Try, try, and try again!? Unanticipated difficult airway in a patient with suspected CHARGE syndrome.

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**Goals:**

1. Review CHARGE syndrome and its implications for the anesthesiologist  
2. Discuss perioperative management of a child with a repaired congenital cardiac defect.  
3. Review airway management strategies for the unanticipated difficult airway.  
4. Discuss the decision making process regarding airway management in an elective case.
Case Description:
An 18 month old male with sensoryneuronal hearing loss and possible CHARGE syndrome is scheduled for a right cochlear implant under general anesthesia. The patient was sent for a preoperative anesthetic evaluation because he developed stridor following his last anesthetic which required an ED admission and a course of steroids.

What additional history would you seek before proceeding with the anesthetic? Does the history of stridor change your plans for airway/anesthetic management for this patient? What specific comorbidities are of concern for a patient with CHARGE syndrome? What are the intraoperative anesthetic requirements for cochlear implant placement?

Additional history and physical exam:
The patient is an 18 month old boy likely born full term – his complete birth history is unknown as he is adopted. He had a valve sparing tetralogy of Fallot repair at 3 months of age and has a note from his cardiologist that he is “stable from a cardiac standpoint and fine to have cochlear implant surgery.” He has a history of GER which is controlled with medications (he takes both zantac and prevacid) and reported seizure-like activity following his TOF repair. His father denies any renal, neurologic, hepatic, or endocrine disorders.

In the preoperative holding area, the nurse asks you to evaluate the patient because he has noisy breathing. On exam he is alert and interactive. 9.8kg, HR 88, RR 26, BP 123/91, O2 sat 100% on RA. His breathing is non-labored. There is significant upper airway noise, but his lower airway is clear. He has facial features consistent with CHARGE syndrome. His has slight micrognathia but his airway appears otherwise grossly normal for age.

What are your concerns regarding his previous cardiac history? Would you require additional information? Echo? Any other concerns from his preoperative evaluation?

Induction of Anesthesia:
Standard ASA monitors are placed and the patient is induced with 7L nitrous, 3L O 2 and 8% sevoflurane. The patient is a grade I mask with an oral airway in place. As peripheral access is being obtained in the left foot the O2 sats and HR both fall to the 70s.

What are your initial thoughts for the bradycardia? Hypoxia? What is your first maneuver to amend the situation? If airway does not appear to be the etiology, could this be related to his cardiac status? How do you proceed?

Airway management:
The heart rate and O2 sat saturation improve after 20mg propofol and 200mcg of atropine. The CRNA performs DL and is unable to lift the epiglottis and visualize the vocal cords. Repositioning of the head and changing the blade are ineffective. The view is not improved with anterior manipulation. DL is performed again by a more experienced practitioner who is still unable to visualize the cords. The ENT surgeon who will be performing the cochlear implant requests to take a look.

Would you allow the ENT surgeon to take a look? If not, why not? At what point would you administer steroids? What airway technique would you go to next?

Airway management continued:
While you are waiting for the glidescope, the ENT surgeon repositions the patient with a larger shoulder role and performs DL x2 with the same blades previously used (RS #1 and Mac 1). He is unsuccessful and reports being only able to see posterior arytenoids and the esophagus. The Glidescope provides a grade II view with anterior manipulation but you are unable to make the anterior angle needed to pass the ETT. A fiberoptic scope is used as a “driveable stylet” without success. The airway is becoming slightly more swollen. There is a trace amount of heme.

What next? At what point do you abort the procedure?
ENT tricks:
The ENT surgeon has requested a rigid scope to attempt intubation. He positions the patient with a large shoulder roll and attempts to visualize. The esophagus is easily viewed and he reports a grade II view with the vocal cords being closed (or swollen?) despite deep sevoflurane and propofol anesthesia. Two additional attempts with the rigid scope are unsuccessful. The CRNA reports the patient is more difficult to mask. The O2 saturation is falling and CO2 is not detectable. You have now called for help.

What next? At what point do you consider a surgical airway?

Your ability to ventilate is somewhat improved by removing the shoulder roll. A more senior colleague arrives and requests to perform a DL and in doing so gets a grade I view with a RS 1 blade. You are humbled. The case proceeds without further difficulty from an anesthetic standpoint.

