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Factsheet 6

Anaesthesia issues in CHARGE syndrome – what are the risks?

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Introduction

Children with CHARGE syndrome often require multiple surgeries throughout their lifetime to manage structural and functional anatomical abnormalities. It is important therefore, to consider any risks caused by anaesthesia. These risks tend to vary based on the individual child's CHARGE syndrome characteristics.

Airway Obstruction

Airway problems are common in CHARGE syndrome. Figure 1 shows the normal anatomy of the airway, as well as associated cranial nerves. Most children with CHARGE syndrome have cranial nerve abnormalities. Pertinent problems are those with the glossopharyngeal and vagus nerves, which are responsible for innervating the pharynx and larynx and allowing swallowing to occur (Blake *et al.* 2008).

Problems dealing with oral secretions can be a result of cranial nerve problems (Blake *et al.* 2009a). A study by Morgan *et al.* (1993) examining 50 patients with CHARGE syndrome found that 86% had upper airway abnormalities, 56% had posterior choanal abnormalities and 42% had retrognathia leading to intubation difficulties.

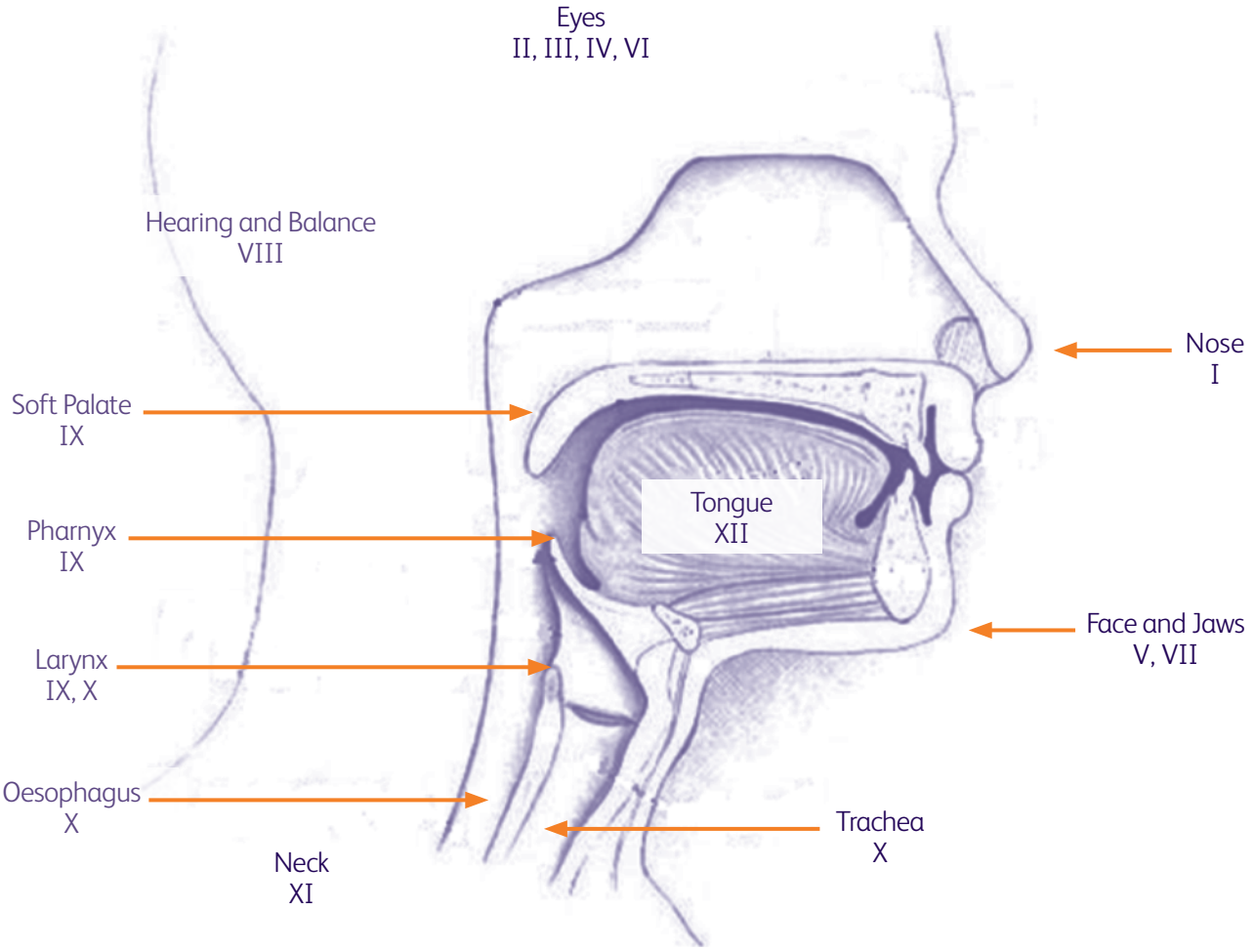
A study done by Naito *et al.* (2007) looked at 10 consecutive patients with CHARGE syndrome who underwent fiberoptic laryngoscopy. All patients had upper airway obstruction at the larynx level, with a hypotonic larynx and hypertrophic arytenoids. They also had uncoordinated movement of the supraglottis, which made it difficult to maintain airway patency. Other problems included glossoptosis, oesophageal fistula and clefts. *See Figure 1, page 2.*

