

Respiratory Concerns in Children with Down Syndrome

Paul E. Moore, M.D.

Associate Professor of Pediatrics and Pharmacology

Director, Pediatric Allergy, Immunology, and
Pulmonary Medicine

Vanderbilt University School of Medicine

Respiratory Concerns in Children with Down Syndrome

- Overview
- Respiratory infections
- Airway issues
- Sleep issues
- Pulmonary vascular issues

Respiratory Concerns in Children with Down Syndrome: Overview

- Most common reason for children to be admitted to the hospital.
- Respiratory infections can be more severe, and hospitalization often results in admission to the intensive care unit.

Anatomical features in DS that contribute to respiratory concerns

- Craniofacial features
 - Narrowed nasopharynx
 - Flattened mid-face
 - Macroglossia
- Adenotonsillar hypertrophy
- Airway size

Other features of DS that contribute to respiratory concerns

- Low tone (hypotonia)
 - Upper airway muscles: dysphagia
 - Airway: malacia
- Gastroesophageal reflux
 - Contribution to adenotonsillar hypertrophy
 - Contribution to airway inflammation
- Cardiac disease
- Obesity

Pearl #1: Anatomical features and physiologic contributors specific to Down Syndrome result in malacia

- Definition: softening or loss of consistency in any of the organs or tissues
- Origin: Greek *malakía*
softness, tenderness, weakness
- Laryngomalacia, tracheomalacia, bronchomalacia, airway malacia

Respiratory Concerns in Children with DS: Respiratory Infections

- Respiratory infection is a significant burden.
 - Bronchiolitis
 - URI/LRTI: upper respiratory infection and lower respiratory tract infection
 - Pneumonia

Respiratory Concerns in Children with DS: Respiratory Infections

- The specific defect in the immune system is not clear, although a number of studies suggest developmental delay.
- Chronic aspiration is a significant contributor.

Pearl #2: Developmental delay in Down Syndrome can refer to more than the nervous system.

- Coordination of swallow
- Immune development

Pearl #3: Water (reflux) damage can be significant in children with DS.



Symptoms of chronic aspiration

- Chronic cough
- Wheezing not controlled by asthma medications
- Pneumonia
- Radiographic findings that suggest chronicity
 - Right middle lobe syndrome
 - Bronchiectasis

Evaluation for recurrent pneumonia

- Swallowing evaluation to look for aspiration
- Studies looking for reflux
- Chest radiograph when well
- CT scan
- Bronchoscopy and bronchoalveolar lavage
- Immune evaluation
- Allergy evaluation

Respiratory Concerns in Children with Down Syndrome: Airway Issues

Symptoms that reflect airway issues:

- Stridor: Inspiratory wheeze that suggests upper airway obstruction.
- Wheeze: Expiratory wheeze that suggests lower airway disease, including asthma.
- Dyspnea: Shortness of breath
- Retractions: Use of accessory muscles

Upper airway abnormalities

- Narrowing of the nasopharyngeal passages
- Macroglossia
- Adenotonsillar hypertrophy
- Laryngomalacia, worsened by reflux

Lower airway abnormalities

- Subglottic stenosis
 - May be more likely following intubation, as tracheal diameter is smaller
- Tracheobronchomalacia
- Tracheal stenosis

Definition of asthma

- Chronic inflammation
- Airway reactivity to specific triggers
- Reversible airway obstruction
- Manifest as symptoms that can include cough, wheeze, and dyspnea

Asthma in Down Syndrome

- Studies have been equivocal about whether there is increased risk of asthma in children with DS.
- Infants with DS who have bronchiolitis, including RSV, are more likely to require hospitalization.

Evaluation for airway issues

- Careful history
- Physical exam
- Chest radiograph
- Bronchoscopy
- Sleep study
- Echocardiogram

Respiratory Concerns in Children with Down Syndrome: Sleep Issues

- Impact on other development
- Obstructive sleep apnea
- Review of anatomic features
- Increased BMI associated with OSA
- AAP recommendation: Screening at age 1
- Importance of sleep study
- Complication: pulmonary hypertension

Respiratory Concerns in Children with Down Syndrome: Pulmonary vascular issues

- Heart disease, with increased pulmonary blood flow
- Hypoxia
- Airway obstruction

Pulmonary hypertension in newborns with DS

- Persistent pulmonary hypertension
- Can occur with or without heart disease
- “Delay” in vascular remodeling

Review of Pearls

- Anatomical features and physiologic contributors specific to DS result in malacia.
- Developmental delay in DS can refer to more than the nervous system.
- Water damage can be significant.

What Health-Care Providers Can Do to Reduce Respiratory Disease

- Look for aspiration.
- Vaccinate against influenza and pneumococcus.
- Address sleep issues.

What Parents Can do to Reduce Respiratory Disease

- Reduce exposure to viral respiratory infections.
- Maintain a healthy lifestyle to boost the immune system and to reduce obesity.
- Reduce exposure to environmental tobacco smoke.

Suggested Reading

Pulmonary Complications of Down
Syndrome during Childhood

Karen M. McDowell and Daniel Craven,
Cincinnati Children's Hospital Medical
Center

Journal of Pediatrics, 2011