ATS publishes clinical practice guideline on ILD in infancy

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The American Thoracic Society has released new clinical practice guidelines on the classification, evaluation and management of childhood interstitial lung disease (chILD) in infants. Childhood ILD includes a diverse group of rare lung diseases found in infants, children and teens that involve the interstitial tissues of the lung, which surround the air sacs (alveoli) in the lung and airways (breathing tubes). It is not known how many children have these disorders. Some types of chILD are caused by other diseases, while the cause is unclear in others, and prognosis varies by disease type.

The new guidelines appear in the August 1 *American Journal of Respiratory and Critical Care Medicine*.

"The characteristics and natural history of ILD differ in children and adults," said Geoffrey Kurland, MD, of the Division of Pediatric Pulmonology at the Children's Hospital of Pittsburgh and a member of the committee that drafted the guidelines. "These new guidelines specifically address the diagnostic evaluation and management of chILD in neonates and infants, as most of the new diagnostic entities that have been recently described affect infants disproportionately."

The guidelines' recommendations include the following:

- **Diagnosis:**
    - Once other possible causes of lung disease symptoms

have been eliminated, "chILD Syndrome" can be diagnosed if at least three of the following four criteria are present: (1) respiratory symptoms (cough, rapid and/or difficult breathing, or exercise intolerance), (2) respiratory signs (tachypnea, adventitious sounds, retractions, digital clubbing, failure to thrive, or respiratory failure), (3) hypoxemia, and (4) diffuse abnormalities on a chest radiograph or computed tomography (CT) scan.

- In patients with chILD Syndrome, diagnostic testing should be performed to determine the exact chILD diagnosis and echocardiography should be performed as part of the initial evaluation to rule out structural cardiovascular disease and pulmonary hypertension.
- If thin section CT is performed, the lowest radiation dose that provides adequate diagnostic information should be used.
- For neonates and infants with chILD Syndrome in whom other diagnostic investigations have not identified the precise chILD disease, or in whom there is clinical urgency to identify the precise chILD disease, surgical lung biopsy should be performed, preferably using video-assisted thoracoscopy rather than open thoracotomy.
- Appropriate genetic testing should be performed.

- **Management:**

  - As there have been no controlled trials of any therapeutic interventions in chILD Syndrome, management is based upon indirect evidence, case reports and clinical experience.
  - For infants with severe, life threatening chILD diseases, referral to a pediatric lung transplantation center after

discussion with the family is recommended.
    - Given the limited evidence of a beneficial effect on clinical outcomes and the well-known side effects of immunosuppressive medications, the decision about whether or not to initiate a trial of immunosuppressive therapy must be made on a case-by-case basis. Considerations include the severity of disease, rate of progression, prognosis without treatment, co-morbidities and family values and preferences.
    - All patients with chILD Syndrome should receive supportive and preventive care, including treatment of co-morbidities and prevention of infection.
    - Families of patients with chILD Syndrome should receive education and support from care providers.

- **Research priorities:**

    - Establish accurate incidence and prevalence rates of specific chILD diagnoses.
    - Determine the natural history and clinical phenotypes of specific chILD diagnoses and their relationships to adult pulmonary disease through international databases.
    - Further delineate the mechanisms underlying chILD diagnoses.
    - Conduct multicenter studies of protocol-driven diagnostic, therapeutic and quality approaches to chILD Syndrome to ascertain the optimal methods of clinical evaluation and management.
    - Create high-quality, accessible tissue repositories and biobanks to enhance research efforts
    - Promote common terminology for chILD diagnoses and their continued inclusion in future revisions of The International Classification of Diseases (ICD).

"As our understanding of the distinctions between adult and child ILD developed, the need for clinical practice guidelines on the diagnosis and management of chILD became clear," said Dr. Kurland. "These guidelines should help clinicians navigate the evaluation and management of children with this complex constellation of diseases."

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