What roll has head position play in the pediatric airway? Is there for or against the use of a shoulder roll? What is your typical practice? What would be your extubation criteria for this case? What do you tell the family regarding future airway management/anesthetics?

Discussion:

This patient presents several challenges for the anesthesiologist related to his presumed diagnosis of CHARGE syndrome and the associated cardiac and airway anomalies. His history of stridor following a previous anesthetic further raises concern regarding potential airway complications for this anesthetic. Airway management for a patient with CHARGE syndrome can be difficult and planning and preparation of resources is essential.

CHARGE syndrome

CHARGE association is characterized by congenital anomalies including coloboma (a hole or gap in one part of the eye), heart defects, choanal atresia, growth retardation, genitourinary problems, and ear abnormalities. Its incidence is approximately 1 in 10,000 to 12,000 live births and its etiology is unknown. Cardiac defects occur in 50-70% of patients with CHARGE and most often include conotruncal (outflow tract) and aortic arch defects. Patients often have growth retardation associated with cardiac disease, nutritional deficiencies and inadequate growth hormone. Visual and hearing defects often predispose developmental delay. Airway difficulty most often exists due to choanal atresia and cleft lip or palate. However, additional airway abnormalities including retrognathia, glossoptosis, tracheolesophageal fistula, laryngomalacia, enlarged tonsils and adenoids, and laryngeal paralysis. Patients with CHARGE syndrome often present for various surgical interventions throughout infancy. Because of their cardiac comorbidities and potential for airway difficulty, these procedures should be combined to limit the number of anesthetics. Airway difficulties tend to be more pronounced as the patient ages, and induction and airway management should weigh the potential complications of airway difficulty with the potential increased risk of aspiration.

Tetralogy of Fallot

Tetralogy of Fallot (TOF) is one of the most common congenital cardiac malformations and consists of (1) pulmonary artery stenosis; (2) interventricular communication; (3) overriding aorta; and (4) right ventricular hypertrophy. This cardiac defect occurs in approximately 3-6 of 10,000 live births and is often associated with 22q11 deletion. Subsequently TOF may occur along with other congenital anomalies including craniofacial and developmental abnormalities. Repair of this defect is now being performed earlier in the neonatal period (<1 year) to minimize secondary insults to the heart and other organ systems. Total correction is the goal of surgical intervention, and consists of VSD closure with either a valve-sparing repair or a transannular patch. Patients are often left with chronic pulmonary regurgitation and potential RV dysfunction and may necessitate future intervention.

Airway

Dysmorphic patients present multiple challenges and more often than not, a major concern is the potential for a difficult airway. In CHARGE syndrome, there are multiple possible etiologies for airway obstruction and difficult intubation. Choanal atresia is a major criteria for the diagnosis of CHARGE syndrome and is a congenital unilateral or bilateral bony
or membranous obstruction of the nasopharynx. Because infants are obligate nasal breathers, this condition may contribute to respiratory distress or asphyxia in the neonate. Alternatively, unilateral or partial choanal atresia or stenosis may initially go undetected. Both complete and partial nasopharyngeal obstruction exacerbates airway obstruction with relaxation of upper airway tone after sedation or induction of general anesthesia.

Micrognathia increases the difficulty of direct laryngoscopy and predisposes the patient, particularly those young in age, to upper airway obstruction. In such a patient, mandibular hypoplasia creates a limited resting spot of the tongue causing a relative macroglossia and increased incidence of obstruction and desaturation.

Optimal head positioning during laryngoscopy is essential to provide optimal visualization of laryngeal structures. In older children and adults, the classic teaching is to raise the occiput with a folded towel or blanket (thus displacing the cervical spine anteriorly) and extend the head at the atlanto-occipital joint to produce the classic sniffing position. These maneuvers align the axis of the mouth, oropharynx and trachea and thus allow best visualization for intubation. In infants and young children, the pronounced occiput raises the cervical spine adequately and head extension alone should provide adequate visualization. Anesthesiologists and otolaryngologists frequent place a rolled towel under the infants’ shoulders to facilitate visualization – this maneuver may be more beneficial for a practitioner in the seating position and might actually limit visualization for a standing practitioner. There is limited anatomic evidence addressing the use of the use of a shoulder roll or other positioning device to optimize airway visualization in the pediatric population.

References:


