Caring for children and adolescents with developmental disabilities, such as those with cerebral palsy or traumatic brain injury, is challenging because of their susceptibility to varying degrees of respiratory morbidity and mortality. This morbidity is a consequence of several factors including ineffective cough, dysphagia, gastroesophageal reflux (GER), and lung restriction from chest wall abnormalities. In addition, these children may also have other underlying respiratory conditions, such as asthma or bronchopulmonary dysplasia, which may make evaluation and management more complicated. This article discusses the common pulmonary problems affecting children with developmental disabilities. Topics to be discussed include GER, drooling, and dysphagia and their relation to aspiration and aspiration pneumonia; upper airway obstruction and obstructive sleep apnea; and the role of airway clearance. Case studies are used to introduce a discussion of the underlying etiology, currently accepted methods to evaluate the conditions, and evidence-based treatment options. The goal of managing these problems is to use anticipatory guidance when possible and choose therapies that improve the child’s quality of life with minimal side effects.

CASE 1

AJ is a 7-month-old boy admitted to the hospital with his second episode of right upper lobe pneumonia since discharge from the NICU 3 months ago. He 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 a patent ductus arteriosis that closed spontaneously. He has recently been weaned from continuous oxygen.

Developmental milestones show he is not sitting, has poor axial control, rolls front to back but not back to front, and has increased lower-extremity tone. He has increased wheeze or noisy respirations at the start of feeding or often immediately following feeds or occasionally between feeds. Feeding can take up to 1 hour to take a 4-oz bottle with frequent gagging and choking noted.

On physical examination he is a small, irritable infant in mild respiratory distress with intercostal retractions. His temperature is 38.5°C, respiratory rate is 32, heart rate is 110, and weight is 4.4 kg. Chest auscultation reveals crackles in the right upper lobe posteriorly and diffuse wheezes. Cardiac examination is normal. The abdomen is slightly distended. He is observed to have occasional opisthotonic posturing during the examination and increased lower-extremity tone. Oropharyngeal examination shows a prolonged gag with pooling of secretions and some drooling.

From the history and physical examination suspected diagnoses include developmental delay with spastic diplegia, feeding problems, aspiration or bacterial pneumonia, and possible bronchopulmonary dysplasia. From the feeding history it seems that aspiration is likely.

**Feeding Problems**

Feeding problems are common in children with developmental disabilities and can result in failure to thrive. Some have increased oral loss because of poor lip closure, jaw instability, and primitive reflexes. Feeding times can be prolonged, greater than 30 minutes, with multiple swallows needed to clear a bolus. Some children pocket food because of defective lateral tongue control or texture aversion. Coughing, choking, and gagging may be texture-specific. If these symptoms occur at the start of a feed it usually indicates suck-swallow incoordination. Symptoms at the end of a feed may be caused by GER. Children with severe cerebral palsy may be at risk of hypoxemia during upright oral feedings.

**Aspiration**

The evaluation of aspiration needs to determine if it is antegrade caused by oral-motor dysfunction, or retrograde caused by GER. Factors contributing to recurrent aspiration include swallowing disorders (dysphagia); GER; excess oral secretions and drooling (sialorrhea); and upper airway obstruction. Aspiration into the lungs results in a symptom complex that might be diagnosed as viral or bacterial pneumonia. Common symptoms of aspiration are gagging, coughing, wheezing, and congestion. Aspiration may also exacerbate underlying conditions, such as asthma or bronchopulmonary dysplasia. Chronic or recurring aspiration may result in chronic tracheitis or bronchitis, bronchiectasis, pneumonia, and empyema with increased risk of mortality. One study of 58 children with cerebral palsy age 6 months to 12 years found that 92% had gastrointestinal symptoms. Sixty percent had swallowing disorders, 32% had regurgitation or vomiting, and 41% had chronic aspiration. Of 45 with symptoms suggesting GER, 41 had abnormal esophageal pH probe studies or esophagitis. Twelve of 18 children who had gastric emptying studies demonstrated significant delay and 11 had abnormal esophageal motility. A study of 34 children with severe neurodisabilities, age 7 months to 16 years, assessed 24-hour pH probe monitoring and feeding video-fluoroscopy and related findings to frequency and severity of lower respiratory tract...
infections. Five of 10 children with no history of lower respiratory tract infections had GER and none of the 10 had evidence of antegrade or direct aspiration. Two of eight with infrequent lower respiratory tract infections had GER and four had direct aspiration. Sixteen had chronic recurrent lower respiratory tract infections with one having GER, seven with direct aspiration, and eight with both.

