Tuesday, 2:30 – 4:00, C1

**Pulmonary Complications in Children and Adolescents with Developmental Disabilities**

*John Marks, MD*

**Objective:**

Identify advances in clinical assessment and management of selected healthcare issues related to persons with developmental disabilities

**Notes:**
Respiratory Problems in Children with Developmental Disabilities

John Marks, MD, FAAP, FCCP

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Common Respiratory Problems in Children with DD

- Etiology of different respiratory problems
- Studies that help define the problems
- Evaluation and diagnosis
- Treatment options
- Case Studies

Common Respiratory Problems in Persons with DD

- Recurrent aspiration
- Upper airway obstruction, obstructive sleep apnea (OSA)
- Lower airway obstruction
- Restrictive lung disease
Case #1

AJ is a 7 month old male infant who is admitted to the hospital with his 2nd episode of RUL pneumonia since discharge from the NICU 3 months ago. According to his mother AJ has had persistent respiratory symptoms since discharge consisting of nearly daily wheezing which waxes and wanes. Past medical history consists of severe prematurity (27 weeks, 850 g), hyaline membrane disease with 26 days of mechanical ventilation, grade II intraventricular hemorrhage, and PDA (closed spontaneously). AJ was discharged from the NICU at 3 months of age (wt 4 lbs) and has been weaned recently from continuous oxygen.

Case #1. AJ 7 months, RUL pneumonia. What additional history would be useful in your evaluation?

Case #1 - Additional History

- **Developmental milestones** - not sitting, poor axial control, rolls front to back but not back to front, increased lower extremity tone.
- **Relationship of feeding to respiratory symptoms** - increased wheeze and/or noisy (wet) respirations at the start of feeding or often immediately following feeds or occasionally between feeds.
- **Feeding** - up to 1 hour to take a 4 ounce bottle with frequent gagging and choking on feeds.
- **Neurological history** - irritable but no seizure disorder.
Case #1-Physical Examination
- Small, irritable infant in mild respiratory distress with intercostal retractions. T 101F, RR 32, HR 110. Wt. 5 lbs 1 oz.
- Chest-crackles RUL posteriorly and diffuse wheezes.
- Cor-RRR with no murmur, sinus tachycardia.
- Abdomen-slightly distended.
- Occasional opisthotonic posturing during the exam and increased lower extremity tone.
- HEENT-prolonged gag with pooling of secretions in the oropharynx, some drooling.
- From the history and physical examination you suspect:

Case #1-Working Diagnosis?
- Developmental delay, spastic diplegia.
- Aspiration pneumonia?
  - Antegrade due to oral-motor dysfunction?
  - Retrograde-due to gastroesophageal reflux?
- Bacterial pneumonia?
- Infantile asthma?
- Bronchiolitis?
- Bronchopulmonary dysplasia?
- What are your plans for treatment and further evaluation?

Case #1-Treatment and Further Evaluation
- Obtain blood culture and respiratory culture and start antibiotics for aspiration/bacterial pneumonia.
- Aerosolized β-adrenergic agonist, IV or PO corticosteroids.
- Oxygen as needed.
- IV fluids.
- After a few days AJ is improved. Which test(s) would you order to evaluate possible aspiration?
Case #1 - Further Evaluations to Consider

- **Barium esophagram, Upper GI** - best for anatomic evaluation of upper GI tract. Specific but relatively insensitive for GER.
- **Flexible bronchoscopy with BAL** - Good for obtaining specific bacterial cultures from LRT; inflammation and lipid laden macrophages suggests chronic aspiration.
- **Swallowing videofluoroscopy** - best test to evaluate oral-motor dysfunction, presence of feeding specialist important, anatomical information is obtained.
- **Endoscopy with esophageal biopsy** - specific for esophagitis.
- **Esophageal pH probe** - most specific and sensitive test for GER.
- **Radionuclide “milk scan”** - low sensitivity, but can show aspiration.

Lipid laden macrophage
Case #1-Further Evaluation and Treatment

- You elect to do a modified barium esophogram with video fluoroscopy and a speech therapist present followed by a 24 hour pH probe.
- Results of the swallowing study: Delayed oral preparatory phase (I) with anterior leakage. Adequate oral transport (II) but abnormal pharyngeal transfer (III) with multiple swallows. Vacular pooling of bolus with subsequent aspiration during inspiration accompanied by cough. Adequate esophageal transport (IV).
- This is noted to be more severe with thin liquids vs thick liquid (puree consistency).

Case #1-Further Evaluation and Rx (cont.)

