Diagnostic Approach to chILD, Treatment and Future Directions

Alicia Casey, MD
Boston Children’s Hospital
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Courtesy of chILD Foundation
First Steps

- Recognizing ‘chILD’ is important and not always easy
- Diagnosis often delayed or missed
- Symptoms overlap with other respiratory diseases
- Look for respiratory symptoms OUT OF PROPORTION to more common or known underlying disease

A Boston Children’s Hospital patient
Diagnostic Team

- Multi-disciplinary team with knowledge of chILD
- Individual patient & family centered approach

Boston Children’s Hospital chILD Program Team

A Boston Children’s Hospital patient
National Team: Children’s Interstitial Lung Disease Research Network

Slide courtesy of Robin Deterding
Team Objectives

• Primary role of diagnostic process is to answer a clinically relevant question

• Team’s responsibility to weigh risk and benefit appropriately
Diagnostic Approach

- Guided by team experience and chILD expert guidelines
- Rule out other common causes of diffuse lung disease
- Radiographic imaging
- Lung function testing
- Bronchoscopy
- Genetic testing
- Lung biopsy
- Look for coexisting complications: infection, immune dysfunction, aspiration, environmental exposures, autoimmune disease, pulmonary hypertension, and other
- Consultation with other subspecialists
Expert Guidelines: Diagnostic Approach

Clinical Practice Guidelines
AJRCCM, August 2013
Pulmonary Function Testing

• A series of breathing tests to determine how much, and how well, the lungs take in and expel air

• Exercise testing to determine exercise tolerance and oxygen levels during exercise

http://www.chp.edu/our-services/pulmonology/patient-procedures/infant-pulmonary-function-testing
Radiographic Imaging: Chest Xray

Courtesy of Geoff Kurland
Radiographic Imaging: Chest CT

- Challenges in children
- May need anesthesia
- Radiation considerations
- Some diagnostic
- Biopsy planning

http://www.childrenshospital.org
Bronchscopy

• Examination of airways to look for:
  – Structural abnormalities
  – Infection
  – Signs of Inflammation
  – Bleeding
  – Aspiration
  – Other
Genetic Testing

• Some chILD diseases have known genetic causes
• Various genetic tests on blood and other tissue to detect
  – Specific disorders
  – Panels
  – Whole exome/whole genome

Courtesy of Larry Nogee
Lung Biopsy

- Involvement may be patchy
- Try to avoid when sick with intercurrent infection or on therapy that may change results
- Thoracoscopic if possible
- Need to collapse the lung to obtain with scope
- Minimize risk

Courtesy of Craig Lillehei
Pathology Review

Courtesy of Sara Vargas

Dishop, PAIP 2010

Courtesy of Geoff Kurland

Dishop, PAIP 2010
ChILD Treatment

**Supportive Care**

- Oxygenation
- Ventilation
- **Nutritional support**
- Immunizations
- Avoid Smoking
- Bronchiectasis therapy
- Exercise therapies/pulmonary rehab
- Referral for transplant

**Anti-Inflammatory Therapies**

- Steroids
- Azithromycin
- IVIG
- Hydroxychloroquine
- Steroid sparing anti-inflammatories
- Rescue therapies
Respiratory Support

Breathing Support

- Supplemental oxygen
- Non-invasive Ventilation
  - CPAP
  - BiPAP
- Invasive Ventilation
  - Intubation
  - Tracheostomy
  - ECMO
  - Artificial lung devices

A Boston Children’s Hospital patient
Nutritional Support

• Children with ILD often work harder to breathe and need to consume more calories

• Nutritional support often needed:
  • High calorie foods
  • Fortified liquids
  • Caloric supplements
  • NG or NJ tubes
  • G or GJ tubes
  • TPN

• Difficulty swallowing/oral aversion can be present and requires expertise

A Boston Children’s Hospital patient
Other Supportive Care

• Prevent further damage to the lungs
  – Avoid infections as much as possible
  – Keep all vaccines up to date (this includes other family members)
  – No smoke exposure
• Monitor for problems in other organ systems

A Boston Children’s Hospital patient
Treatment Challenges

• Lack of randomized controlled trials
• Individual/network experience
• Based on adult treatment
• Treat primary and coexisting conditions
• Multi-disciplinary team needed
• More options needed with data to support use
Lung Transplant for ChILD

• ChILD 2nd most common reason for pediatric lung transplant
  – Pulmonary fibrosis, interstitial lung disease, surfactant abnormalities, non-lung transplant BO, & lung growth abnormalities

• Younger patient population of the pediatric transplant recipients

• Similar outcomes to other populations
  – May be less rejection (younger vs. disease)

*Ped Pulm 48:490*
ChILD Research Network

Because disorders are rare, collaboration between multiple centers and families is imperative

Interdisciplinary and Multi-Center Collaboration:
Improve care for children and families
Finding new treatments and ultimately cures
- Clinical Care
- Research
- Education
- Advocacy
ChILD Research Network

- Collaboration chILD Foundation
- Monthly phone meetings
- Conferences
- Patient data registry
- Multiple chILD publications
- Grants
- New research
- Educational materials/events
- Planning treatment/interventional studies

Courtesy of chILD Foundation
ChILD Partnership

• Families can help by:
  – Participating in chILD foundation activities
  – Supporting each other
  – Asking questions
  – Raising awareness
  – Enrolling in studies
  – Fundraising
  – Participate in the #CartwheelsForKids Challenge!

Research Network Family Member
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