**Antegrade Aspiration**

Swallowing is a complex physiologic process requiring coordination of oropharyngeal, laryngeal, and esophageal muscles. Swallowing consists of four phases. In the oral preparatory phase food is taken into the mouth and processed into a small bolus. In the oral transport phase the bolus is pushed toward the pharynx by the tongue. The pharyngeal transfer phase involves a peristaltic wave in the hypopharynx that propels the bolus into the esophagus. During this phase the nasal and laryngeal airways are protected by the soft palate and epiglottis. In the esophageal transport phase the bolus moves down the esophagus to the stomach. Dysphagia results when there is impaired coordination of the phases of swallowing. Children with developmental disabilities are at increased risk of aspiration caused by dysphagia. Aspiration may occur before, during, or after a swallow (Fig. 1). Although often obvious because of respiratory symptoms, antegrade or direct aspiration can be difficult to diagnosis clinically because aspiration episodes can be “silent.” One study reviewed videofluoroscopic modified barium swallow studies in 186 children with developmental disabilities and found that 48 (26%) had direct aspiration, mostly with liquids, and that in 94% the aspiration was “silent,” that is without coughing or gagging. Another study also describes aspiration as being silent in 68% of children with severe cerebral palsy.

**Evaluation and Management of Antegrade Aspiration**

A video esophagram or modified barium swallow study is the best way to evaluate the swallowing in children suspected of having antegrade aspiration. Fluoroscopy is done while the patient swallows barium-containing liquids and foods of various textures. The video can be reviewed by a speech pathologist to assess the phases of swallowing and determine if laryngeal pooling or penetration or aspiration has occurred and if there is adequate clearing of any aspirated material by gagging or coughing or if aspiration was silent. Flexible fiberoptic bronchoscopy with bronchoalveolar lavage can be done if there is evidence of pneumonia to obtain specimens for culture and analysis for lipid-laden macrophages. A positive lipid-laden macrophage finding is not specific for aspiration and in the author’s experience has not been very helpful to make that diagnosis.

Management of antegrade aspiration depends on the severity and frequency of episodes. Thickening liquids and giving solids with textures that were well tolerated during a video swallow study may be all that is necessary in mild cases. Oral sensormotor therapy may help improve dysphagia in some patients. Twenty-seven children with cerebral palsy with associated feeding problems (7 aspirators, 20 nonaspirators) underwent 10 weeks of therapy and were found to have significant improvement in feeding, chewing, and swallowing, but no change in drinking skills. For those with more severe aspiration despite dietary manipulation and oral therapy, stopping oral feedings may be necessary. Alternatives are nasogastric tube feeding, gastrostomy, or jejunostomy. Nasogastric tubes can be uncomfortable and exacerbate problems with oral secretions and GER and are often not well tolerated. A gastrostomy tube is the best option to avoid aspiration and improve nutrition. Gastrostomies can be performed surgically or by the percutaneous endoscopic gastrostomy technique. The percutaneous endoscopic gastrostomy procedure is generally preferred over...
surgery because of less intraoperative and postoperative morbidity. A retrospective study of 63 neurologically impaired children who underwent gastrostomy compared a group of 30 who had the percutaneous endoscopic gastrostomy procedure with a group of 33 who had a Stamm (surgical) procedure. The two groups were comparable in age range, cause of impairment, and indication for gastrostomy. None in either group had symptomatic GER. During a 23-month follow-up period minor complications occurred in 30% of cases, but major complications occurred only in the surgical group, including two postoperative deaths. Thirty-nine percent of the surgical patients eventually required fundoplication after developing symptomatic reflux, whereas only 10% of the percutaneous endoscopic gastrostomy patients required antireflux surgery.

Three questions arise regarding gastrostomy tubes in developmentally disabled children. First, is there any increased risk of respiratory morbidity following a gastrostomy? Second, are caregivers satisfied with the results of having a gastrostomy placed? Third, should antireflux surgery be done routinely with either type of gastrostomy procedure? A prospective study of 57 children with cerebral palsy measured the number of chest infections and hospitalizations in the year before and the year after gastrostomy. Results showed a 50% reduction in chest infections and 80%
reduction in hospitalizations in the year after compared with the year before gastrostomy, suggesting there is no increased risk of respiratory morbidity. Another prospective multicenter study measured growth and general health in 57 children with cerebral palsy age 5 months to 17 years, comparing these parameters before and 12 months after gastrostomy placement. Complications of gastrostomy feeding were also assessed. Half of this group were significantly underweight at baseline and had significant weight gain during the study period. There was no evidence of increased risk of respiratory complications. Most parents reported a significant decrease in feeding time and improvement in their child’s general health after gastrostomy tube placement.

Using the same cohort of 57 children with cerebral palsy described previously, the authors also reviewed the impact of gastrostomy tube feeding on the quality of life of the caregivers of the children. Caregivers reported improvements in social functioning, mental health, energy, and general health perception. Another study involved a questionnaire sent to 38 caregivers of neurologically impaired children regarding their perceptions of the outcome after feeding gastrostomy placement. Of the 29 who replied, most reported improvement in coughing, choking, and vomiting, and decrease in feeding time. Weight gain also improved in those who had poor nutritional status at baseline. Only one parent regretted the procedure.

Disabled children who require a gastrostomy and have symptomatic GER should have an antireflux procedure before or at the time of gastrostomy placement. The use of routine or protective antireflux procedures when placing a feeding gastrostomy in children without symptomatic GER has been controversial. A study of 107 neurologically impaired children age 1 month to 16 years undergoing a feeding gastrostomy procedure assessed preoperative and postoperative incidence of GER. Of 98 patients who had preoperative assessment, 44 had documented reflux and 33 of those had antireflux surgery at the time of gastrostomy placement. Seven of 11 patients who did not have antireflux surgery developed symptomatic reflux, and five required an antireflux procedure. Fifty of 54 patients without preoperative evidence of reflux who did not undergo antireflux surgery were followed for 20 months after gastrostomy placement. Twenty-two developed symptomatic reflux and 17 eventually had antireflux surgery. The risk of developing reflux was not related to age, gender, indication for gastrostomy, underlying diagnosis, or method of gastrostomy. The authors concluded that routine protective antireflux surgery is not justified in those patients without preoperative evidence of GER.

A prospective study of 148 neurologically impaired children undergoing feeding gastrostomy found that 105 had preoperative evidence of reflux based on upper gastrointestinal contrast and esophageal pH probe studies. All of these patients had antireflux surgery. Thirty-seven of 43 patients without evidence of reflux were doing well at a mean follow-up of 21 months. Six (14%) of the 43 developed symptomatic reflux and five required antireflux surgery. Another study of 19 neurologically impaired children undergoing percutaneous endoscopic gastrostomy placement at a mean age of 34 months without an antireflux procedure found that over a 21-month follow-up no child developed choking, gagging, or retching. The results of these two studies show that between 14% and 44% percent of neurologically impaired children undergoing gastrostomy placement eventually develop symptomatic reflux, but this does not support the idea that routine antireflux surgery should be done in all cases.

**Case 1 Follow-Up**

AJ underwent a modified barium esophagram with video fluoroscopy and a speech therapist present followed by a 24-hour pH probe. Results of the swallowing study
showed delayed oral preparatory phase with anterior leakage, adequate oral transport
but abnormal pharyngeal transfer with multiple swallows, and valecular pooling of
bolus with subsequent aspiration during inspiration accompanied by cough. Esophageal
transport was adequate. Aspiration was noted to be more severe with thin liquids
versus thick liquid (puree consistency). The 24-hour pH probe showed moderate GER
(pH <4 for >5% of 24 hours recording). The management plan included oral sensory
therapy with speech therapy, small frequent feedings, upright positioning after feeds
with gentle handling, and oral ranitidine. Eight weeks later AJ’s weight increased 1 lb,
he is less irritable, and wheezing has decreased from multiple daily episodes to about
twice per week. Because of his significant improvement gastrostomy and antireflux
surgery were not considered.

