

International chILD survey 2017

Introduction

The aim of the survey was to find out more about the challenges that parents and caregivers of infants/children/teenagers diagnosed with interstitial lung disease (chILD) face and to highlight these to healthcare professionals so as to inform future practice, activities and research.

The survey was developed with members of the European Lung Foundation (ELF) chILD patient advisory group and made available in the following languages: English, French, German, Italian, Spanish, Polish, Portuguese, Greek and Russian. The survey was available online for three months between August to October 2017.

The survey findings were presented at the ENTeR-chILD (European network for translational research in children's and adult interstitial lung disease) Inaugural Conference, COST Action in Prague at the beginning of November 2017.

A summary of the core findings is on page 2 followed by a more detailed report on pages 3-11 which shows some of the variations and more detailed examples provided by the survey respondents.

We hope that these survey findings, although not telling the whole story, will help to deepen understanding and demonstrate a range of views and experiences. Thank you to all the individuals who took the time to answer the questions and share their experiences.

A number of quotes from survey respondents have been used throughout the document to illustrate the themes and have been translated from the original source language into English.

Summary of findings

Main themes

Three key over-arching themes emerged from analysis of both the survey statistics and open comments to highlight the most challenging aspects of living with chILD:

1. Lack of information about disease progression and what the future holds.
2. Lack of knowledge and consistency of approach across the medical profession.
3. Lack of support in practical day-to-day living and in dealing with anxiety and uncertainty.

Most difficult aspects to manage:

- Unknown factors about progression and prognosis.
- Lack of knowledge from healthcare professionals.
- Lack of research into chILD.
- Feeding and nutritional issues.
- Oxygen use practicalities and breathing difficulties.
- Managing everyday sickness and risk of infections.
- Child being 'different' from their peers and resulting issues.
- Living with stress/anxiety and difficulties of life revolving around medical care.

"I cannot express enough the anxiety of having a NEHI baby" (UK)

Information needs:

- About the condition, treatment options and support.
- Tips on how to manage daily living and challenges.
- Targeted information for individuals with chILD (at all ages), siblings and other family members and education providers.
- How to find chILD medical experts and get second opinion.
- Best treatment plans for conditions.
- Other people's experiences - especially success stories.

"We as parents feel helpless because we have no information about the expected illness of our child - there is a lack of information all along the line" (Undiagnosed, Germany)

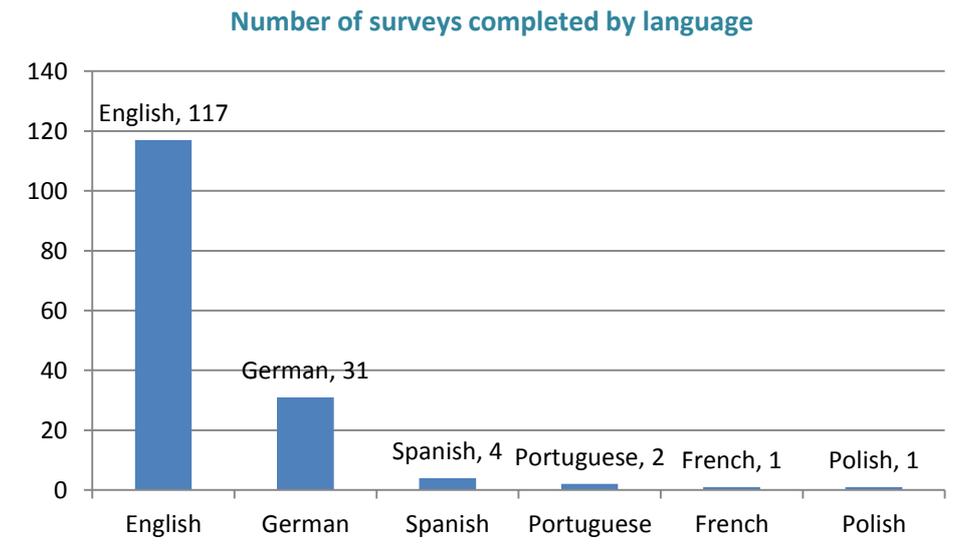
Research is needed into:

- Causes including genetics, environmental and other risk factors.
- How the conditions will progress
- Less invasive diagnostic techniques
- Current and potential new treatments
- Feeding issues
- Reducing impact on bodies over the longer-term

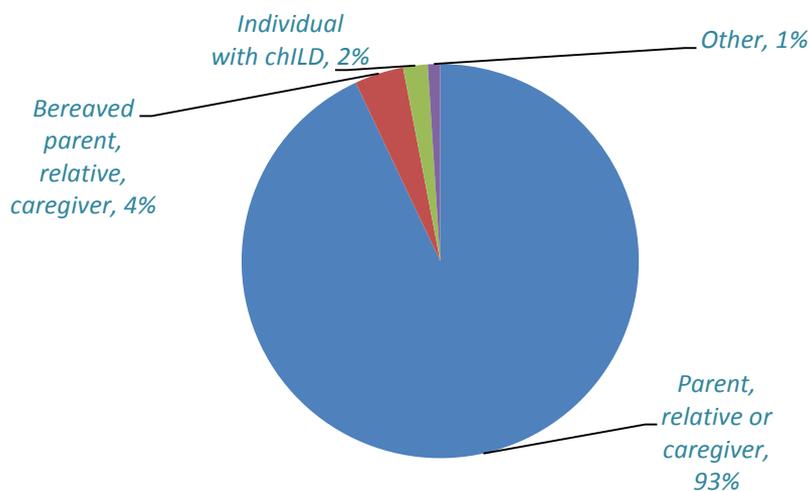
Detailed findings

A. Survey respondents

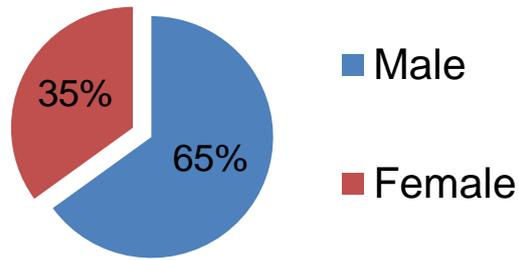
- 156 surveys were completed as shown below:



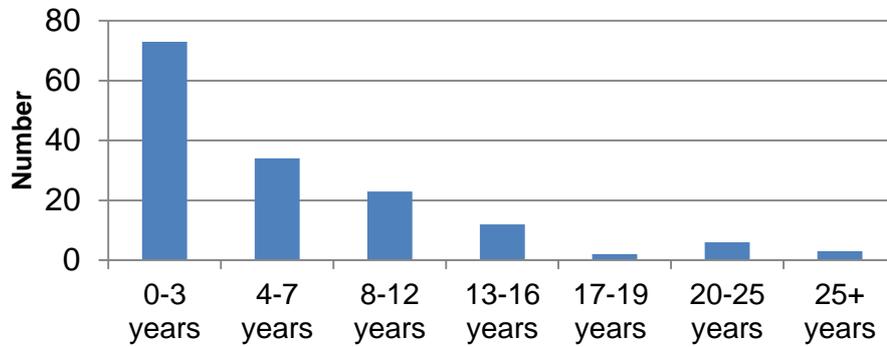
- Respondents were resident in the following 13 countries/areas: Australia, Bulgaria, Canada, Estonia, France, Germany, Ireland, Poland, Portugal, Spain, UK, USA and Venezuela.
- 93% of respondents identified themselves as a parent, relative or caregiver with the remaining 7% identified as shown in the chart below:



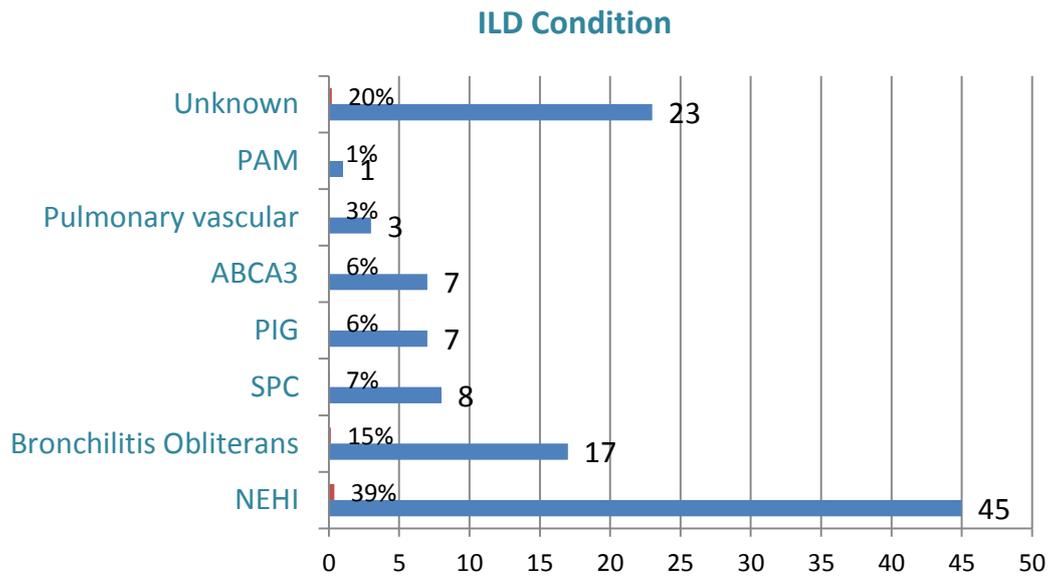
- More of the respondents (65%) had a child with ILD that was male:



- The age ranges of the child with ILD was mainly between the ages of 0 to 7:

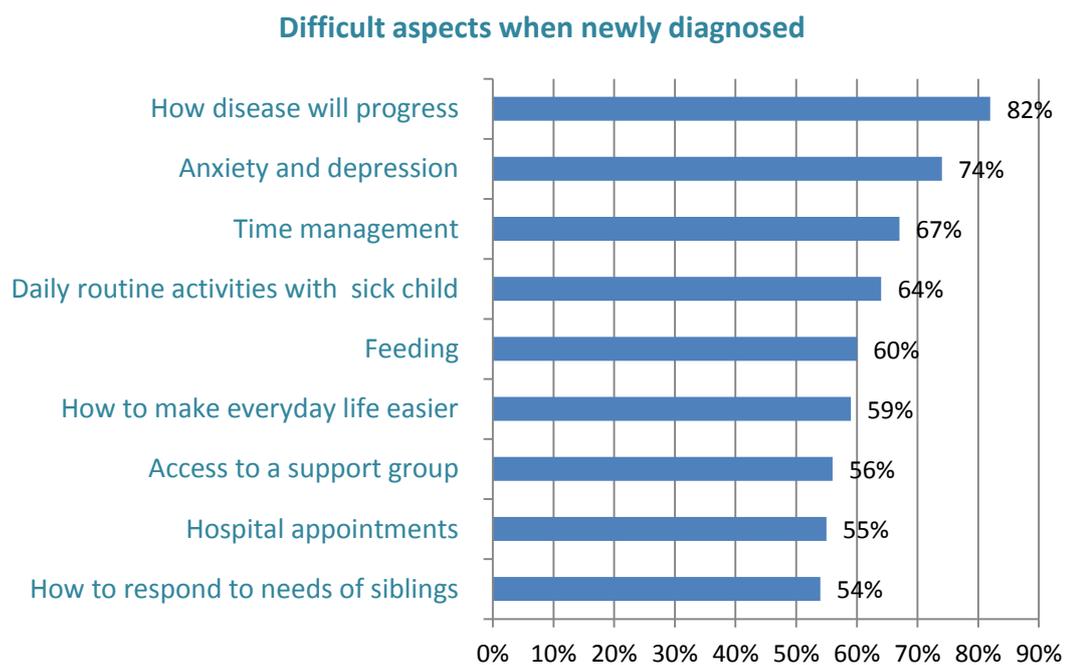


- There was a range of child conditions as shown below with NEHI being the most frequently cited; and one fifth of respondents had no diagnosis:



B. Testing and diagnosis

- Respondents were asked to rate how difficult certain aspects of living with chILD were when their child was newly diagnosed using a 5 point rating scale. All aspects shown in the chart below were rated as either 'Difficult' or 'Very difficult' by over half of all respondents:



(The % is the combined scores of those who ranked as either 'Difficult' or 'Very Difficult')

Suggestions to make the testing and diagnostic period less challenging:

- Tips for dealing with equipment, and especially oxygen, at home.
- Information for family and friends to help them understand.
- More multidisciplinary care.
- Hospitals to be better at dealing with long-term stays.
- Help to balance hospital appointments/life with paid work.

*“The first year was the most difficult time in my entire life”
(Bronchiolitis Obliterans, UK)*

- Respondents were asked to rate how important the aspects below were to them during diagnosis and testing stage for children with ILD, using a 5 point scale. Each of these aspects was rated between 97% and 100% (combined scores of those who ranked them as either 'Important or 'Very Important'):

| | |
|---|------|
| - Support from healthcare professionals | 100% |
| - Tips and advice on healthcare management | 100% |
| - More awareness of disease among paediatricians and other healthcare professionals | 99% |
| - More awareness of the disease among local physicians | 98% |
| - Recognition of disease at an early stage | 98% |
| - Written information | 97% |

Additional comments made about what is important during the diagnosis and testing stage:

- Multidisciplinary care with social work and/or psychology as standard.
- Education for the child and their siblings.
- National care programme/pathway when chILD is suspected.
- Help from healthcare professionals in understanding the benefits versus the risk and pain of testing.
- Greater awareness among hospital emergency staff.
- More information on co-morbidities (additional diseases).

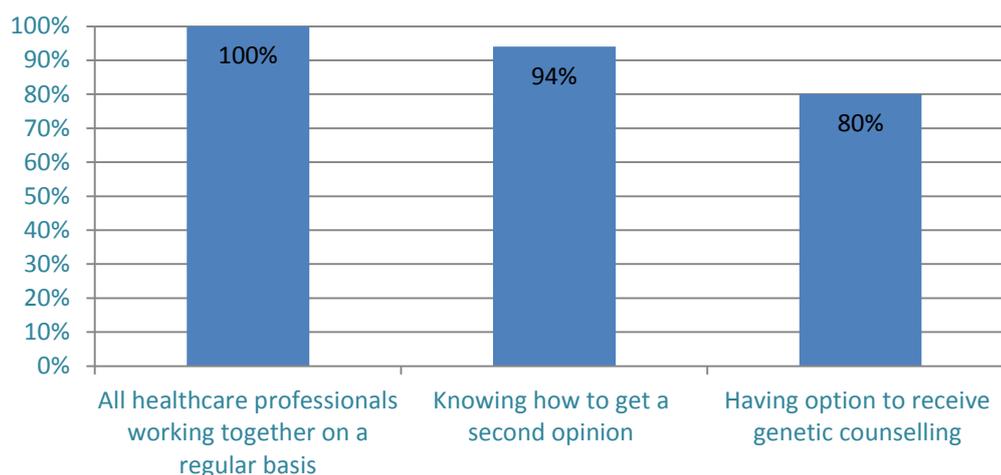
C. Treatment and care

- A wide range of challenges were identified by respondents when accessing both healthcare and emergency care including:
 - Difficulty in finding an expert for chILD in own region.
 - Lack of knowledge among both local healthcare staff and emergency room professionals; therefore having to continually explain about the condition.
 - Plan of action not shared with emergency room professionals.
 - Experience as a parent not viewed as important.
 - Having to drive the inquiry into diagnosis.
 - Lack of co-ordination between healthcare staff.
 - Lack of access to services e.g. psychology, respite care, feeding.
 - Dealing with oxygen-related issues.
 - Health insurance issues.

“I cannot express enough the anxiety of having a NEHI baby”
(NEHI, UK)

- Respondents were asked to rate how important the aspects shown in the chart below, were in chILD treatment and care: 100% felt that ‘All healthcare professionals working together on a regular basis’ was the most important:

How important are these 3 aspects of treatment and care



[The % is the combined scores of those who ranked aspects as either ‘Important or Very Important on a 5 point rating scale]

Respondents also said that the following were important for treatment/care:

- A network of specialist doctors and greater collaboration between hospitals.
 - Information on available treatments and their effectiveness.
 - Nutritional counselling and feeding issues.
 - Practical advice on everyday life e.g. tips on how to make navigating on oxygen easier for new patients.
 - Helping parents to connect with each other.
 - Reimbursement issues.
- **The aspects that respondents identified as most difficult were:**
 - The unknowns about how the condition will progress, how it will affect their child, longer-term prognosis and future prospects.
 - Management of time and life which revolves around medical care (including medication, doctor/hospital appointments, treatment).
 - Feeding, diet and nutritional issues.

- Oxygen use and the management of breathing difficulties.
- Managing sickness and infections.
- Living with the stress and anxiety of the uncertainty and challenges.
- Awareness of child being 'different' from their peers; especially as they get older (limitations and keeping up with peers, depression, lack of information for teenagers, social activities).
- School/education information and support lacking.
- Lack of research around long-term treatments and that there is no cure, including lung transplantation survival statistics.

D. Information needs

“We as parents feel helpless, because we have no information about the expected illness of our child..... There is a lack of information all along the line” (Undiagnosed, German)

- Respondents wanted to receive information about:

- | | |
|--|------|
| - The condition | 100% |
| - All treatment options available | 99% |
| - Support available (locally/nationally) | 99% |

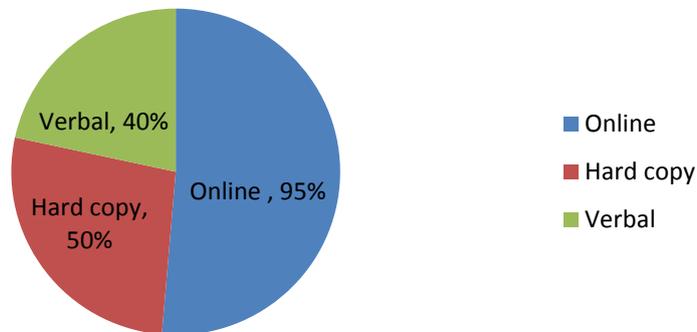
Other types of information that respondents would like to receive:

- Latest research results.
- How to integrate child into school/social environment/family.
- Progression of the disease and different phases.
- chILD events and conferences.
- Details of specialists.
- Financial support available.
- Tips and tricks on dealing with the condition every day.
- How to deal with education providers.
- Children and teenager information targeted for their needs.
- Case studies (especially success stories).

“Information eases decision making and uncertainty” (NEHI, UK)

- Survey respondents wanted information in a range of formats: online, verbal and hard copy e.g. leaflets; 95% wanted it online.

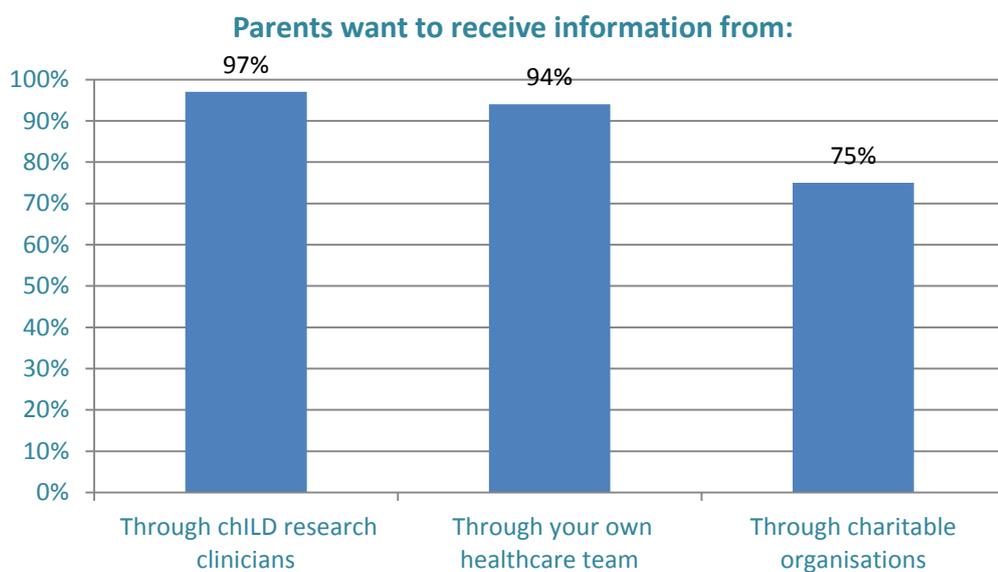
How information should be made available



*“Of course you cannot do without the personal treatment and that we know each other because that can never be replaced by online”
(Undiagnosed, Spain)*

*“Follow-up email regarding specific case, diagnosis, next steps”
(NEHI, USA)*

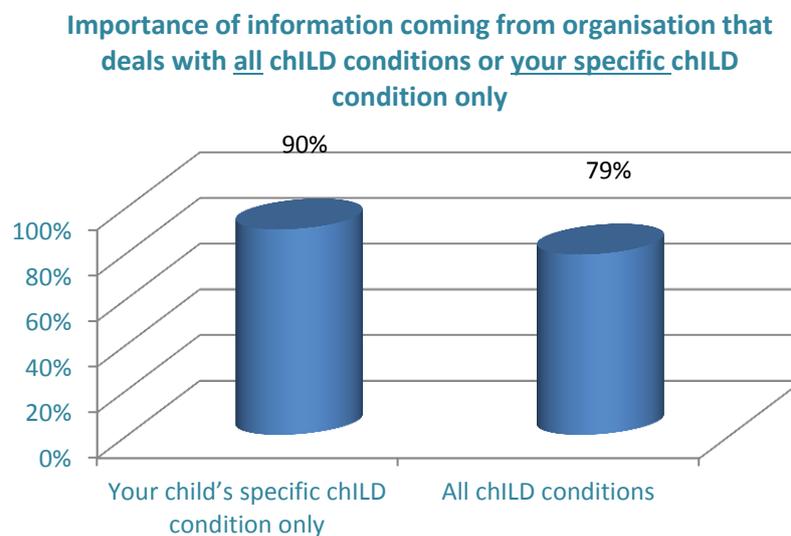
- The survey asked how important it was for parents to have information presented by different people/organisations. Respondents felt it was important to receive information from **all** of these sources with 97% wanting to receive information from chILD research clinicians:



“Medical information should come from referral specialists. Patient associations have a social and supportive purpose for families...There is an urgent need in France for specialists to provide information to families” (Undiagnosed, France)

“This is a very lonely disease. The lack of information and community is very isolating” (PIG, USA)

- The survey asked how important it was to get information /support from an organisation that deals with either your specific chILD condition; or all chILD conditions:



Further comments:

“I would like to be able to speak to other people familiar with my son's disease who are also sharing the same experiences” (USA; PIG)

“As long as the information is accurate, it doesn't matter to me whether the organization deals with all lung diseases or just my child's” (Bronchiolitis Obliterans, USA)

“Both our condition and ILD generally are important, to help understand comorbidities and care options which are common.” (NEHI, Australia)

E. Research

- **Research areas that would most help their own child and/or future generations were identified as follows:**
 - Causes
 - Progression of conditions prognosis
 - Genetics and epigenetics
 - Pregnancy (risk factors, environmental exposures)
 - Diagnostics (lung biopsy specimens, less invasive techniques)
 - Current treatment options (e.g. oxygen need, medication, lung transplantation, long-term effects)
 - New treatments (e.g. stem cells)
 - Feeding issues and why this affects some and not others.
 - Reducing impact over the long-term (e.g. minimising fibrosis, heart implications)
 - Behavioural links.
 - Relationship between bronchiolitis obliterans and solid organ transplants

Respondents also wanted to see long-term studies, more consistent diagnostic tools, best treatment plans, translational global research, more training for paediatricians and the identification of more chILD diseases.

“I have only met one doctor (who diagnosed her) who knows more about my daughter’s condition than me.” (Undiagnosed, UK)

For further information about the survey please email Jeanette Boyd, ELF patient projects at jeanette.boyd@europeanlung.org.