Dysphagia in Infants and Young Children with Congenital Heart Disease

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Disclosure

- I have no relevant financial or nonfinancial relationships within the products and services described, reviewed, evaluated, or compared in this presentation.
Introduction

- 8 cases per 1000 live births
- Number 1 cause of birth defect related deaths
- Each year in the US approximately 40,000 infants are born with congenital heart disease
To understand congenital heart disease, first we must understand the heart.
Anatomy of the Heart
Right side of the heart

- Oxygen depleted blood enters through the superior and inferior vena cava
- Atrium contracts
- Blood flows through tricuspid valve to the right ventricle
- Tricuspid valve closes when ventricle is full to prevent blood flow back to the right atrium
- Right ventricle contracts and blood flows through the pulmonary valve to the lungs for oxygenation
Oxygenation

- Blood travels through the pulmonic valve to the pulmonary artery then to the lungs
- Oxygen moves from tiny air sacs in the lungs, through the capillaries to the blood
- Carbon dioxide moves from the blood to the sacs
- Oxygenated blood then travels to the left atrium through the pulmonary veins
Left side of the heart

- Oxygen rich blood enters left atrium
- Blood flows from the left atrium through the mitral valve to the left ventricle
- When the ventricle is full, the mitral valve closes to prevent blood flow back to the left atrium
- The ventricle contracts and blood flows through the aortic valve to the aorta and to the body
# Types of defects

**Cyanotic**
- Deoxygenated blood is mixed with oxygenated blood
- Results in cyanosis
- Increased heart rate and respiratory rate, cough

**Acyanotic**
- Includes obstructive defects
- Oxygen rich blood is sent to the lungs
- No cyanosis
- Increased heart rate, cough, fatigue, respiratory distress
A cyanotic and obstructive defects:

- Patent Ductus Arteriosus
- Atrial Septal Defect
- Ventricular Septal Defect
- Pulmonary stenosis
- Aortic stenosis
- Coarctation of the aorta
Patent Ductus Arteriosus

- Patent Ductus Arteriosus is failure of the ductus to close
- Symptoms
  - Fast breathing
  - Poor feeding
  - Rapid pulse
  - Shortness of breath
  - Sweating while eating
  - Tiring easily
  - Poor growth
Atrial Septal Defect

- Hole in the wall between the right and the left atria (atrial septum)

- Symptoms
  - Difficulty breathing
  - Shortness of breath
  - Palpitations
  - Frequent respiratory infections
Ventricular Septal Defects

- Hole in the wall between the right and left ventricles (ventricular septum)

- Symptoms
  - Shortness of breath
  - Fast/hard breathing
  - Paleness
  - Failure to gain weight
  - Fast heart rate
  - Sweating while feeding
  - Frequent respiratory infections
Pulmonary Stenosis

- Narrowing of the pulmonary valve
- Symptoms
  - Abdominal distension
  - Chest pain
  - Fainting
  - Fatigue
  - Poor weight gain
  - Shortness of breath
Aortic Stenosis

- Narrowing of the aortic valve or above or below the valve

- Symptoms
  - Breathlessness
  - Chest pain (angina type)
  - Fainting/dizziness
  - Palpitations
  - Easily tired with exertions
  - Failure to gain weight
  - Poor feeding
  - Breathing problems
Coarctation of the aorta

- Narrowing of the aorta, usually in the descending aorta
- Symptoms
  - Chest pain
  - Cold feet/legs
  - Dizziness/fainting
  - Decreased exercise capacity
  - Failure to thrive
  - Leg cramps
  - Nosebleed
  - Poor growth
  - Headache
  - Shortness of breath
Cyanotic Defects

- Due to deoxygenated blood bypassing the lungs and entering the blood system
- Patient appears cyanotic
- Due to right-to-left or bidirectional shunting, or malposition of the great arteries
Cyanotic Defects

- Tetrology of Fallot
- Hypoplastic Left Heart Syndrome
- Malposition of the Great Arteries
- Ebstein’s Anomaly
- Truncus Arteriosus
- Total Anomalous Pulmonary Venous Return
Tetrology of Fallot

- **Four components**
  - A ventricular septal defect (malalignment)
  - Pulmonary stenosis
  - Overriding aorta
  - Right ventricular hypertrophy

- **Symptoms**
  - Cyanosis
  - Clubbing of fingers
  - Difficulty feeding
  - Failure to gain weight
  - Passing out
  - Poor development
  - Squatting during cyanosis
Hypoplastic Left Heart Syndrome