Gastroesophageal Reflux and Retrograde Aspiration

As noted in the discussion of the case study, GER is common in children with develop-
mental disabilities. A study of 23 children with cerebral palsy age 7 months to 12
years found that 70% had severe feeding difficulties, 52% had failure to thrive, 31%
had anemia, and 31% had recurrent chest infections.16 When investigated for GER
using 24-hour pH probe monitoring 16 patients (70%) had abnormal studies with a re-
flux index, the percent of time with esophageal pH less than 4%, greater than 5%
(median, 11.4%). Presence of reflux was unrelated to developmental age or severity
of cerebral palsy. Another study of 32 children with cerebral palsy age 8 months to
19 years who had 24-hour pH probe monitoring found reflux in 78%, 47% mild,
16% moderately severe, and 16% severe.17 Abnormal esophageal radiography and
motility studies were common. Chest radiographs showed consolidations in three of
the patients, presumably caused by retrograde aspiration. There are several reasons
that GER is common in developmentally disabled children. Esophageal dysmotility
with low basal lower-esophageal sphincter tone are common findings. Abdominal
spasticity, scoliosis, and pulmonary hyperinflation can mechanically contribute to re-
flux. Nasogastric or gastrostomy tubes may increase GER.

Evaluation and Management of Gastroesophageal Reflux

Symptoms of GER include vomiting; dysphagia; stridor from laryngospasm; gagging;
coughing; and dystonic movements (Sandifer’s syndrome).18 Hematemesis, anemia,
and iron deficiency may result from severe esophagitis. When associated with aspira-
tion recurrent pneumonia is common. When GER is suspected several methods are
available to confirm the diagnosis. A simple barium esophagram can identify anatomic
abnormalities of the esophagus, but may not demonstrate GER. A radionuclide “milk”
scan can demonstrate reflux, assess gastric emptying, and on delayed scanning may
reveal aspiration into the lung. Esophageal manometry may show abnormal motility
but by itself does not diagnose GER. Endoscopy with esophageal biopsy can identify
chronic inflammatory changes and support a diagnosis of GER. The 24-hour esopha-
geal pH probe study remains the gold standard for diagnosis of GER.

When pathologic GER is documented different treatment options need to be consid-
ered. Dietary changes, such as avoiding spicy and acidic foods, and keeping the child in
an upright position during and after feedings, may alleviate the problem in mild cases. If
these measures do not improve symptoms, then pharmacologic therapy is indicated.
There are few studies assessing the effectiveness of histamine2 receptor blockers in de-
velopmentally disabled children. A study of children with cerebral palsy and GER eval-
uated with endoscopy and esophageal biopsy and pH probe monitoring demonstrated
little improvement in esophagitis after 3 months of ranitidine at a dose of 9.3 mg/kg/d.19
After 3 months at a dose of 14.8 mg/kg/d there was slight improvement in esophagitis, but symptoms improved. Ranitidine only infrequently increased gastric pH.

There are also few studies of proton pump inhibitors for the treatment of GER in disabled children. In one study, 52 institutionalized intellectually disabled children age 4 to 19 years with endoscopically diagnosed esophagitis were treated with omeprazole. Baseline esophagitis was graded as to severity from 1 (mild) to 4 (severe) with 74% having grade 1 or 2 and 26% having grade 3 or 4. Omeprazole dose was 40 mg/day in those weighing greater than 20 kg and 20 mg/day in those weighing less than 20 kg given as a healing dose for 3 months. The dose was decreased to 20 mg/day or 10 mg/day depending on weight for 3 months of maintenance. Omeprazole was effective in healing and maintaining remission in 86% of patients regardless of baseline severity of esophagitis. Relapse occurred in 14% after decreasing the dose, but all became symptom-free with increasing the dose and at 6 months showed endoscopic healing. The authors conclude that omeprazole is highly effective with no adverse effects and remission can be maintained by titrating the dose. Another study of 57 children age 1 to 16 years with erosive esophagitis assessed the safety and efficacy of omeprazole. About 50% of the patients had neurologic impairment and 67% had grade 3 or 4 disease. This was a dose ranging study with healing dose determined by pH probe study showing acid reflux less than 6% over 24 hours. Endoscopy at 3 months revealed healing in 54 of 57 patients and all patients had improvement in symptoms. The healing dose correlated with the severity grade. The authors concluded that omeprazole was effective, safe, and well tolerated.

Two other pharmacologic agents have limited study in the management of GER in developmentally disabled children. Cisapride, a prokinetic agent no longer on the market, was studied in children with cerebral palsy and symptoms of GER. Fifteen of 28 patients had abnormal 24-hour pH probe studies and were started on cisapride, before it was removed from the market. After 3 months of therapy eight patients had improvement in GER symptoms and 8 of 12 who had repeat pH probe studies showed improvement. No electrocardiographic abnormalities were noted. Although this agent is not available, these results demonstrate that newer prokinetic drugs may have a place in the management of GER in disabled children. Baclofen, a γ-aminobutyric acid agonist, has been shown to reduce lower-esophageal sphincter relaxations in normal adults and children with GER. A study of eight neurologically impaired children with GER age 2 months to 16 years assessed the effects of baclofen on GER. Baclofen was administered 30 minutes before meals for 7 days. Twenty-four hour pH probe monitoring was done at baseline and on day 7 and frequency of emesis was recorded. Emesis frequency decreased and the number of reflux episodes greater than 5 minutes decreased, although total time with pH less than 4 did not change as measured on day 7.

If pharmacologic therapy fails to control GER symptoms antireflux surgery has to be considered. The standard procedure is fundoplication or wrap to prevent reflux of stomach contents into the esophagus. The Nissen procedure done through an open incision or more recently laparoscopically is the most common. Several questions need to be asked about antireflux surgery: how safe is it and what are the risks of intraoperative and late complications, is it effective at reducing morbidity and improving quality of life with minimal side effects, and what is the failure rate requiring reoperation?

Eighty neurologically impaired children underwent laparoscopic fundoplication, with 48 having a simultaneous gastrostomy placement, and the intraoperative complication rate was only 5%. The late complication rate was 30% and 6% required reoperation. Another study of 198 children who had antireflux surgery found that there
were significant reductions in the incidence of vomiting and pneumonia for an average of 11 months postoperatively. A total of 71% eventually had recurrence of symptoms, however, whereas only 29% remained asymptomatic. The same authors reviewed a larger series of neurologically impaired children postantireflux surgery and found that 75% developed multiple symptoms of GER, but no single symptom predicted documented recurrence. They recommend maintaining a high level of suspicion for recurrence and doing a contrast study to assess the fundoplication mechanically and a pH probe study to assess physiologically. A prospective study of 20 severely impaired children followed for a median of 3.5 years postfundoplication found a significant decrease in vomiting and gastrointestinal bleeding and decreased reflux index. GER recurred in 30%. The incidence of pneumonia before and after surgery was not different. A retrospective study of 52 neurologically impaired children who underwent fundoplication found that there were fewer hospitalizations and hospital days and improved weight gain in the 6 months following the procedure compared with the 6 months before the procedure. Perioperative mortality was 6%.

A cohort of 153 neurologically impaired children and 81 normal children who underwent antireflux surgery were followed for late postoperative complications. Eighty-six percent of the impaired group and 30% of the normal group also had gastrostomy tubes placed. Late complications occurred in 26% of the impaired group and 12% of the normal group. The impaired children were four times more likely to require reoperation, 19% versus 5% of normal children. Mortality from aspiration occurred in 9% of impaired children versus 1% of normals.

Antireflux surgery can improve GER symptoms and quality of life in developmentally disabled children; however, complications are common and eventual operative failure occurs in 25% to 30% of cases. An alternative to fundoplication is to place a gastrojejunal tube for enteral feeding. A retrospective review of 111 neurologically impaired children with GER found 63 who underwent fundoplication and gastrostomy and 48 who underwent image-guided gastrojejunal tube placement. Both groups were followed for 3 to 5 years and various complications were noted. The gastrojejunal group had a higher incidence of bowel obstruction or intussusception than the fundoplication group, 21% versus 8%. Eighty-five percent of gastrojejunal patients had tube-specific complications requiring tube manipulation almost twice per year of follow-up. Eight percent of these patients eventually required fundoplication, whereas 11% of the fundoplication group required reoperation. The authors concluded that gastrojejunal tube placement is a reasonable alternative to fundoplication, although both have high complication rates.

If fundoplication or jejunal tube feedings fail to relieve GER another surgical procedure, esophagogastric separation or dissociation, can be considered. A total of 44 severely impaired children in three different series underwent esophagogastric dissociation with few complications. There was improved nutrition, fewer respiratory infections, improved quality of life, and no recurrence of GER. The most common problem following the procedure was salivary secretion intolerance.

CASE 2

RF is a 12-year-old girl admitted to the hospital with her third pneumonia this year. Her chest radiograph shows severe scoliosis and increased perihilar markings consistent with chronic bronchitis. She also has a right middle lobe infiltrate that seems to be new. RF has severe developmental delay with cognition at infantile levels. She has significant contractures of extremities as a result of spastic quadriplegia. Because of documented aspiration of food stuffs and GER during her first few years RF
underwent Nissen fundoplication and gastrostomy tube placement. She gets no oral feedings. 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.

Her examination shows a small, preadolescent, girl 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. Auscultation of the chest reveals coarse rhonchi and right-sided crackles.

From the history and physical examination suspected etiologies of the pneumonia include chronic aspiration of saliva, incompetent Nissen fundoplication with GER and retrograde aspiration of feedings, and attempted covert feeding by mouth.

**Drooling (Sialorrhea) and Oral Aspiration: Evaluation and Management**

Drooling or sialorrhea is a common finding in neurologically impaired children. Excess salivary gland output can be caused by GER and some medications, such as some anticonvulsants. Some children have a normal amount of saliva production, but have impaired swallowing resulting in pooling of saliva in the oropharynx and hypopharynx. Anterior spilling of saliva is caused by a combination of poor lip seal, thrusting of the tongue, and impaired swallowing. Macroglossia can also contribute. Although anterior spilling does not result in respiratory symptoms it often results in perioral skin irritation and increased work for caregivers to maintain oral hygiene. Pooling of oral secretions in the hypopharynx results in upper airway congestion with a gurgling sound. If the posterior pooling is benign (ie, there is no aspiration) then careful oral suctioning can help clear pooled secretions and relieve the congestion. The most serious respiratory complication of sialorrhea is chronic aspiration not associated with feeding (Fig. 2).

The best method to evaluate and document aspiration of oral secretions is the radionuclide salivagram. A study of 31 children with neurologic impairment and who were predisposed to aspiration had sublingual placement of a radionuclide substance in a drop of saline and then had serial scans over 60 minutes. Eight patients had aspiration distal to the carina with six demonstrating bilateral aspiration. Other reports have demonstrated the usefulness of this method to evaluate salivary aspiration. In children with tracheostomies instilling a small amount of a colored dye in the mouth may demonstrate aspiration if the dye is found in tracheal secretions.

A study of 63 severely disabled children with cerebral palsy compared the salivagram, barium videofluoroscopy, and milk scan as tests for identifying aspiration. The salivagram was positive in 56%, videofluoroscopy in 39%, and milk scan in only 6% of cases suggesting that nonfeeding antegrade aspiration is more common than aspiration because of dysphagia and that many children are prone to both types of antegrade aspiration. The low incidence of positive milk scans probably indicates that it is an insensitive test to detect retrograde aspiration.

As with treatment of GER, treatment of drooling and nonfeeding oral aspiration involves both pharmacologic and surgical methods. Because salivary gland secretion is controlled by cholinergic stimulation by way of muscarinic receptors, anticholinergic agents are commonly used to lessen anterior drooling and aspiration from salivary pooling. Although these medications are effective, anticholinergic side effects, such as, tachycardia, blurred vision, constipation, urinary retention, dry mouth, flushing, and behavioral problems, commonly occur. Atropine and benztropine both cross the blood-brain barrier and can cause central side effects. A study of 20 children and adults with cerebral palsy and severe drooling were given benztropine in a single
daily dose in a placebo-controlled crossover 4-week trial. As scored by observers there was a 65% to 70% decrease in drooling. Three patients discontinued the medication because of side effects. Glycopyrolate was studied in a 40 children and young adults with cerebral palsy and drooling in a prospective, open-label design. Thirty-six (90%) had a reduction in drooling, whereas two had no benefit. Eleven (28%) stopped the drug because of side effects, but 26 (65%) continued the drug because of perceived benefit. A survey of caregivers of 37 children with cerebral palsy and drooling treated with glycopyrolate found that 95% reported significant improvement. Side effects occurred in 44%, but less than one third reported discontinuation of the drug.

Transdermal scopolamine patches have also been used to reduce drooling in developmentally disabled children. Patches are replaced every 3 days and only come in one dose with no specific pediatric dose. In the author’s experience the dose can be semi-titrated by cutting the patch in half and securing it with an occlusive bandage. A placebo-controlled study of 10 developmentally disabled children treated with a scopolamine patch (1.5 mg) found that over one half had significant reduction of drooling with no reported side effects.

A newer treatment for drooling is salivary gland injection with botulinum toxin. Twenty children age 6 to 16 years with cerebral palsy and drooling underwent parotid and submandibular gland botulinum toxin injection under ultrasound guidance.

