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# Epidemiology of childhood interstitial lung disease (chILD) in France : The RespiRare cohort

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## Introduction

**Childhood interstitial lung disease (chILD)** is a heterogeneous group of rare lung diseases that share common radiological features of interstitial signs on chest X-ray or chest computed tomography (CT)-scan.

chILD can present throughout life, from birth to adolescence, **with important variations in severity from pauci-symptomatic diseases to end-stage respiratory failure leading to pulmonary transplantation or death.** chILD classification usually distinguish two age groups: below 2 years and 2 to 18 years.

Limited number of patient series with chILD have been reported worldwide by single centers or large networks (Table 1).

**The present study aims** at describing the epidemiology of chILD in France in terms of prevalence, incidence, respective frequencies of the different etiologies of ILD and age of onset.

Table 1: chILD series reported to date

Country / Area	Patients (n)	Period of time (years)	References
United Kingdom and Ireland	46	3	Dinwiddie et al. 2002
Denmark	884	10	Kornum et al. 2008
Germany	56	1	Griese et al. 2009
France	205	4	Nathan et al. 2012
Australia and New Zealand	71	5	Casamento et al. 2016
USA	93	18	Soaers et al. 2013
China	133	5	Tang et al. 2020
Europe and above	575	1	Young et al. 2018
Spain	381	2	Torrent-Vernetta et al. 2022

## Methods

### Type of study

- Observational
- Retrospective (2000 - 2022)
- Prospective (Feb 2022-Feb 2023)
- National, multicentric
- Legal issue: ethics committee gave their authorization

### Inclusion criteria

- Patients with ILD defined following the ATS Clinical Practice Guideline
- Born between 2000 to 2022 and aged under 18-year-old at diagnosis

### Exclusion criteria

- Bronchopulmonary dysplasia,
- No available data

### Data collection

- e-Respirare
- Multidisciplinary team meetings (MDT) for chILD
- Direct emails to the centres

### Localisation of the study

All the RespiRare centres in France

## Results

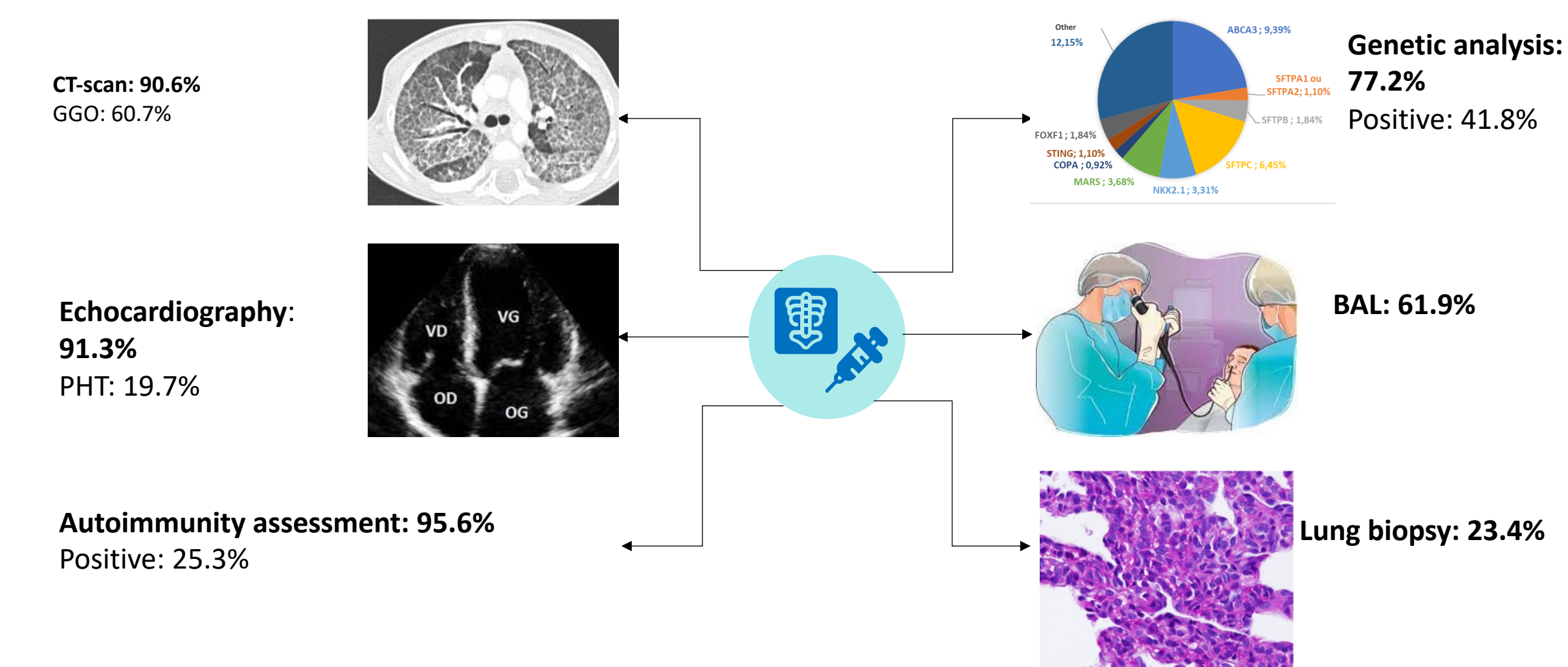
### A total of 790 patients were included

Median age at diagnosis: 3 m      Sex ratio: 1  
Premature birth :17.7%      Family history of ILD: 16.9%

**The prevalence** of chILD in 2022 was 44.04/million of children

**The incidence** of chILD in 1 year was 4.44/million of children

### Investigations performed:



### chILD etiologies

**Below 2 years**, the main ILD etiologies were surfactant metabolism disorders and proteinosis alveolar (23.7%) and NEHI (11.8%).

**Between 2 and 18 years**, the main etiological diagnoses were alveolar hemorrhage and pulmonary hemosiderosis (12.2%), connective tissue diseases (11.4%), hypersensitivity pneumonitis (8.8%), sarcoidosis (8.8%) and vasculitis (7%).

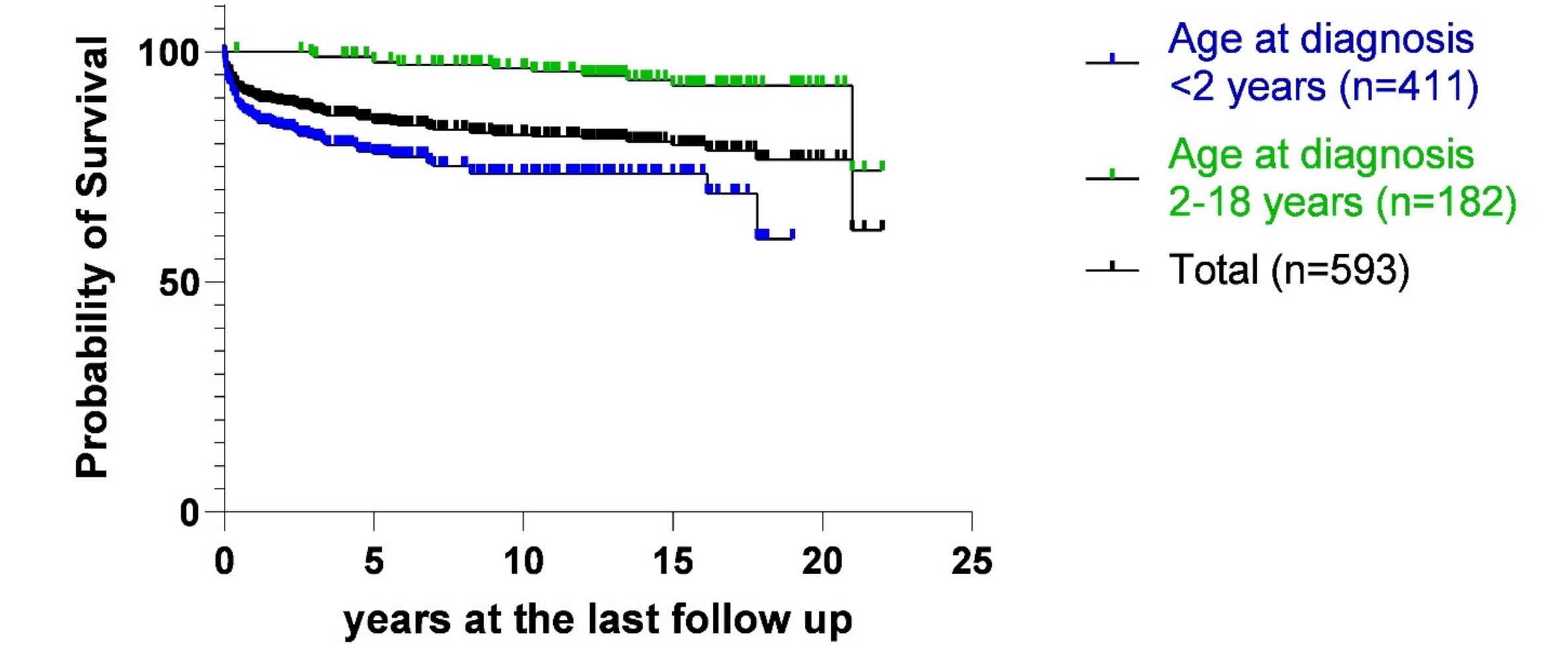
## Results

### Evolution

**At follow up:** Deceased patients: 17.7%

**5 years survival (Figure 1):**

- For patients diagnosed < 2 years: 57.3%
- For patients diagnosed > 2 years: 86%



Kaplan Meier survival according to the age at diagnosis.

## Discussion and conclusion

This large epidemiological study of chILD confirms a **higher incidence and prevalence than previously reported.** Our findings are similar to the 2022 Spanish study.

The next challenge will be to take advantage of well-organized international networks such as the CRC chILD and the ERN-lung to develop systematic cohorts and registers – at least for some chILD etiologies - to develop and test new therapeutic options and to prepare new guidelines for the management and transition of care of these patients.

