Saturday, August 3, 2019

Breakout Session D22 • 4:00-5:00 pm • Chantilly Ballroom East

Airway and Anesthesia Risks for Individuals With CHARGE Syndrome. How Can You Advocate for Best Practice?

Dr. Kim Blake, Dalhousie University

Dr. Catherine Hart, Cincinnati Children's Medical Center

with Jessica MacLean and Emily Chedrawe

Presenter Information

Dr. Kim Blake is a professor of pediatrics at Dalhousie University in Nova Scotia, Canada. She has been researching in CHARGE syndrome over the last 35 years and has published extensively. She has explored post-operative airway events, sleep apnea, bone health, cranial nerve abnormalities, and gastrointestinal issues. In the last 10 years Dr. Blake has partnered with Dr. Jason Berman and they have developed a zebra fish model of CHARGE syndrome to answer further research questions. With this model we have been able to understand about the abnormalities of the vagus nerve and gut mobility in CHARGE syndrome which has influenced our knowledge of gut motility. Anesthesia has also been researched in our zebra fish model. This support the clinical findings that individuals with CHARGE syndrome have increased risk following anesthesia and should have combined procedures where possible in one anesthesia. Kim is very proud of the CHARGE syndrome checklist which has been developed both for families, individuals, and professionals to use as a guide and a teaching tool for anybody dealing with CHARGE syndrome.

Dr. Catherine Hart is a faculty member in the Division of Otolaryngology at Cincinnati Children's Hospital Medical Center. She has fellowship training in the management of pediatric airway disorders and specializes in the management of children with complex aerodigestive disorders. She is a member of the CHARGE Clinic at Cincinnati Children's and cares for multiple children with CHARGE syndrome every

Presentation Abstract

Dr. Blake and Dr. Hart will give this platform presentation, which will help families recognize risks associated with airway problems and anesthesia. Individuals with CHARGE syndrome can have a variety of airway problems that can lead to airway obstruction, obstructive sleep apnea and difficulty managing the airway in an emergency. They also experience both minor and severe adverse advents during and following sedation and general anesthesia. We will review the anatomic considerations and characteristic airway findings in children with CHARGE syndrome and discuss both clinical knowledge and basic science research in this area. This knowledge will empower families and individuals to advocate for best practice and care when interacting with anesthesiologists and surgeons. Key messages will be to combine surgeries and procedures under a single anesthesia whenever possible to minimize the number of anesthesia episodes.

Learning Objectives

- Identify anatomic reasons for airway obstruction in children with CHARGE syndrome.
- Attendees will have knowledge and advocacy tools to help them prepare for future general anesthesia and sedation.
- Research from our CHARGE syndrome zebrafish model will be shared with families and individuals so supporting them in their knowledge base when discussing serious concerns with the anesthesiologist.

Airway and Anesthesia risk for individuals with CHARGE Syndrome.

How can you advocate for best practice?

Dr. Catherine Hart[1] and Dr. Kim Blake[2]
Co-presenters Jessica MacLean[2] and Emily Chedrawe[2]
[1]Cincinnati Children's Hospital Center, University of Cincinnati, Cincinnati, OH, USA
[2]Dalhousie University, IWK Health Centre, Halifax NS.

Objectives

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2. Attendees will have knowledge and advocacy tools to help them prepare for future general anesthesia and sedation.

3. Research from the Dalhousie CHARGE syndrome zebra fish model will be shared to supporting families in their knowledge base when discussing serious concerns with the anesthesiologist.

Anatomic reasons for airway obstruction

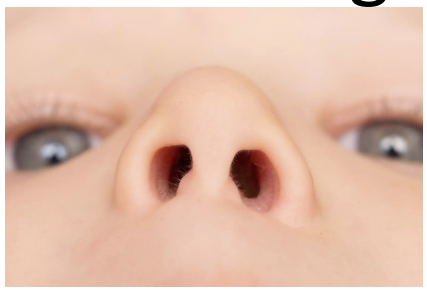
1. Induction of anesthesia
2. Intubation
3. Maintenance of anesthesia
4. Emergence from anesthesia/Post-operative recovery

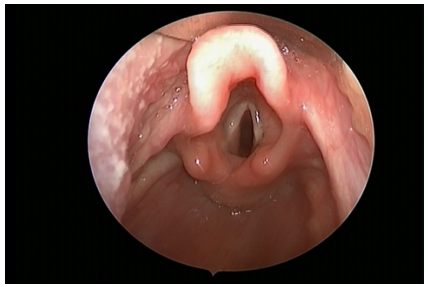
Induction and Excitement—Stages 1 and 2

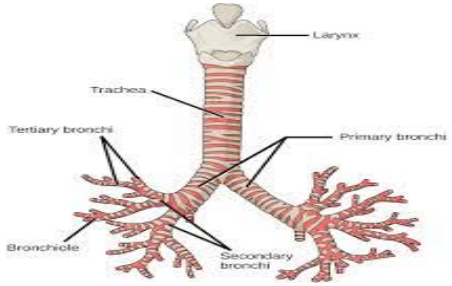
- Stage 1: Begins with administration of anesthesia and lasts until loss of consciousness
- Stage 2: From loss of consciousness until start of automatic breathing
  - Breathing can be irregular with breath holdi
- Issues during these stages:
  - Difficulty ventilating (trouble moving air)
  - Airway obstruction
  - Laryngospasm/bronchospasm

Reasons for difficult ventilating

- Issues can be due to
  - Nose
  - Throat
  - Voicebox (larynx)
  - Airway (trachea/bronchi)

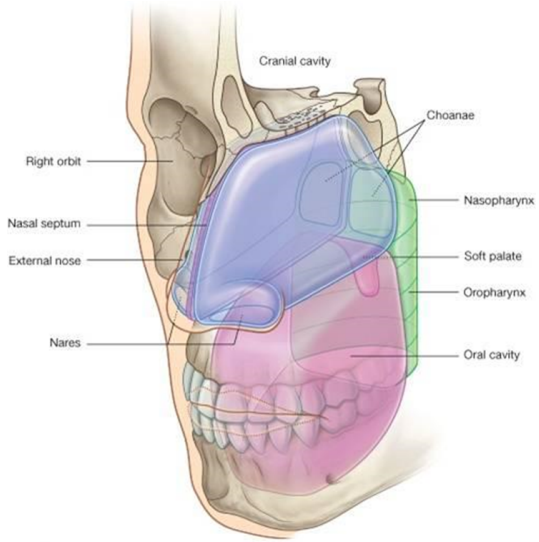






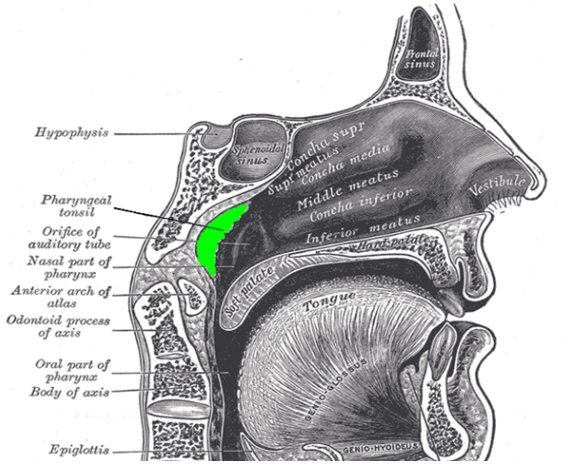
Nose: Choanal atresia

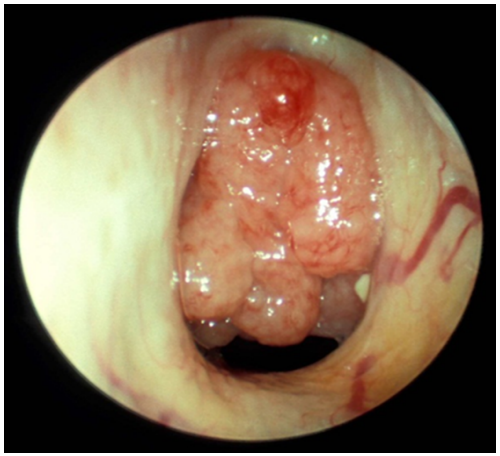
- 50-60% of patients
- Unilateral or bilateral
- Interferes with breathing
- Interferes with ability to eat by mouth



Nose: Adenoid hypertrophy

- Present at birth but usually quite small
- Start to enlarge between 3-6 months
- Continue to grow until approx 6 years





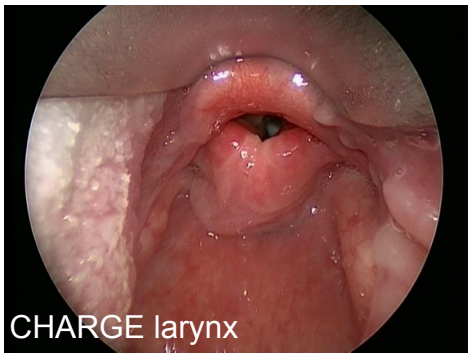
Nose/Throat: Cleft Lip/palate

- Present in 25%
- Difficult to seal the mask
- Tongue can fall back in throat and may block airflow

Voicebox/Larynx

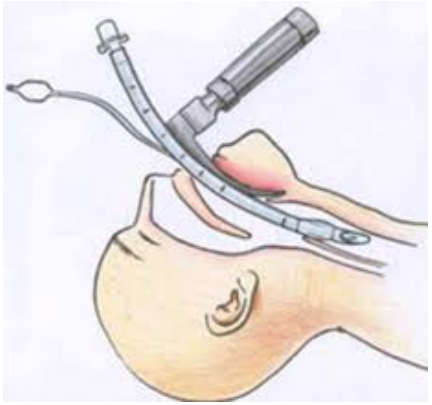
In CHARGE:

- Prominent, anteriorly placed arytenoids
- Shortened aryepiglottic folds
- Foreshortened appearing vocal folds
- May block airflow



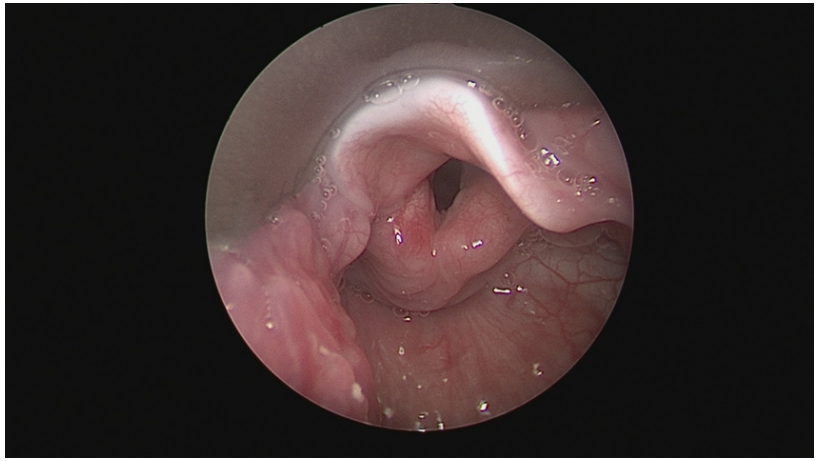
Intubation

- Passing the breathing tube into the airway can be difficult in children with CHARGE



Intubation in CHARGE

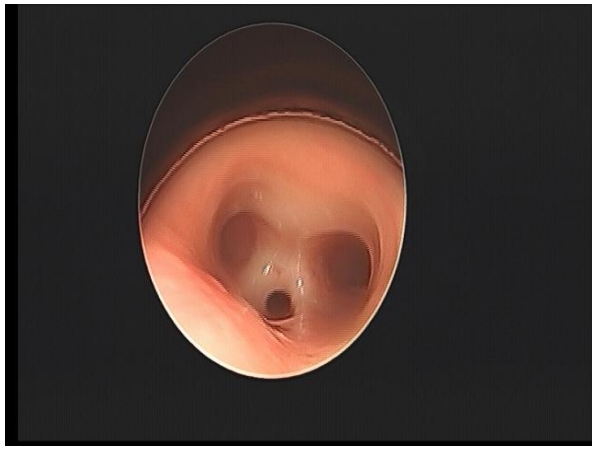
- Can be difficult to get a view of the larynx
- Can be difficult to pass the breathing tube into the larynx due to extra tissue above the voice box
- Or due to narrowing below the voice box



Add video of airway here

Tracheoesophageal Fistula

- Connection between trachea and esophagus
- Present in 15-20%
- Breathing tube can end up in the hole (if not repaired) or the pouch (if repaired)



Difficult Intubation

- If your child is difficult to intubate:
- Make sure new providers are aware
- Schedule elective procedures at experienced center

Maintenance of Anesthesia

- Desaturations (dropping Oxygen levels)
  - Aspiration→damage to lungs making it more difficult to tolerate anesthesia
  - Cardiac Issues
  - Bronchospasm/airway compression

Bronchospasm/compression

- The smaller parts of the airway can collapse preventing good movement of air
- Similar to asthma
- Can be due to bronchomalacia or compression of the small airways

Insert video of bronchial compression

Summary of Anatomic Reasons for Airway Obstruction

- Airway obstruction can occur at all levels of the airway
- Individuals with CHARGE often have multiple levels of airway obstruction
- This increases the risk of anesthesia

There are Always Risks of Complications with Anaesthesia

- "…you sign a consent"
- Are you informed?
- Are Individuals with CHARGE Syndrome More at Risk?

If yes, what are the risks and who should know?

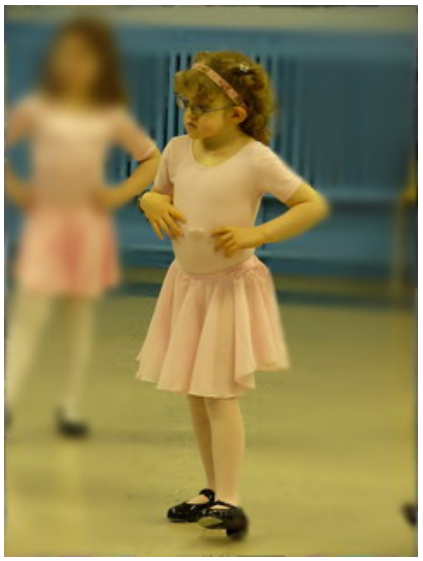
Halifax, NS 2006

Growing up With CHARGE Syndrome and *ICU admissions*

Age 0-2 years: 7 surgeries

- *2 weeks – open heart surgery*
- *6 months – G-tube/fundoplication extubation attempted (x 3)*
- *18 months – aspiration pneumonia*

Age 2-4 years: 3 surgeries



Age 4-6 years: 6 surgeries

- *6 yrs. – heart surgery – pneumonia after heart surgery*

Postoperative Airway Events of Individuals with CHARGE Syndrome

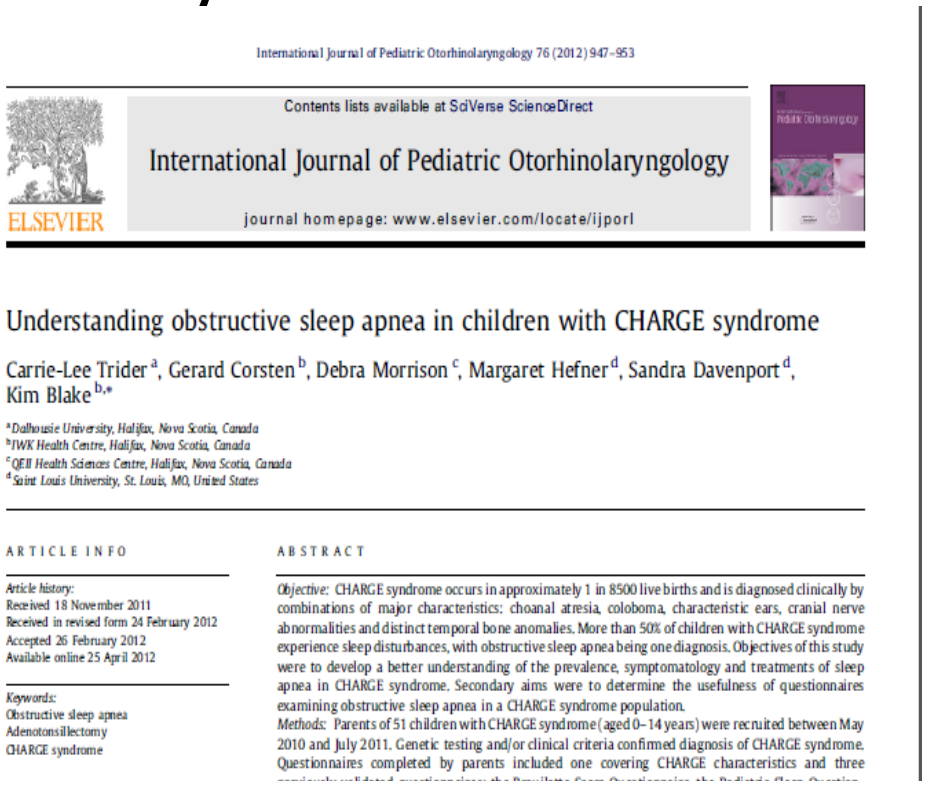
Population n=9

Mean age 11.8 years (± 8.0)

215 surgeries (mean 22 per child)

147 anesthesia's (mean 16 per child)

Postoperative events (e.g. reintubation for apneas and desaturations, airway obstruction due to excessive secretions, failed extubation and decreased respiratory rate needing ICU admission)



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Understanding obstructive sleep apnea in children with CHARGE syndrome

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ABSTRACT

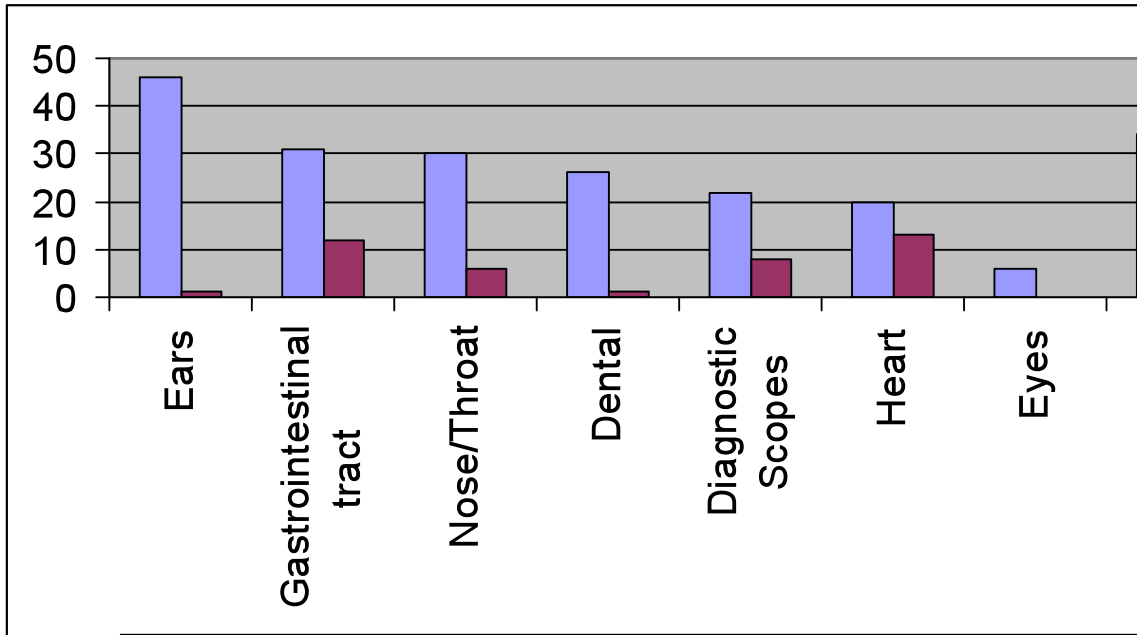
Objective: CHARGE syndrome occurs in approximately 1 in 8500 live births and is diagnosed clinically by combinations of major characteristics: choanal atresia, coloboma, characteristic ears, cranial nerve abnormalities and distinct temporal bone anomalies. More than 50% of children with CHARGE syndrome experience sleep disturbances, with obstructive sleep apnea being one diagnosis. Objectives of this study were to develop a better understanding of the prevalence, symptomatology and treatments of sleep apnea in CHARGE syndrome. Secondary aims were to determine the usefulness of questionnaires examining obstructive sleep apnea in a CHARGE syndrome population.

Methods: Parents of 51 children with CHARGE syndrome (aged 0–14 years) were recruited between May 2010 and July 2011. Genetic testing and/or clinical criteria confirmed diagnosis of CHARGE syndrome. Questionnaires completed by parents included one covering CHARGE characteristics and three

Blake K, et al Int. jnl Ped Otorh 2009

Results

Anaesthesia related events occurred most often with heart, diagnostic scopes, gastrointestinal tract procedures.



Results

- 35% (51/147) of anesthesia's resulted in post-operative events (>60% were major)
- Combining multiple procedures under one anesthesia did not lead to an increase in post-operative events.
- Having a G/J tube or Nissens fundoplication increased the child's risk of post-operative airway events

MacKenzie & Kennedy sharing lunch

Summary

- Post-operative events are common for individuals with CHARGE syndrome cardiovascular, diagnostic scopes, and gastrointestinal tract procedures result in the most events.
- High risk of complications in individuals with Nissen fundoplication and/or gastrostomy/ jejunostomy tube

- Having multiple procedures with one Anaesthetic does not increase adverse events

What about individuals who have mild clinical criteria?

Will they be at risk in the future?

Have they actually been challenged with surgeries?

MacKenzie's Story

- 27 surgical procedures
- 18 anesthesia's
- 4 complications
- Multiple ICU admissions

Following post tonsillectomy and adenoidectomy there were no anesthesia problems.

Botulinum Toxin Injections into Salivary Glands to Decrease Oral Secretions in CHARGE Syndrome: Prospective Case Study. AJMG Part A Med Genet 158A:828-831 Mar 2012

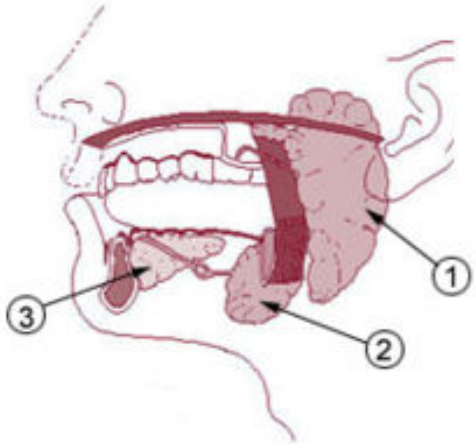
Freddy's Story

Freddy at 2 Months

- Difficulty with intubation
- ToF repair, vascular ring repair, PDA ligation
- Increased oral secretions
- Multiple attempts at extubation

Botox Injection

Submandibular Gland Via Ultrasound and Parotid Gland by Palpation



1. Parotid glands
2. Submandibular glands
3. Sublingual glands

Botox 10 Units/gland

Summary - Botox

- Botox injections into the salivary glands may help reduce oral secretions (needs repeating every 4-5 months.
- Reduction in oral secretions may help prevent aspiration and pneumonia
- May help prevent and/or removal of tracheostomy

The CHARGE Information Pack for Practitioners

sense
for deafblind people

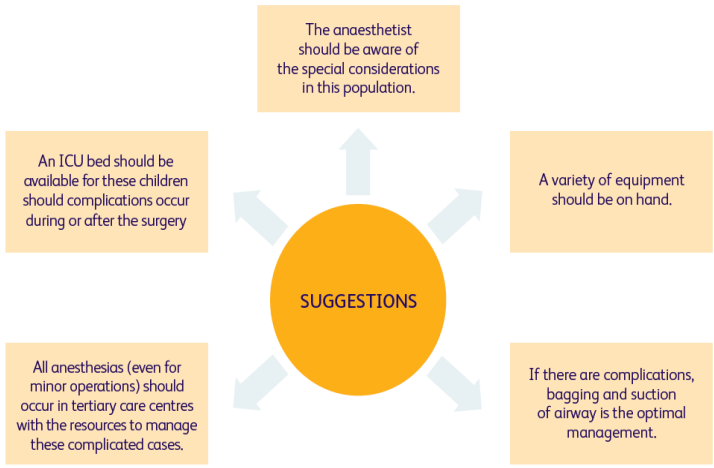
Factsheet 6

Anaesthesia issues in CHARGE syndrome – what are the risks?

CARRIE-LEE TRIDER, MD, Dalhousie University
KIM BLAKE, MD, MCS, MRCP, FRCP(C), Professor Paediatrics, IWK Health Centre, Canada

Anaesthesia Issues

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.

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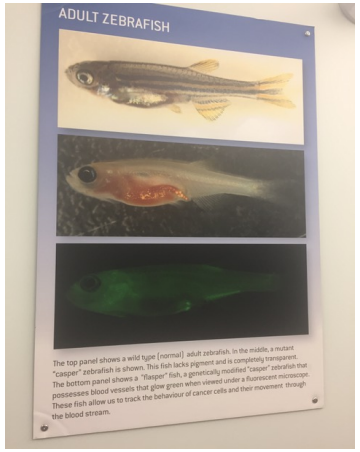
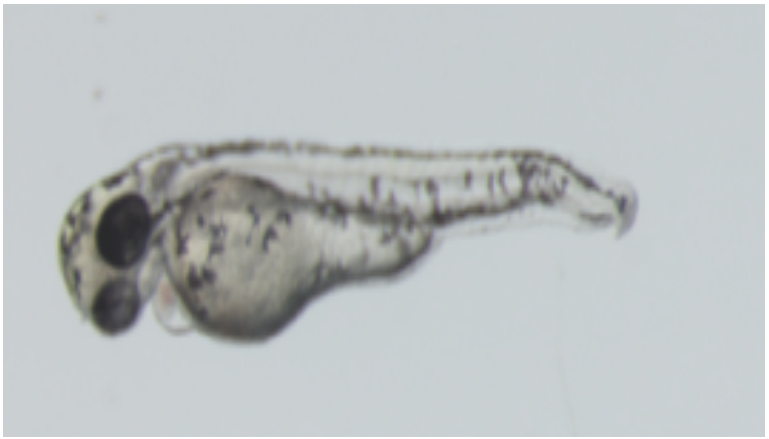
Research from the Dalhousie CHARGE syndrome zebra fish model will be shared to supporting families in their knowledge base when discussing serious concerns with the anesthesiologist.



Response to anesthesia in a zebrafish model of CHARGE syndrome

- Berman lab at Dalhousie University made a zebrafish model of CHARGE syndrome using a technique called CRISPR
- CRISPR acts like a pair of scissors and snips out a specific segment of DNA
- We used it to remove chd7 from these zebrafish



Response to anesthesia in a zebrafish model of CHARGE syndrome

- Individuals with CHARGE syndrome have trouble undergoing anesthesia
- Investigated whether this was the case in our CHARGE zebrafish by looking at:
    1. Time to become anesthetized (lose response to touch)
    2. Time to recover from anesthesia (gain response to touch)
    3. Heart rates under anesthesia
    4. Respiratory rates under anesthesia
- We compared the CHARGE zebrafish to a control fish, with intact chd7

CHARGE zebrafish respond differently to anesthesia

1. CHARGE zebrafish required more time in anesthetic to become anesthetized
    - CHARGE zebrafish took 89 seconds vs 31 seconds in other fish
2. There was no significant difference in time to recover from anesthetic
    - CHARGE fish took 212 seconds vs 173 seconds in other fish
3. CHARGE zebrafish had lower heart rates when exposed to anesthesia compared to other fish
    - 168 bpm in CHARGE fish vs 182 bpm in other fish
4. The respiratory rates of CHARGE zebrafish did not drop as low as control fish when anesthetized, and they were higher during the recovery period form the anesthetic
    - 164 breaths/min in CHARGE fish vs 84 breaths per min in other fish

Vagus nerve connection

- CHARGE syndrome is associated with many issues postulated to be connected to vagal nerve dysfunction
- Vagus nerve involved in autonomic nervous system (Control of heart and respiratory rates)
- Vagus nerve dysfunction may contribute to differences in heart and respiratory rates when CHARGE fish are exposed to anesthesia

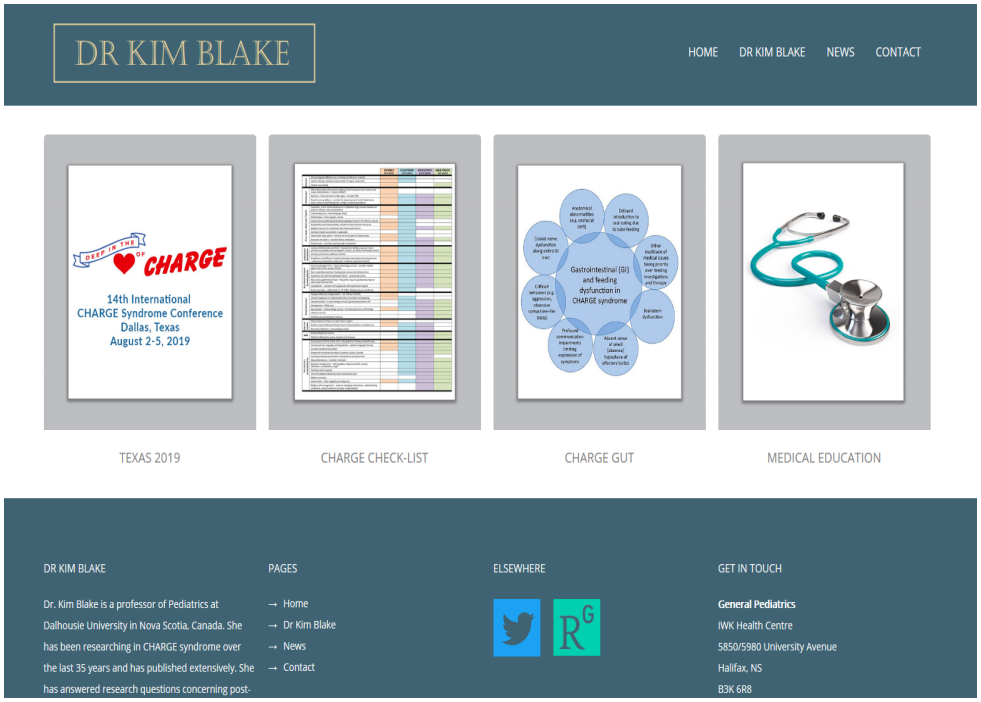
Observations and Future directions

- We have shown that CHARGE zebra fish respond differently to anesthesia similar to what is observed clinically
- Investigate differences in expression of other genes between CHARGE zebra fish and zebra fish with intact chd7
- By finding these differences we can get a better idea of how diseases develops which can help inspire new interventions

Take home messages

- Your children are at high risk for anesthesia related complications. Combining procedures during one anesthesia does not increase the risk of airway events.
- The anesthesiologist needs to be aware that, even with simple procedures, Individuals with CHARGE syndrome are at high risk for anesthesia events.

Questions and Answers



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