Fig. 2. Suggested approach for evaluating and treating drooling. (Adapted from Toder DS. Respiratory problems in the adolescent with developmental delay. In: Homnick DN, Greydanus DE, editors. Pulmonary disorders in the adolescent. Adolescent medicine: state of the art reviews, vol. 11. Philadelphia: Hanley and Belfus; 2000. p. 622; with permission.)
12 weeks there was significant improvement in drooling, number of bibs required, and quality of life. Eighty-nine percent of parents and children would undergo future injections. Another study of nine children with excessive drooling who had ultrasound-guided injection of the parotid glands with botulinum toxin showed objective improvement in drooling, but subjective improvement in only three, with functionally significant improvement in only one. The authors conclude that higher doses and including the submandibular glands may result in improved efficacy. This treatment is not permanent and may require repeated injections.

When medical treatment does not reduce drooling, aspiration and surgical procedures should be considered. The most common procedure is bilateral submandibular gland excision and parotid duct ligation. A retrospective review of 16 neurologically impaired children age 16 months to 18 years who underwent bilateral submandibular gland excision and parotid duct ligation revealed a significant reduction in number of lower respiratory tract infections and hospitalizations in the postoperative period compared with before surgery. Eight of 11 families contacted reported improved quality of life with decreased care requirements. Another study reported on 36-month follow-up of 31 children with neuroligic disabilities and drooling who had submandibular gland excision and parotid duct ligation. Results showed 87% excellent or good results in drooling control with insignificant morbidity. A larger retrospective study of 93 children who had salivary gland excision and parotid duct ligation found only three postoperative complications. Seventy-two families interviewed from 1 to 10 years postprocedure showed 62 (87%) reported significant improvement in or no drooling.

If these treatments are ineffective in preventing aspiration other surgical procedures should be considered. A tracheostomy alone does not prevent aspiration, but does facilitate airway clearance by giving caregivers access to suction secretions from the lower airways. A more extreme procedure separates the larynx from the lower trachea to prevent aspiration by creating a distal tracheostomy with a laryngeal blind pouch, preventing vocalization. Twenty-three neurologically impaired children had a laryngotracheal separation or diversion procedure done to prevent recurring aspiration of oral secretions. All had reduced lower respiratory infections and hospitalizations by eliminating oral aspiration.

**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 in 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 boy followed since infancy with spastic cerebral palsy. He has done quite well to this point and receives regular occupational and physical therapy and attends preschool. His disability stems from neonatal asphyxia and he has mild cognitive and 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, his mother notices that this has been decidedly worse lately and he awakes from sleep several nights per week. 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.
TS’s physical examination shows a small but well-nourished youngster with mild to moderated disabilities. His examination 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. Nasal decongestants have not helped relieve his symptoms. Sinus radiographs and a barium swallow study are performed that are both negative and a work-up for obstructive sleep apnea (OSA) is begun. A polysomnogram is ordered, which shows and apnea-hypopnea index of 9.5 per hour of total sleep time, normal less than 5, and occasional hemoglobin-oxygen desaturations to 85% without change in heart rate or seizure activity. Treatment of OSA is recommended.

Obstructive Sleep Apnea in Children Who have Developmental Disabilities

Children with developmental disabilities, such as cerebral palsy, are at an increased risk of developing OSA. Factors that contribute to airway obstruction during sleep include tonsillar and adenoid hypertrophy, pharyngeal collapse caused by palatal hypotonia, macroglossia, glossoptosis, mandibular hypoplasia, subglottic stenosis, laryngomalacia, tracheomalacia, and obesity. A survey of parents of 233 children with cerebral palsy age 1 to 5 years found the prevalence of snoring was 63%, nasal obstruction 20%, apneic episodes 20%, and stridor 15%. Of the 48 children who had sleep studies, 27% had abnormal apnea-hypopnea index and 58% desaturated to less than 85% on pulse oximetry. Pharyngeal collapse was the most common cause of obstruction followed by adenoid or tonsillar hypertrophy. A review of polysomnographic findings in nine children with cerebral palsy, mean age 36 months, showed that five were diagnosed as having OSA.

Consequences of OSA include hypoxemia, hypercarbia, pulmonary hypertension, hypoxic seizures, headaches, daytime somnolence, developmental delay, behavior problems, and increased risk of aspiration. Children suspected of having OSA based on chronic snoring and observed apnea should have an overnight polysomnogram. Upper airway fluoroscopy can reveal dynamic upper airway collapse and is available in some sleep laboratories. Flexible fiberoptic bronchoscopy can identify airway anomalies, such as tracheomalacia and laryngomalacia. Otolaryngologic evaluation can help identify nasal and pharyngeal obstructions.

Treatment of OSA ranges from simple measures, such as supplemental oxygen for mild cases, to extensive surgical procedures in more severe cases. As with normal children with OSA, tonsillectomy or adenoidectomy may be all that is needed to relieve symptoms. A review of 27 children with cerebral palsy who had had surgical treatment for OSA showed 19 had adenotonsillectomy, 3 of whom also had uvulectomy, and 6 had adenoidectomy alone. After 34 months of follow-up 76% were doing well without need for further surgery. Four patients eventually required tracheostomy, but overall 84% remained tracheostomy-free. A study of 18 children with cerebral palsy and OSA looked at results of preoperative and postoperative polysomnograms to assess response to aggressive upper airway surgery. Patients ranged in age from 9 months to 17 years and underwent various surgical procedures including tonsillectomy and adenoidectomy in 9, uvulopalatoplasty in 13, plus other tongue and mandible procedures. Postoperatively there were improvements in apnea index, respiratory disturbance index, and lowest oxygen saturation. Fifteen remained tracheostomy-free at a mean follow-up of 30 months. Five patients, three with cerebral palsy and two with Down syndrome, all with OSA, had custom-made, adjustable and easily inserted maxillary and mandible splints that retracted the mandible forward with elastic bands. The oral appliance was well tolerated and all had improved apnea index on polysomnogram.
If surgery does not completely relieve OSA symptoms a trial of nasal or face mask continuous positive airway pressure or bilevel continuous positive airway pressure is indicated. If these measures are not tolerated then tracheostomy is the only other option.

**Case 3 Follow-Up**

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

**AIRWAY CLEARANCE**

The most common respiratory complication in children with developmental disabilities is aspiration. Chronic aspiration results in inflammation of the lower airways and excess airway secretions. Clearing of lower airway secretions is often impaired in children with disabilities because of ineffective cough resulting from such factors as weak respiratory and accessory muscles, abnormal chest wall mechanics from scoliosis, and limited ambulation. Standard postural drainage, percussion, and vibration help to mobilize secretions from peripheral to central airways where they can be expectorated by coughing. The head down position should not be used in patients with GER. Several independently administered airway clearance devices have been shown to be effective in aiding airway clearance in cystic fibrosis patients, including positive expiratory pressure, the intrapulmonary percussive ventilation device, the high-frequency chest wall oscillation devices, and various other devices. These devices have not been well studied in children with developmental disabilities, but children capable of mastering the use of an airway clearance device may benefit. The positive expiratory pressure device and other devices generate patient-controlled positive expiratory airway pressure to open plugged peripheral airways, whereas other devices add airway oscillations. Patients have to have the ability to form a seal at the mouth, have a good inspiratory effort, and sustain a prolonged expiration. Those unable to achieve an adequate seal can use the device with a face mask. Those with weak respiratory muscles may benefit from using the percussive ventilation device or the devices that actively generate oscillating positive pressure, used with a mouthpiece or mask. The high-frequency chest wall oscillating vest does not require cooperation and can be used in most patients. One study found that using the vest in seven children with cerebral palsy in a long-term care facility resulted in a decreased incidence of pneumonia and hospitalization. Secretions are mobilized by transmitting pressure oscillations to the airways from the chest wall. Children unable to generate a forceful cough may benefit from using an insufflator-exsufflator device. Used with a face mask, the device generates a sustained inspiratory pressure followed by a negative expiratory pressure to mobilize secretions during exhalation, assisting a cough. This device has been well studied in both children and adults with neurologic impairment.

**SUMMARY**

Children and adolescents with developmental disabilities are at risk of significant respiratory morbidity and mortality mainly caused by chronic or recurrent aspiration. Antegrade aspiration caused by dysphagia, retrograde aspiration caused by GER, and aspiration caused by sialorrhea can lead to chronic airway damage, recurrent pneumonia, and death. Some patients may suffer from all forms of aspiration resulting in more severe morbidity. These children and adolescents are also at risk of upper
airway obstruction and OSA and its resulting morbidities. Health care providers caring for these patients should have a high index of suspicion for these problems to evaluate them appropriately. Once a problem is identified treatment options should be reviewed and effective measures initiated, avoiding significant side effects. Caregivers should be closely involved in making treatment decisions. Improved quality of life for patient and caregiver results from careful evaluation and management of these problems.

REFERENCES


