systemic opioids may be advantageous in patients with airway obstruction (e.g., OSA). There have been reports of successful neuraxial blocks, although the presence of scoliosis may make neuraxial anesthesia difficult.

Be very cautious with deep sedation for procedures in Crouzon patients as sedation can precipitate airway obstruction. Dexmedetomidine has emerged as a valuable alternative in this scenario as it provides sedation and analgesia with minimal respiratory depression. However, data are very limited.

Necessary additional preoperative testing (beside standard care)

Look for indicators of a difficult airway during preoperative anesthetic assessment: Evaluate possibilities for alternative airway devices (e.g., supraglottic airway): assess mouth opening [6].

The personal, surgical, and anesthetic history may help guide anesthetic management or prompt further investigations: description of snoring and/or sleep apnea, history of difficult bag-valve-mask ventilation and any adjuncts used.

Corrective craniofacial surgery must be considered genuine major surgery, and an extended preoperative assessment is advisable (including blood work such as full blood count and coagulation studies, planning for transfusion requirements, postoperative intensive care arrangements, etc.).

In suspected or confirmed PDA or AoC, an echocardiogram and assessment for heart failure should be performed.

Also investigate for any history of seizures and the use of anticonvulsant medication.

Particular preparation for airway management

When anesthetizing a patient (adult or child) with Crouzon syndrome, it is advisable to anticipate and prepare for a difficult airway. In children, airway obstruction may occur early during induction due to the tongue blocking a hypoplastic oral cavity; thus, airway adjuncts such as a nasopharyngeal or oropharyngeal airway or a laryngeal mask may be required. The mandible is usually not affected by the disease, and therefore (direct) laryngoscopy is often feasible. In accordance with current guidelines, video laryngoscopy is the preferred option [7,8].

However, there are special situations where laryngoscopy may be more difficult. Following a series of mid-facial distraction procedures to achieve midface advancement (which enlarges the nasopharyngeal space), difficult laryngoscopy may be encountered – particularly during anesthesia for removal of the distraction device. This can occur even if intubation was uneventful during the device insertion.

The anesthetist should be familiar with how to remove the RED frame quickly in case of an emergency. Removal is accomplished by cutting the horizontal wires and using a screwdriver to remove the remaining screw and vertical central bar. The necessary tools must, of course, be immediately available whenever a patient with a RED frame is anesthetized.

A difficult airway trolley, with equipment such as a range of oropharyngeal and nasopharyngeal airways, laryngeal mask airways, a video laryngoscope, cricothyroid puncture (cricothyrotomy) sets, and a fiberoptic bronchoscope, should be kept ready nearby. Prepare correctly sized airway adjuncts in advance.

In any doubtful case, management of the airway by an awake fiberoptic intubation (planned a priori) is advisable.

Particular preparation for transfusion or administration of blood products

Crouzon syndrome is not associated with a bleeding diathesis, and there is no evidence to suggest any specific issues related to blood product administration. However, craniofacial procedures can be associated with significant blood loss, particularly when surgery is prolonged and in younger, lower-weight children [9]. As such, patients should have a full blood count, coagulation screen, and cross-matched blood available prior to surgery. If possible, optimize hemoglobin levels pre-op.

Employ strategies to reduce blood loss. Measures such as the use of anti-fibrinolytics (e.g., tranexamic acid), surgical adrenaline infiltration, meticulous surgical hemostasis, and cell salvage are effective and recommended to reduce transfusion requirements [10]. Invasive monitoring is indicated for craniofacial surgery, and regular sampling for arterial blood gas analysis or a thromboelastogram (TEG) will help guide fluid and blood product management.

Particular preparation for anticoagulation

There is no evidence of any increased risk of venous thromboembolism in the perioperative period, compared to the normal surgical population.

Particular precautions for positioning, transportation and mobilization

Extra care should be taken to protect the patient's eyes intraoperatively. Exophthalmos may be present and full eyelid closure may not be possible. The use of lubricating eye ointment and carefully applied eye pads is advisable.

Interactions of chronic disease and anesthesia medications

None known.

Anesthetic procedure

Once the airway is secure, anesthesia can proceed with either inhalational or intravenous agents, as appropriate. A multimodal analgesic approach may help to reduce opioid requirements in patients with OSA. At the end of craniofacial surgery, the airway should be re-assessed prior to extubation.

Particular or additional monitoring

The type of surgery will dictate the extent of additional monitoring required. Craniofacial surgery will require large-bore peripheral IV access (or central venous access), invasive arterial blood pressure monitoring, and urinary catheterization. Regular sampling for blood gas analysis or TEG will help guide fluid and blood product management during such cases. For other types of surgery, an arterial line may also be useful to allow monitoring of blood gases and hemodynamics in the recovery period.

Cerebral near-infrared spectroscopy (NIRS) can be used to monitor regional cerebral oxygenation during cranial vault surgeries – a drop in NIRS could indicate low cardiac output or significant venous air embolism (VAE) affecting cerebral perfusion.

Possible complications

Children or adults with Crouzon syndrome and suspected or diagnosed OSA may not tolerate a spontaneous-breathing anesthetic technique. This is due to a rightward shift of the CO_2 response curve under anesthesia, compounded by the respiratory depressant effect of opioids. As such, following securement of the airway, positive-pressure ventilation should be used. Consider the use of shorter-acting agents, such as desflurane, propofol, and remifentanil, to facilitate a reliable return of airway reflexes and spontaneous ventilation at the end of the procedure. Avoid benzodiazepines.

In craniofacial procedures, as with any surgery during which the anesthetist does not have immediate access to the airway, be vigilant for evidence of endobronchial intubation, accidental extubation or disconnection, or blockage of the endotracheal tube by blood or secretions. There are no reports of other disease-specific manifestations intraoperatively.

Vigilance for VAE: In surgeries where the head is elevated and venous sinuses opened (like cranial vault expansions), be vigilant for VAE [11].

Postoperative care

Plan for a controlled, awake extubation in most cases. Consider an airway leakage test. Children who used nasal or face-mask CPAP preoperatively should have it available in the recovery area or intensive care unit (ICU). Plan for post-op ICU or high-dependency monitoring for the youngest, major cases and advanced pathology.

Disease-related acute problems and effect on anesthesia and recovery

There are no known syndrome-specific acute emergencies.

Be vigilant for post-extubation airway obstruction and respiratory compromise. Monitor and prevent secretions, swelling, or positional asphyxia.

Ambulatory anesthesia

Children without significant comorbidity or sleep apnea – for example, those with mild manifestations of the syndrome or those who have had successful corrective craniofacial surgery – and who are undergoing minor procedures may be suitable for day-case surgery. Caution is advised for adults with partially treated or suspected sleep apnea, particularly those who require opioids.

Obstetrical anesthesia

Crouzon syndrome is not known to be associated with complications in pregnancy. There is no contraindication to neuraxial techniques, although scoliosis may pose practical problems for neuraxial block placement. If general anesthesia is required, supraglottic airway edema may impede mask ventilation and laryngoscopy. Special consideration should be given to performing an awake fiberoptic intubation in this subgroup of patients [12].

Breastfeeding and genetics: Not directly an anesthetic issue, but the FGFR2 mutation has 50% chance of passing to child (if mother has it). Ensure pediatrician is aware that baby might have the syndrome for any immediate newborn care differences.

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