- You proceed with the 24 hour pH probe which shows moderate GER (pH < 4 for > 5% of 24 hours recording).
- Your therapeutic plan includes oral sensory therapy with speech therapy, small frequent feedings, upright positioning after feeds with gentle handling, and oral ranitidine (Zantac).
- AJ returns to your office 8 weeks later. His weight is increased 1 lb, he is less irritable, and wheezing has decreased from multiple daily episodes to about twice/week.

Gastrointestinal (GI) Disorders in DD - How Common are They?

- 58 children with CP (6 mo. to 12 yrs.).
- 92% had GI symptoms.
  - 60% swallowing disorders.
  - 32% regurgitation/vomiting.
  - 41% chronic aspiration.
- 41/45 with GER symptoms had abnormal pH probe studies and/or esophagitis.
- Symptoms not related to CT/MRI brain findings.

Factors Contributing to Recurrent Aspiration in DD

- Swallowing disorders (dysphagia)
- Gastroesophageal reflux (GER)
- Excess oral secretions, drooling (sialorrhea)
- Upper airway obstruction

Feeding Problems in Children with DD

- 1/3 will develop feeding problems severe enough to cause FTT and interfere with social development.
- Problems encountered:
  - Increased oral loss due to poor lip closure, jaw instability, primitive reflexes (suckling).
  - Prolonged feeding time (>30 minutes) often with multiple swallows to clear bolus.
  - Pocketing of food due to defective lateral tongue control or texture aversion.
  - Coughing, choking, gagging—may be “texture specific”. If it occurs at start of feed it indicates suck-swallow incoordination; if at end, possible GER.

Aspiration of Oral Contents

- Up to 90% of those children found to aspirate on barium videofluoroscopy do so *silently*!
- Signs of aspiration:
  - Infant—apnea, bradycardia, recurrent or persistent wheezing.
  - Older child and adolescent—cough, increased congestion, wheezing during meals.
- Complications—includes recurrent tracheobronchitis, pneumonia, hypoxemia, and bronchiectasis due to chronic secondary bacterial infection.
Swallowing Disorders and Aspiration
- Treatment and Prevention

- Oral sensorimotor therapy
- Feeding gastrostomy tube
  - Retrospective survey of caregivers of NI children following feeding gastrostomy (some with antireflux surgery)
  - 29 replies to 35 questionnaires
  - Coughing, choking, and vomiting improved in most cases
  - Feeding time reduced
  - Only one parent regretted the surgery
Feeding Gastrostomy (FG)- Is Antireflux Surgery Necessary?

- FG in 107 NI children, 98 had pre-op radiographic GER evaluation
- 44 with GER
  - 33 FG + ARS
  - 11 FG only - 5 needed ARS
- 51 without GER - FG only
  - 22 (44%) developed GER
  - 17 needed ARS


Swallowing Disorders - Summary

- 60% of DD/NI patients have swallowing disorders
- Aspiration is most significant complication
- Aspiration may be “silent”
- Evaluate with modified barium video swallowing study
- Gastrostomy, including PEG (percutaneous endoscopic gastrostomy), effective in preventing aspiration
- “Protective” antireflux surgery not always necessary

Gastroesophageal Reflux & Retrograde Aspiration
**GER in Children with CP**

- 23 children (0.6 to 11.8 yrs.)
- 70% had feeding problems
- 52% had FTT
- 31% recurrent pneumonia
- 31% anemia
- 70% had abnormal pH probe studies
- GER not related to developmental or chronologic age


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**GER in Children with CP**

- 32 children (0.7 to 19 yrs.)
- pH probe and esophageal manometry
  - 15 (47%) had mild GER
  - 10 (31%) had moderate to severe GER
  - 7 (22%) no GER
  - 10 (31%) had abnormal manometry
  - 13 iron deficient, 5 anemic
  - 2 died presumably from aspiration


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**Factors Associated with GER in Neurologically Impaired Children**

- Low basal tone of gastroesophageal sphincter.
- Supine positioning.
- Abdominal spasticity.
- Scoliosis.
- Seizures.
- Medications.
- Nasogastric intubation.
- Pulmonary hyperinflation.
Evaluation of GER

- Barium esophagram-specific but relatively insensitive-technique very important!
- Radionuclide scan ("milk scan")-intermediate sensitivity and specificity-may require N/OGT.
- 24 hour pH probe- “gold standard”.
- Endoscopy with esophageal biopsy-reveals inflammation-but not a dynamic study.
- Eosophageal manometry-suggests risk for GER.
Treatment of GER

- **Mechanical measures** - positioning, thickened feedings, frequent small feedings, reduce medications that promote low GE sphincter tone, decrease lung hyperinflation.
- **Pharmacologic therapy** - H2 blockers (ranitidine), prokinetic agents (metaclopromide), proton pump inhibitors (omeprazole).
- **Antireflux surgery** - Nissen fundoplication with or without feeding gastrostomy tube.

