orphanaesthesia

Anaesthesia recommendations for patients suffering from

Crouzon syndrome

Disease name: Crouzon syndrome

ICD 10 code: Q75.1

Synonyms: Craniofacial dysostosis, first branchial arch syndrome

Crouzon syndrome is a congenital disorder characterised by premature closure (synostosis) of the coronal sutures, and less frequently sagittal or lambdoidal skull sutures. This results in a dysmorphic appearance of 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. 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 (1). It may be inherited in an autosomal dominant fashion or occur sporadically as a spontaneous mutation. It has a male:female predominance of 3:1. The clinical appearance of Crouzon syndrome may vary significantly, from subtle facial features to severe dysplasia and significant comorbidity.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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

Disease summary

Premature synostosis of cranial sutures can produce a number of effects in the growing child, although the degree of severity is variable. The combination of a reduced intra-cranial capacity and a growing brain may result in raised intracranial pressure (ICP), optic atrophy, deafness, seizures and rarely mental impairment.

Extended dysostosis of facial and cervical bones and subsequent soft tissue abnormalities may comprise the upper airways and obstructive sleep apnoea (OSA) is common in Crouzon Syndrome. Spine abnormalities may be present, can reduce cervical movement and together with nasal and pharyngeal obstructions, a difficult airway scenario has to be anticipated (2;3).

Crouzon syndrome may be associated with a patent ductus arteriosus (PDA) and aortic coarctation (AoC).

Crouzon, Apert and Pfeiffer syndromes are the most recognizable of the syndromic craniosynostoses.

Typical surgery

Corrective craniofacial neurosurgery is typical in children with Crouzon syndrome. If possible, these major procedures are postponed till late infancy when bone growth is more advanced.

In the neonatal period non-craniofacial procedures such as tracheostomy or insertion of a ventriculo-peritoneal shunt are more common. Posterior vault expansion may be carried out in the first 6 months of life nonetheless, to achieve cranial decompression, if necessary.

Fronto-orbital advancement is intended to protect the orbitae from subluxation. Complex hypoplasia of the cranial vault, orbitae and midface may require correction with extended procedures such as a fronto-facial advancement, a so-called Le Fort III osteotomy, and/or distraction osteogenesis with application of a rigid extraction device (RED frame). Again, in case of severe hypoplasia these procedures may take place earlier in life, if indicated.

The mentioned reconstructive procedures will probably only be carried out in tertiary, specialized centres and represent an extensive surgical trauma that requires appropriately advanced anesthesiological and intensive care resources and expertise.

In those cases afflicted with a patent ductus arteriosus or aortic coarctation paediatric cardiosurgical procedures may be indicated.

Naturally, patients may present themselves anywhere later in life for any kind of surgery or may even require sedation for elective diagnostic procedures or minor surgery in cases of mental impairment.

Type of anesthesia

There are no known contraindications to specific anesthetic drugs, and surgery can proceed with regional or general anesthesia. The avoidance of general anesthesia and systemic opiates may be advantageous in patients with airway obstructions (e.g. OSA).There have been reports of successful neuroaxial blockages (5). However, the presence of scoliosis may make neuroaxial anesthesia difficult.

Due to the possibility of a difficult airway and especially an upper airway obstruction (see below) inhalational techniques are not suitable for the induction of anesthesia.

In the case of increased intracranial pressure the use of Nitrous Oxide is not advised. Any further increase in ICP should be avoided.

In well-developed countries most adult patients with Crouzon will have had corrective craniofacial surgery and anesthesia will then most likely not pose any difficulties. In other situations, such as in the developing world, adults may be encountered with untreated severe disease.

Necessary additional diagnostic procedures (preoperative)

During the pre-operative anesthesiological assessment scrutiny towards any indicators of a difficult airway is essential. Possibilities for alternative airway devices such as supraglottic masks and tubes should be carefully assessed (Mouth opening?).

There may be important clues in the personal, surgical or anesthetic history: description of snoring and/or sleep apnoea, difficult bag-valve mask ventilation and adjuncts used. Such knowledge may help guide anesthesiological management or further investigations.

Corrective cranio-facial surgery has to be considered as genuine major surgery and extended preoperative assessment in accordance to local practice guidelines is advisable (blood work including coagulation, transfusion requirements, surveillance in intensive care etc.).

In suspected or diagnosed PDA or AoC an echocardiogram and heart failure assessment should be performed or paediatric cardiology be consulted alternatively.

Investigate into a possible history of seizures, seizure medication etc.

Particular preparation for airway management

When anesthetising adults or children 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, and thus require airway adjuncts such a naso- or oropharyngeal tube or laryngeal mask or tube. The mandibula is *usually* not affected by the disease process and therefore laryngoscopy is *usually* feasible.

However, there are special situations where laryngoscopy may be made more difficult: Following a course of staged distractions to allow mid-facial advancement, which is done to enlarge the nasopharyngeal cavity, difficult laryngoscopy may be encountered particularly for anesthesia to remove the distraction device. This can occur even if intubation had been uneventful for the device insertion (6). Another specific situation to be aware of is anesthetising children who have a RED frame in place: Conventional application of a face mask is impossible during induction, but turning the mask upside down can maybe allow a tight fit and positive pressure ventilation if required. Although laryngoscopy may also be impeded in these cases, a laryngeal mask airway should be possible to place as a back-up device.

