Childhood Interstitial Lung Disease (chILD): shining a new light on childhood!

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Childhood interstitial and diffuse lung diseases (chILD) are a heterogeneous and diverse group of rare lung disorders presenting in childhood. Unfortunately, there are few therapies and poor understanding of the natural history for most disorders. Patients encounter high morbidity and mortality. Through the establishment of national research networks and registries such as chILD EU in the European Union, chILD Research Network in the USA and the chILDRANZ: children's interstitial lung disease research in Australia and New Zealand, progress has begun to better understand patients with chILD. The chILD EU group published two landmark manuscripts in *Thorax* demonstrating the value of these registries to further advance the care of these patients through evidence-based studies.

Defining acute exacerbations (AEs) and understanding their clinical impact and outcome is essential to moving chILD research forward. Seidl *et al* in their manuscript 'Acute Exacerbations in Children's Interstitial Lung Disease-A chILD-EU Cohort Study'[1] provides a definition for chILD AEs that can be used as standardised criteria in future natural history studies and clinical trials. This study demonstrates that AEs in chILD, similar to AEs in cystic fibrosis, asthma and pulmonary fibrosis, can have a significant and negative impact on clinical course and quality of life. This article also sheds light on the clinical spectrum and variability of responses to AEs based on underlying diagnosis with overrepresentation of patients with diffuse developmental and surfactant protein dysfunction disorders in the 81 patients who died during the observational period.

With the majority of AEs lasting longer than 2–3 weeks, one-third of children with an AE requiring admission, and death being associated with an AE in 60%, it is clear that chILD AEs are serious. The severity of chILD AEs delineates a need for prevention measures and the establishment of AE treatment protocols. There are no established treatment protocols for treatment of AEs in chILD, but some providers have used courses of glucocorticoids and antibiotics with only anecdotal evidence. Prevention with immunisations and even wearing masks are important considerations. Having a clear definition of an AE in chILD as provided by this study is useful to capture and monitor key natural history events and to define outcome measures for future AE treatment and prevention studies. This definition of AEs should be used in further chILD treatment protocols as a reduction of AEs given their significance would be a positive treatment outcome.

In addition to the significant health burden on children with chILD, the manuscript by Seidl *et al* 'Healthcare resource ultilization and medical costs for children with interstitial lung disease (chILD) in Europe'[2] illustrates the substantial financial and personal costs chILD creates for health systems and families across different chILD disorders. Medical costs, excluding drugs, constituted the largest category and were driven by hospitalisations and diagnostics. Major cost burdens were also born by the families through direct non-medical (travel and other care costs) and indirect costs (job productivity and financial loss). Though medical costs dropped significantly after the first year, the costs for families persisted. The magnitude of persistent financial losses are even greater when they are sustained early in the life of a child and their young family, creating the potential for lifelong earning impact. Medical costs differed by country. Patients from the USA were not part of this study. Costs for chILD would have been expected to have been even higher as traditionally US medical costs in 2018 were nearly twice other comparable countries.[3]

The health and financial impacts of chILD reported by the EU chILD collaborative are a call to action for countries, insurers and funding agencies to do more to study new treatments and care paradigms. Fortunately, there is a roadmap to accomplish this as chILD is coming of age with new opportunities and organisational structure around the world.[4] The first world-wide study in chILD fibrosis was successfully completed with a new antifibrotic, nintedanib[5] and more promising drug treatments for chILD hopefully will follow. The European Medicines Agency (EMA) and Food and Drug Administration (FDA) should require paediatric trials when companies are seeking approval for drugs used for conditions in adults that could also be beneficial in chILD disorders. New genetic insights, mechanisms for intervention, diagnostics and registry study potential are only limited by prioritising funding opportunities. We applaud the EU chILD Collaborative for shining new light on chILD. We have more to do to stop robbing childhood from children and families who are living with chILD.

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