Childhood Interstitial Lung Disease: Family Experiences

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Summary. Aims: To present experiences of UK families of children diagnosed with ILD, to inform clinical practice and service development. Methods: Thirty seven such families completed an anonymous web-based survey between February and March 2014. Results: Median time from first symptoms to diagnosis was 25 weeks. Most reported that they were happy with the overall management of their child. Areas highlighted for development included improved communication especially the need for written information; psychological support (91% reported anxiety). Feeding issues (not described in current literature) were reported by 77% and persisted in 35%. Other requests included better written communication between hospitals with training for smaller hospitals, and improved specialist nurse support for children with ILD. Conclusions: These family perspectives need to be addressed by professionals looking after these children as well as when planning of future services. **Pediatr Pulmonol.** © 2015 Wiley Periodicals, Inc.

Key words: interstitial lung disease; gastrostomy; anxiety; psychological support.

Funding source: European Union's Seventh Framework Programme; Number: FP7/2007–2013.

INTRODUCTION

Interstitial lung diseases (ILD) in children are rare and disparate.[1,2] As a consequence, even in large centers clinicians will have seen only a handful of cases. We considered that clinicians may be able to provide better care to parents and children with ILD if they have a broader understanding of parent experiences and perspectives. Our aim was to present the experience of parents of children diagnosed with interstitial lung disease in order to inform current clinical practice, and future planning of health care.

METHODS

Between February 2014 and March 2014, families with children affected with ILD completed a 118 question web-based survey. Participants were recruited through the family support forum and internet mailing list. The questionnaire was developed by the chILD Lung Foundation and piloted on three volunteer families. The survey consisted of mainly closed questions, with some open qualitative questions. Participation criteria were: family based in the UK; parents of child with a diagnosis of ILD.[1] Responses were collected anonymously; however, the Internet Protocol (IP) address of the participant was checked to ensure that the survey was limited to UK participants. We used month/year of birth and diagnosis to ensure that only one survey per family was completed. The project was funded by chILD Lung Foundation (http://childlungfoundation.org/). Ethical approval was thought not to be required for this parent conducted voluntary survey of parents within a patient organization. Access to the survey can be found at https://www.surveymonkey.com/s/N8WQV3B, and the questions and number of responses to the survey is available as online supporting material.

RESULTS

Family Data

Forty three participants accessed the online survey, of whom six were excluded (diagnosis chronic lung disease of prematurity n = 2; non-UK residence, n = 1; incomplete questionnaires, n = 3). Data is reported for the remaining 37 completed questionnaires. Seventy percent [26/37] of children diagnosed were male, with the

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Conflict of interest: None.

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Received 14 October 2014; Revised 25 January 2015; Accepted 30 January 2015.

DOI 10.1002/ppul.23168
Published online in Wiley Online Library (wileyonlinelibrary.com).

majority (89%) born at term (≥37 weeks) and only 11% (n = 4) preterm.

The reported diagnoses were unknown ILD 38% (n = 14), neuroendocrine cell hyperplasia of infancy (NEHI) 16% (n = 6), *ABCA3* mutations 8% (n = 3), obliterative bronchiolitis (OB) 24% (n = 9), follicular bronchiolitis 3% (n = 1), pulmonary interstitial glycogenosis (PIG) 3% (n = 1), surfactant protein C mutations (*SP-C*) 5% (n = 2), and chronic bronchiolitis 3% (n = 1). There was a family history of respiratory disease in 51% (n = 19) and of autoimmune disease in 19% (n = 7).

Presentation of ILD

At birth, 9 of the 37 children had been admitted to a neonatal unit, where five required treatment for respiratory distress (*ABCA3* (n = 2), *SP-C* (n = 1), PIG (n = 1), unknown ILD (n = 1)). The other four children were admitted for antibiotic treatment of Streptococcus B infection (n = 2; both unknown ILD), meconium aspiration (n = 1; NEHI) and antibiotics due to delivery at 34 weeks via caesarean section (n = 1; OB).

The reported median age of first symptoms was 10 weeks (range 0–22 months). At time of first presentation, children had a broad range of symptoms (Table 1), but respiratory symptoms were most common: 51% (n = 17) of children had a cough, upper respiratory or other infection and 41% (n = 15) had increased work of breathing. The mean number of symptoms per child was 2 (range 1–4).

The initial treatment included oxygen (n = 27; 74%), antibiotics (n = 11; 29%), intubation and mechanical ventilation (n = 6; 17%). Ventilation was instituted at birth (n = 4); in a further two cases with OB, intubation was at 8 and 11 weeks. In four other cases (11%) parents were provided with reassurance that there was nothing wrong with child by medical staff at initial review. Most parents were happy about their child's current status (68%, n = 25 responses).

TABLE 1—Signs and Symptoms Reported at First Presentation of ILD (n = 35)[1]

Sign and symptom	No. reported
Central and peripheral cyanosis	5
Respiratory tract infection	17
Failure to thrive	4
Feeding issue	10
Grunting	4
Low oxygen saturations	3
Hypothermia	1
Intercostal retractions	6
Lethargy	2
Pale Skin	2
Sweating	1
Tachypnoea	15
Wheezing	2

[1]Some participants reported more than one sign or symptom.

Diagnostic Delays

The median age at diagnosis of ILD was 35 weeks (range 1 week to 8 years). The median time from first reported symptoms to diagnosis was 25 weeks (range 1 week to 8 years). As expected CT scan (78%; n = 29), combined with either bronchoscopy (22%; n= 8) or lung biopsy (11%; n = 4) or all three procedures (19%; n = 7) were the main diagnostic tests. Fifty three percent (n = 19) parents could not recall receiving any written information on ILD or the child's disease type. Eleven percent (n = 4) of initial ILD diagnoses were changed subsequently.

Feeding Issues

Feeding issues were reported in 73% (n = 28). Over half (54%, n = 19) were prescribed reflux medication and 43% (n = 16) required nasogastric tube calorie support, progressing to gastrostomy in 11. Persistence of feeding problems were common and present in 35% (n = 13), mostly long-term gastrostomy dependency (22%; n = 8) and oral aversion in 14% (n = 5).

Communication and the Medical Team

As may be expected, a large number of healthcare professionals provided input to the child (mean 8, range 2–14). The reported best relationships between the families and clinical team were with the respiratory consultants and physiotherapists, and 70% (n= 26) of participants reported that they were very happy/happy with the overall management of their child. A care plan/treatment strategy was provided by a respiratory consultant in approximately half of cases (51%; n = 19). Disappointingly, over half of families considered they had been provided with no information on future prognosis (57%; n = 21).

Family Support Systems

Participants reported that they felt most supported by patient organizations, other parents affected by ILD, and their clinical team. Support was perceived to be less strong from friends and primary care. The most prevalent concern for families was the potential for future deterioration (81%, n = 30). Significant anxiety (52%, n = 17) and moderate anxiety (39%, (n = 13) were common. Psychological support was reported as offered to only seven families.

Participant's Comments

Participants were given the opportunity to give their opinion on how they would like to see ILD services develop. Common health services themes included: more

proactive psychological input, written summaries of disease specific discussions from healthcare professionals (including prognosis), help talking to the child about ILD, better written communication between shared care hospitals and training for smaller hospitals, and better specialist nurse support of children with ILD (often present for those oxygen but not those managed in air (n = 14)).

DISCUSSION

This is the first opportunity for families of children diagnosed with ILD to discuss their needs; this is important for professionals looking after these children as well as for the planning of future services. The salient features are (i) the high prevalence of feeding issues is highlighted; this is not described in the ILD literature[3,4]; (ii) the need for written information at diagnosis and psychological support throughout; (iii) ongoing psychological support around feeding issues; and (iv) the need for specialized nursing support for these families.

There are of course weaknesses in this study. It is retrospective, there was no objective verification of the accounts of the parents, and it is likely that this represents a highly selected sample of the families. There are approximately 70 families known to the group, but we cannot exclude the possibility that others unknown to us may have found the questionnaire and completed it. Furthermore, the questionnaire is not a validated instrument, and the nature and number of the questions may have influenced the answers. We also have no objective verification of the answers; so parents may in fact have received information but just forgotten it. However, if this was the case, it still emphasizes the need for improved information sharing with these families. Nonetheless, we believe it provides useful data to inform current and future practice. In particular, psychological support should at least be offered to all families, and if at all possible, a respiratory nurse should be involved to give ongoing support, albeit that the nurse may have to learn about the particular ILD with the family, given the wide spectrum of rare conditions.[1] Finally, there is a need for more fact sheets; one of the outputs of the current FP-7 chILD grant[5] will be better patient information leaflets.

ACKNOWLEDGMENTS

The research leading to these results has received funding from the [European Union's] Seventh Framework Programme ([FP7/2007–2013]) under grant agreement n° [305653] (Dr. Steve Cunningham, Prof. Andrew Bush). Prof. Bush was also supported by the NIHR Respiratory Disease Biomedical Research Unit at the Royal Brompton and Harefield NHS Foundation Trust and Imperial College London.

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Supporting Information

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