Securing an Airway

There are reports of cancelled surgery and aborted anaesthesia from failure to secure airway control in children with CHARGE syndrome (Hara *et al.* 2009). An early study showed greater difficulty with tracheal intubation that increased with age (Stack and Wyse, 1991). Laryngomalacia may contribute to upper airway collapse during light anaesthesia and may lead to a requirement of CPAP during induction.

Since the pharynx and larynx of CHARGE syndrome patients may be much smaller then expected from their physique, a smaller sized laryngeal mask airway (LMA) should be used first (Hara *et al.* 2009). A wide range of equipment sizes should be on-hand, including that to perform a tracheotomy (Stack and Wyse, 1991). It is suggested that a paediatric

FIGURE 1 ANATOMY OF THE AIRWAY AND ASSOCIATED CRANIAL NERVES



Cranial Nerve		General Function
I	Olfactory	Sense of smell
II	Optic	Sight
III	Oculomotor	Eye movement
IV	Trochlear	Eye movement
V	Trigeminal	Facial sensation and chewing
VI	Abdicems	Eye movement
VII	Facoa	Facial movement and sensation to pharynx and taste
VIII	Vestibulocochlear	Hearing and balance
IX	Glossopharyngeal	Pharynx sensation and movement
X	Vagus	Pharynx and larynx taste sensation and movement, and airway protection
XI	Accessory	Head, neck and shoulder movements and swallowing
XII	Hypoglossal	Movement of tongue, chewing and swallowing

anaesthetist and/or laryngologist be present in the operating room to anticipate possible intubation problems (Blake *et al.* 2009b).

A case study by Hara *et al.* (2009) presented a strategy to secure an airway in children with CHARGE syndrome, beginning with an assessment of the anatomy with a fiberoptic scope. These children should then be intubated with an endotracheal tube under bronchofiberscopic guidance and a ProSeal-type laryngeal mask should be inserted. Atropine is recommended to prevent saliva entering the airway (Hara *et al.* 2009).

Anaesthesia Drug Management

In a chart review study that examined patients with CHARGE Syndrome from a tertiary referral centre (Blake *et al.* 2009b), it was noted that Isoflurane, Halothane and Sevoflurane were the most commonly used inhalation induction agents. Propofol, Ketamine and Thiopentol were the most common hypnotics. Succinylcholine and Rocuronium were the most commonly used muscle relaxants. Fentanyl, Sufentanyl and Morphine were the most commonly used opiods. Experts in the field report the requirement of greater doses of anaesthetic to achieve sedation for these children.

Premedication may be important in cases where secretions are expected to be problematic. Glycopyrrolate can be administered via J-tube to control excessive secretions (Blake *et al.* 2009a). Botox is also effective in controlling secretions when injected into submandibular and parotid glands as a preventative measure (Blake *et al.* 2009a). Atropine has also been recommended (Stack and Wyse, 1991).

Post-op Airway Events

Post-operative airway difficulties are common in children with CHARGE syndrome. In a study by Blake *et al.* (2009b), 35% of anesthetic procedures resulted in post operative airway difficulties. The most common was decreased oxygen saturation requiring intervention. Other problems included obstruction from excessive secretions with desaturations requiring management, prolonged wheezing, heart arrhythmias, decreased respiratory rate, stridor and failed extubation.

Post-operative airway difficulties were most likely to occur following surgical procedures involving the cardiovascular and/or the gastrointestinal systems, or after diagnostic scopes. Given these risks, children with CHARGE syndrome should be monitored longer after surgery than the general paediatric population.

In terms of anaesthetic related risk, there are some protective factors for these children. The same study by Blake *et al.* (2009b) found that individuals with cleft palates had a decreased anaesthetic related risk, which would seem to be due to the widened nasopharyngeal space allowing better drainage of secretions. Also, two of three study participants had fewer airway problems following tonsillectomy and adenoidectomy. This requires further study. Anaesthesia related risks did not increase with the combination of surgical procedures so procedures should be combined for risk reduction.

Suggestions

Expert witnesses note that most of the litigation involving CHARGE syndrome surrounds problems during operations and anaesthesia. *See Figure 2 on page 4 for conclusions and suggestions.*

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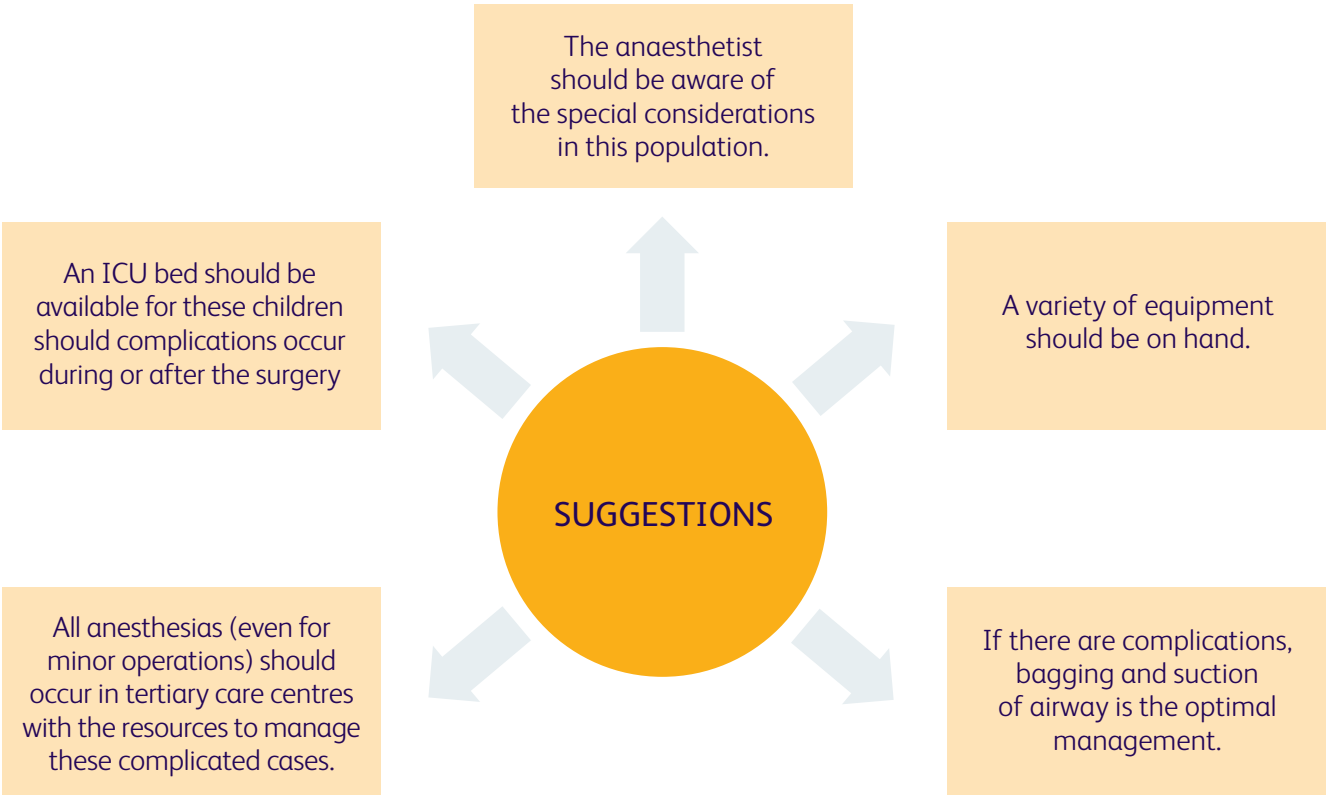
FIGURE 2 CONSIDERATIONS PRIOR TO SURGERY



Combined surgical procedures (i.e. tubes, examination of eyes, dentist, G-Tube change, etc.) should be considered to decrease the number of anaesthesias.

GLOSSARY

Arytenoids: two small pitcher-shaped cartilages at the back of the larynx to which the vocal cords are attached.

Cranial nerves: nerves that emerge directly from the brain rather than the spinal cord.

Choanal anomalies: anomalies affecting the passageway between the nose and the pharynx.

Fistula: a permanent abnormal passageway between two organs in the body or between an organ and the exterior of the body.

Glossoptosis: retraction or downward displacement of the tongue.

Laryngomalacia: a soft floppy larynx, due to softening of the cartilage.

Retrograthia: a condition in which either or both jaws recede with respect to the frontal plane of the forehead.

Supraglottis: the area of the pharynx above the glottis as far as the epiglottis.

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