- Underdevelopment of the left side of the heart, including the left atrium and ventricle, mitral valve, aortic valve, and aorta

- Symptoms
  - Cyanosis
  - Lethargy
  - Cold hands/feet
  - Poor pulse
  - Poor suck/poor feeding
  - Pounding heart
  - Rapid breathing
  - Shortness of breath
Malposition of the great arteries

- The aorta and the pulmonary artery are inverted
- The aorta carries deoxygenated blood from the right ventricle to the body
- The pulmonary artery carries oxygenated blood back to the lungs
- Symptoms
  - Cyanosis
  - Clubbing of fingers or toes
  - Poor feeding
  - Shortness of breath
Ebstein’s Anomaly

- Malformation of the tricuspid valve
- Either an atrial septal defect or a patent foramen ovale is present
- In some patients, a pulmonary valve stenosis or pulmonary valve atresia is also present
- Symptoms
  - Cough
  - Failure to grow
  - Fatigue
  - Rapid breathing
  - Shortness of breath
  - Fast heartbeat
Truncus Arteriosus

- The aorta and the pulmonary artery are not fully separated
- A single arterial trunk arises above a VSD
- **Symptoms**
  - Cyanosis
  - Failure to grow
  - Fatigue
  - Lethargy
  - Poor feeding
  - Rapid breathing
  - Shortness of breath
  - Clubbing of fingers or toes
Total Anomalous Pulmonary Venous Return

- Pulmonary veins drain into the right atrium rather than the left atrium
- Symptoms
  - Cyanosis
  - Frequent respiratory infections
  - Lethargy
  - Poor feeding
  - Poor growth
  - Rapid breathing
- Acyanotic defects
  - Difficulty breathing
  - Shortness of breath
  - Respiratory infections

- Obstructive defects
  - Also difficulty breathing and shortness of breath
  - Chest pain
  - Respiratory infections

- Cyanotic defects
  - Cyanosis
  - Lethargy
  - Failure to grow

- Feeding difficulty reported as a symptom across defect types
Jadcherla and colleagues (2009)

- Retrospective study assessing feeding abilities in neonates with congenital heart disease
- 76 infants (29 acyanotic and 47 cyanotic)
- Cyanotic CHD required 3 times longer use of mechanical ventilation, narcotics and vasopressor use
- Acyanotic group: prolonged respiratory support correlated linearly with time to attain maximum gavage feeds and nippling
- Cyanotic group: delayed initiation of gavage feedings and prolonged respiratory support correlated linearly with time to attain maximum gavage feedings and nippling
Recurrent Laryngeal Nerve

- Loops around the aorta
- Positioned to increase manipulation or injury during surgical intervention involving the aorta
- Injury can result in left vocal fold paralysis
Pereira and colleagues (2006)

- VCP after PDA ligation
- 100 infants enrolled – 7 with VCP
- 2 patients with stridor and feeding difficulty
- 5 patients with average follow-up 9 months post surgery
- At last follow-up, none had feeding difficulty
Truong and colleagues (2007)

- Retrospective study from 4 pediatric hospitals
- 109 children with VCP were identified
- 35% recovered VC function in 6.6 months
- 65% continued to have VC dysfunction at 16.4 months (at follow-up)
- 45% had laryngeal penetration or aspiration on a modified barium swallow study
Clement and colleagues (2008)

- Case-control study compared 23 infants undergoing PDA ligation (12 with UVCP and 11 without paralysis)
- 52% of infants had UVCP following PDA ligation
- Patients with UVCP required longer use of feeding tubes, supplemental oxygen, and ventilatory support
Skinner and colleagues (2006)

- Assessed laryngopharyngeal dysfunction after the Norwood procedure
- 33 of 36 infants received postoperative fiberoptic laryngoscopy and a MBS
- LVFP noted in 3 patients
- Abnormal MBS noted in 16 patients (8 with aspiration)
- Of patients with LVFP, 2 had normal MBS
Sachdeva and colleagues (2007)

- Investigated vocal cord dysfunction and feeding difficulties
- 38 children out of 2255 had postoperative VCD
- A swallow study confirmed swallowing dysfunction in 27 patients
- Gastrostomy tube was placed for 18 patients
- At discharge, 18 patients were fed by gastrostomy, 13 orally, 3 by nasogastric tube, and 4 by a combination of oral/NG
Davies and colleagues (submitted)

- Laryngopharyngeal dysfunction BEFORE and AFTER surgery
- Pre-operative oral feeding evaluation revealed dysfunction in 33% of assessments
- 30/57 patients without VCP had an abnormal oral feeding evaluation
- Multivariate modeling of the inability to consume nutrition orally at discharge included preoperative intubation, an abnormal preoperative oral feeding evaluation, and age (larger patients were at lower risk)
Assessment Protocol

- Pre (when possible) and post-operative assessment by ENT and SLP for all infants undergoing surgical intervention surrounding or involving the aortic arch.

- MBS is frequent in infants with left vocal fold involvement but also necessary in infants with normal vocal fold movement.
Predictive Factors

- An abnormal preoperative oral feeding evaluation was predictive of an abnormal postoperative evaluation.
- Other predictive factors of an abnormal oral feeding evaluation:
  - Genetic syndrome
  - Preoperative mechanical ventilation
  - Preoperative shock
  - Vocal fold paralysis
Predictive Factors (MBS)

- Previous arch intervention
- Previous palliative procedure
- Abnormal preoperative oral feeding evaluation
- Abnormal postoperative oral feeding evaluation
- Patients intubated preoperatively
- Vocal fold paralysis was not predictive of an abnormal MBS in this study
Conclusions

- Postoperative swallow function following surgeries surrounding the aortic arch is more complicated than the presence or absence of vocal fold paralysis
- Influenced by intrinsic patient characteristics and perioperative care
- Preoperative evaluations identified sensory and motor dysfunction which influenced feeding success
- No single assessment identified all patients at risk of swallow dysfunction
Unilateral (Left) Vocal Cord Paralysis - Conclusions

- Literature is varied, but appears to support a variety of factors which influence oral feeding success
- Left vocal cord paralysis should be considered as a factor, but viewed as one of many influencing factors on potential feeding and swallowing outcomes
The nonnutritive assessment will be the foundation on which oral feeding is built.
The nutritive assessment is the first of many assessments in route to successful oral feeding.
Oral Feeding Evaluation

- Root, latch, and suck
  - Extraction with sequential sucking
    - No signs of aspiration: initiation of oral feedings
  - Limited extraction, refusal, elimination
    - Signs of aspiration, limited to no oral trials
    - Limited to no oral trials
Assessment Outcomes

- Normal interest, coordination without signs of aspiration
- Signs of aspiration
- Hypersensitivity
- Poor coordination
- Poor stamina
- Prolonged food refusal
Signs of Aspiration

- Cough
- Congestion
- Refusal
- Increased respiratory rate
- Silent
Completion of a VFSS

- When a patient is mobile
- Trial of interventions
  - Least invasive: nipple changes
  - Moderately invasive: position changes (sidelying)
  - Most invasive: thickening of liquids or non-oral feeding
Goal of every therapist should be to maximize skill and minimize risk simultaneously.
Establish Oral Feeding Plan

- Identify an intervention that decreases risk of aspiration
- May have poor coordination or food refusal in addition to aspiration
- Provide ongoing monitoring of progress through regular observation of bottle feeding
- Requires coordination and communication between all team members
Hypersensitivity

- Increased irritability in response to handling
- Gagging with attempts to stimulate suck
- Facial grimace with attempts to stimulate a suck
- Gagging or facial grimace with oral stimulation in older children
Intervention for Hypersensitivity

- Start where infant or child can maintain a calm state
- Calming strategies such as containment can increase tolerance in infants
- Oral stimulation progressing to taste stimulation can increase skills in older children
Poor Stamina

- Initial active root and interest
- Suck vigor and overall alertness decreases as the feeding progresses
- Intake is limited
Intervention for Poor Stamina

- Pacing can improve stamina
- May improve after medical intervention for cardiac complications
- Education of family members and staff on reading infant’s cues
Poor Coordination

- Increased respiratory rate
- Can affect airway protection
- Affects stamina
Intervention for poor coordination

- Nipple changes
  - Slow flow nipples
  - Bionix bottle
- Pacing
- Horizontal bottle position
- Monitoring of signs of stress
Prolonged Food Refusal

- Frequently observed across ages
- Infants refuse to latch or refuse after limited amounts
- Young children push away food and sometimes liquids
Intervention for prolonged food refusal

- Establish a mealtime routine
- Provide scheduled meals and snacks
- Positive reinforcement for accepted bites
- Calming strategies to increase acceptance of bottles
Disorders of swallowing impact development, health, and social interactions.
THE OUTLOOK AHEAD....