The anesthetist should be familiar with how to remove the RED frame quickly in case of an emergency (4), which is done easily by cutting the horizontal wires and using a screwdriver to remove the remaining screw and vertical central bar. The necessary tools need of course to be present.

Prior to induction of anesthesia for any patient with Crouzon, proximity of skilled help is mandatory, and this may include a second experienced anesthetist and an (ear-nose-throat) surgeon. A difficult airway trolley, with equipment such as a range of oropharyngeal, nasopharyngeal and laryngeal mask airways, video laryngoscope, cricothyroid puncture sets and fibre optic bronchoscope should be in close proximity. Prior to induction, prepare correctly sized airway adjuncts. Full monitoring should be instituted, for example as per the Association of Anaesthetists of Great Britain and Ireland guidelines (7).

In any doubtful cases, airway management by an a priori awake fibre optic intubation is advisable.

Particular preparation for transfusion or administration of blood products

Crouzon is not associated with 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 where surgery is prolonged, and in younger and lower weight children (8). As such, patients should have a full blood count, coagulation screen, and cross matched blood available prior to surgery. Pre-operative use of erythropoietin and iron has also been described.

Measures such as use of anti-fibrinolytics, surgical adrenaline infiltration, and cell salvage have all been successful in reducing transfusion requirements (9). Invasive monitoring is indicated for craniofacial surgery, and regular sampling for arterial blood gas analysis or a thrombelastogram (TEG) will help guide fluid and blood product management. Apart from red cells, other products which are likely to be required include fresh frozen plasma, cryoprecipitate and platelets. Liaison with a paediatric haematologist before or during the procedure may also be helpful.

Particular preparation for anticoagulation

There is no evidence of increased risk of venous thromboembolism peri-operatively, compared to the normal surgical population.

Particular precautions for positioning, transport or mobilisation

Extra care should be taken protecting the patients' eyes intraoperatively. Exophthalmos may be present and lid closure may not be easy. Use of lubrication and carefully applied eye pads is advisable. There is no report of increased risk of fracture or skin ulceration; therefore patients should be handled with the same precautions as other surgical patients.

Probable interaction between anesthetic agents and patient's long term medication

None known

Anaesthetic procedure

Once the airway is secure, anesthesia can proceed using volatiles or intravenous agents. Multimodal analgesia, including use of local anesthetic infiltration may help to reduce opiate requirement in patients with OSA. At the end of craniofacial surgery, the airway should be re-assessed prior to awake extubation.

Particular or additional monitoring

The type of surgery will dictate the extent of additional monitoring required. Craniofacial surgery will require large bore or central venous access, invasive blood pressure monitoring and catheterisation. Regular sampling for blood gas analysis or TEG will help guide fluid and blood product management. For other types of surgery an arterial line may also allow monitoring of gas exchange in recovery.

Possible complications

Children or adults with Crouzon and suspected or diagnosed sleep apnoea may not tolerate a spontaneous-breathing anesthetic technique. This is due to right shift of the carbon dioxide response curve which occurs under anesthesia, and compounded by the effect of opiates. As such, following securement of the airway, positive pressure ventilation should be used. Consider also 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. Abstain from benzodiazepines!

In craniofacial procedures, as with any surgery where the anesthetist does not have immediate access to the airway, be vigilant at all times for evidence of endo-bronchial intubation, dislocation, disconnection, or blockage of the endotracheal tube by blood or secretions. There are no reports of other common disease specific manifestations intra-operatively.

Postoperative care

Children with Crouzon are at risk of upper airway obstruction on emergence and in recovery. The combination of reduced conscious level and/or excessive opiates, underlying sleep disordered breathing, and any co-existent soft tissue oedema or secretes can be potentially hazardous. Obstruction post extubation may be relieved by simple airway manoeuvres such as a chin-lift/head-tilt and suction. Insertion of a nasopharyngeal airway, if possible, is also effective. Children who used nasal or facial CPAP pre-operatively should also have it available in recovery or ICU.

Information about emergency-like situations / Differential diagnostics

No reports.

Ambulatory anesthesia

Children without significant comorbidity or sleep apnoea, for example those with mild syndrome characteristics or those who have had successful corrective craniofacial surgery and who are undergoing minor operative procedures may be suitable for day case surgery. Caution is advised for adults with partially treated or suspected sleep apnoea, particularly those who require opioides.

Obstetrical anesthesia

Crouzon syndrome is not known to be associated with complications in pregnancy.

There is no contraindication to neuroaxial techniques, but scoliosis may pose practical problems.

If general anesthesia is required, supraglottic airway oedema may impede mask ventilation and laryngoscopy. Special consideration should be given to awake fibre optic intubation in this subgroup of patients (10).

Literature and internet links

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Online Resources:

Johns Hopkins Pediatric Neurosurgery Information Online:
http://www.hopkinsmedicine.org/neurology_neurosurgery/centers_clinics/pediatric_neurosurgery/conditions/craniosynostosis/

Family and patient support group:
https://www.facebook.com/pages/International-Crouzon-Syndrome-Support-Group/146204398727264

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