GER Pharmacotherapy - Ranitidine

- Children with DD and esophagitis
  - endoscopy and pH probe monitoring
  - two doses - 9.3 mg/kg/day or 14.8 mg/kg/day given for 3 mo.
  - lower dose: no improvement in esophagitis
  - higher dose: improved symptoms, but only slight microscopic improved
  - infrequent increase in gastric pH

GER Pharmacotherapy - Omeprazole

- Increases gastric pH
- Decreases total gastric secretion volume
- Facilitates gastric emptying

**Omeprazole in NI Children with Esophagitis**

- 52 patients (4 to 19 yrs.).
- Endoscopically proven esophagitis.
- Omeprazole 40 mg/day (20 mg if <20 kg) as healing dose for 3 mo., then 20 mg/day (10 mg) for 3 mo.
- 44 (86%) remission, 7 (14%) relapsed but improved with higher dose.
- Also improved vomiting, regurgitation, anemia.

**Antireflux surgery carries significant complication rate for children with DD**

- Borgstein et al. 50 children with ARS. 14 pts had early and 9 late complications, 12 required reoperations, 9 deaths (2 at reoperation).
- Pearl et al. 153 children with ARS. Post operative complications in 26%, 19% required reoperation, 9% mortality, combined failure rate (reoperation + deaths) was 28%.
- Rice et al. 52 children with ARS. 6% perioperative deaths.

**GER in Persons with DD**

**Summary**

- GER is common in persons with DD
- Complications of GER include:
  - recurrent aspiration with risk of death
  - esophagitis
- Evaluation should include:
  - esophagram, pH probe monitoring, and sometimes endoscopy
- Treatment with omeprazole will help esophagitis
- For those at risk for recurrent aspiration, antireflux surgery benefits outweigh the risks
Case # 2
RF is a 12 year female admitted to the hospital with her 3rd pneumonia this year. Her chest xray shows severe scoliosis and increased perihilar markings consistent with chronic bronchitis. She also has a RML infiltrate which appears to be new. RF has significant developmental delay with cognition at infantile levels and unmeasurable fine and gross motor age. She has significant contractures of extremities as a result of spastic quadraplegia. Because of documented aspiration of food stuffs and gastroesophageal reflux during her first few years RF underwent Nissen fundoplication and gastrostomy tube placement. She is fed entirely by G-tube. Besides her acute illness, of concern to the family is her constant drooling which, despite frequent oral suctioning has caused maceration of the skin around her mouth.

Case #2-Physical Examination
Her examination shows a small, preadolescent, white female with significant hypertonia, moderate clonus when stimulated, protuberance of her right anterior chest wall, distant heart sounds, and mild contractures of both upper and lower extremities. There is significant pooling of saliva in the posterior pharynx and gag is prolonged and exaggerated when stimulated. Her last two pneumonias have both shown *Pseudomonas aeruginosa* from respiratory cultures obtained during deep posterior pharyngeal suctioning.

What are the possible causes of her recurrent pneumonias?
Is scoliosis a contributing problem?
Case #2-Possible Causes of Recurrent Pneumonia

- Chronic aspiration of saliva.
- Incompetent Nissen fundoplication with aspiration of feedings.
- Covert feeding by mouth.
- Contribution of scoliosis.
  - Decreased chest wall compliance contributes to ineffective cough, aggravates muscle weakness, distorts relationship of diaphragm to thoracic structures, may affect bronchial caliber if severe.

What studies and/or evaluations would clarify the possible causes?

Case #2-Evaluation

You elect to first evaluate the fundoplication by putting barium into the gastrostomy tube—it is competent.

You have your local speech therapist (or other feeding specialist) evaluate RF and find that her anterior drooling is due to poor lip closure but also that she is pooling significant amounts of posterior pharyngeal secretions that put her at risk of aspiration. Her recurrent pneumonias would suggest that this is the case.

The therapist suggests a program or oral training and something to reduce the amount of pooled secretions. What are the options?

Drooling (Sialorrhea) and Oral Aspiration

- Drooling common in patients with DD/NI
- Swallowing disorders and excess oral secretions can result in chronic aspiration not associated with feeding
- “Salivagram” - oral radionuclide scan can demonstrate pulmonary aspiration
- Treatment options are medical and surgical
Options for Treatment of Drooling

- Pharmacologic agents and side-effects
  - Benzotropine (Cogentin) - blurred vision, tachycardia, constipation, urinary retention.
  - Scopolamine patch (Transderm) - toxic psychosis.
  - Trihexyphenidyl - behavioral changes, urinary retention.
  - Glycopyrrolate (Robinul) - irritability, constipation.

Drooling - Pharmacologic Treatment

- Anticholinergic agents - glycopyrrolate
  - Survey of 54 parents (41 responded) regarding benefit of glycopyrrolate.
    - 37 used glycopyrrolate.
    - 95% improvement in drooling.
    - 44% had side effects (dry mouth, thick secretions, urinary retention).
    - <33% discontinued due to side effects.

- 40 patients treated with glycopyrrolate.
  - 36 (90%) had decreased drooling.
  - 11 (28%) discontinued due to side effects.
Options for Treatment of Drooling

- Surgical treatments and side-effects:
  - Parasympathetic denervation - transient effect.
  - Bilateral submandibular gland excision and parotid duct ligation - parotitis, xerostomia, dental problems.
  - Tracheostomy - vocalization problems, infection.
  - Laryngotraheal separation - no vocalization, permanent.

Drooling - Surgical Treatment

- Bilateral submandibular gland excision and parotid duct ligation.
- 16 patients (16 mo. to 18 yrs.) 6 had tracheostomy at time of surgery.
- Number of pneumonias and hospitalizations 1 yr. before and 1 yr. after procedure.
  - Pneumonias: 2.3+/1.4 before vs. 0.9+/1.2 after.
  - Hospitalizations: 1.2+/0.8 before vs. 0.4+/0.8 after.
- Improved quality of life in most.
- Effect of tracheostomy on results.


Drooling - Surgical Treatment

- Laryngeal diversion.
- 14 patients with life-threatening aspiration.
- Reduced aspiration.
- Improved pulmonary function.

Case 2 Follow-up

RF had a salivagram that documented aspiration not associated with feedings. She was treated with glycopyrolate by gastrostomy tube and a scopolamine patch with significant reduction of her oral secretions. Dose titration helped reduce side effects of dry mouth and constipation. Aspiration episodes were reduced and she did not require any surgical procedures.
Case #3

TS is a 5 year old male who you have followed since infancy with spastic cerebral palsy. He has done quite well to this point and receives regular OT and PT and attends preschool. His disability stems from neonatal asphyxia probably due to placenta abruptio and he has mild cognitive as well as significant fine and gross motor delays. He feeds by mouth and has never had a documented pneumonia. Although he has always had noisy breathing at night, mother notices that this has been decidedly worse lately and he awakes from sleep several nights per week.

What additional history would be useful?

Case #3-Additional History

- Important additional history would include:
  - ? Snoring, apnea, arousal sequence-OSAS, nasal obstruction
  - ? Daytime somnolence-OSAS (less rare in children)
  - ? Cough or wheeze at night-GERD, nocturnal asthma, sinusitis
  - ? History of seizure disorder and increase in frequency-seizures
  - ? Trial of medications, eg. decongestants

He has no seizure history and his mother relates that often his snoring is accompanied by short pauses in respiration followed by a long deep, noisy inspiration that sometimes wakes him. She has not noted increased daytime sleepiness but his appetite has decreased. He seems more irritable as well.

Case #3-Physical Examination and Evaluation

TS’s physical examination shows a small but well nourished youngster with mild to moderated disabilities. His exam is basically unchanged from previous visits although he seems more impatient and irritable. The only additional noted abnormality is 3+ tonsillar hypertrophy and some mild clear nasal discharge. You try nasal decongestants and there is no change in symptoms. You do a sinus x-ray and UGI that are both negative and begin a work up for OSAS.

You order a polysomnogram which shows and apnea/hypopnea index of 9.5 (per hour of TST, normal < 5) and occasional desaturations to 85% without change in HR nor seizure activity. You recommend treatment of OSAS.
Factors Associated with Upper Airway Obstruction/OSA

- Tonsilar and adenoid hypertrophy.
- Pharyngeal collapse (palatal hypotonia).
- Macroglossia.
- Glossoptosis.
- Mandibular hypoplasia.
- Subglottic stenosis.
- Laryngomalacia.
- Tracheomalacia.

OSA in Children with CP

- 233 surveyed for OSA symptoms.
  - 63% snore.
  - 20% nasal obstruction.
  - 20% observed apnea.
  - 15% snoror.
- 48 had sleep studies.
  - 27% increased apnea/hypopnea index.
  - 58% had $\text{SpO}_2 < 85\%$.


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<td>Pulmonary hypertension or polycythemia</td>
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Diagnosis of OSAS

- Polysomnogram ("sleep study") - apneas, hypopneas, cardiac changes, gas exchange abnormalities, seizures during various sleep stages.
- Upper airway fluoroscopy - abnormal airway motion.
- Flexible bronchoscopy - airway abnormalities and tone.
- Pulmonary function testing - airway obstruction.
- Rhinoscopy - allergic rhinitis, sinusitis, polyps, vocal cord dysfunction.

OSAS Treatment Options

- Adenotonsillectomy.
- Uvulopalatopharyngoplasty (UPPP), tongue reduction, tongue-hyoid advancement, aryepiglottoplasty.
- Skeletal expansion procedures - mandibular advancement or distraction.
- Oral appliances
- NPPV-BIPAP, CPAP.
- Tracheostomy.
OSA - Surgical Treatment

- 25 children with CP and OSA.
  - 19 had T&A, 3 with uvulectomy.
  - 6 had adenoidectomy alone.
- F/U mean 34 mo.
  - 76% better, no further surgery.
  - 6 had further surgery.
    - 1 revision of adenoidectomy.
    - 1 tonsillectomy and uvulectomy.
    - 4 tracheostomy.

Magardino & Tom. Laryngoscope 1999;109:1611-15

OSA - Surgical Treatment

- 18 Patients CP and OSA
  - 9 T&A
  - 13 UPPP
  - other - tongue mandible procedures
  - All improved
  - polysomnogram, SpO2
  - 2 (11%) needed tracheostomy


- 10 patients with palatal hypotonicity without significant adenotonsillar hypertrophy
  - UPPP and T&A
  - 8 improved
  - 2 further surgery
OSA - Oral Appliances

- Simple oral airway.
- Elastic-retracted appliance.
  - 5 patients (3 CP, 2 Down’s) with OSA.
  - Custom made maxillary and mandibular splints retracted forward with elastic bands, adjustable, easily inserted.
  - All had improved apnea index on polysomnogram.

OSA - Treatment with CPAP

- 4 patients with NI (6 to 16 yrs.).
- Continuing OSA after T&A.
- All improved 12–48 months after starting CPAP.
  - Better sleep parameters.
  - Improved SpO₂.
Tracheostomies

- Help with airway hygiene by direct access.
- Infection not uncommon.
- Can cause negative psychological, communicative, and social effects on patient and family.
- Increases caregiver burden regarding time spent in administering respiratory care.
- May be avoided in many cases with aggressive surgical alternatives.

Infections associated with chronic aspiration and tracheostomies.

- Tracheostomies often colonized with *Staphylococcus sp.* and *Pseudomonas sp.*
- Both anaerobic and aerobic organisms should be covered (*Peptostreptococcus, Bacteroides fragilis* group, *Fusobacterium sp., Klebsiella, Pseudomonas sp., Morganella, and α-hemolytic streptococcus*).
- Antibiotics should cover penicillin resistant anaerobes as well as aerobes.
Case #3-Treatment

TS underwent tonsillectomy and adenoidectomy with considerable improvement in daytime somnolence, night waking, appetite and irritability. Polysomnogram was repeated 6 weeks after the T & A and found to be normal with no desaturations, an apnea-hypopnea index of 3.5.

Upper Airway Obstruction/OSA Summary

- Upper airway obstruction and obstructive sleep apnea are common in persons with DD
- Any patient with snoring and observed apnea should have a polysomnogram
- Treatment includes oral appliances in few, upper airway surgery in most, and CPAP or tracheostomy in severe cases

Airway Clearance

- Recurrent aspiration results in chronic airway inflammation and excessive airway secretions
- Removal of excess secretions reduces inflammation and risk of infection
- Patients with DD or NI have reduced ability to clear airway secretions
Airway Clearance Techniques

- Chest Physiotherapy, Percussion and Postural Drainage
- Oscillating positive pressure
  - Intrapulmonary percussive ventilation
  - PercussivNeb
  - Acapella
- High frequency chest wall oscillation (the Vest)
- Insuflation-exsuflation (Cough Assist)

Summary

- Children with neurological disabilities are at increased risk of respiratory or related illness most often due to mechanical causes.
- Failure to detect (and correct these problems when possible) leads to increased morbidity most often due to chronic inflammatory pulmonary disease.
- Patients are often at an increased mortality risk from untreated pulmonary or related disease.
- Subtle signs associated with silent aspiration and OSAS leads to delayed diagnosis in many cases.