Assessment of fibrosis in lung biopsies from the European childhood interstitial lung disease (chILD) registry

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BACKGROUND

- Childhood interstitial lung disease (chILD) comprises >200 rare respiratory disorders[1]
- There is some overlap with entities observed in adults but also many differences[2]
  - Many are exclusive to infants and children
  - Idiopathic pulmonary fibrosis is exclusive to adults
- There is little data on pulmonary fibrosis in chILD

- European chILD registry (chILD-EU; NCT02852928)[3]
  - International network of expert centres specialised in chILD
  - Cooperatively assesses and follows patients with chILD
  - Broad spectrum of patients are classified into disease categories[1]

AIMS

- To assess the prevalence of fibrosis in lung biopsies from chILD-EU
- To correlate paediatric lung fibrosis with radiological findings

METHODS

Chest CT

- Scored by paediatric radiologist
- Fibrosis features:
  - linear or reticular opacities
  - traction bronchiectasis
  - honeycombing
  - architectural distortion

Lung biopsies

- Scored by paediatric pathologist
- Fibrosis features:
  - interstitial fibrosis
  - fibroblastic foci
  - honeycombing
  - histological diagnosis of CPI

RESULTS

PRESENCE OF FIBROSIS IN BIOPSIES

825 chILD cases

149 Lung biopsies

426 CT scans

60 Biopsies with features characteristic of fibrosis

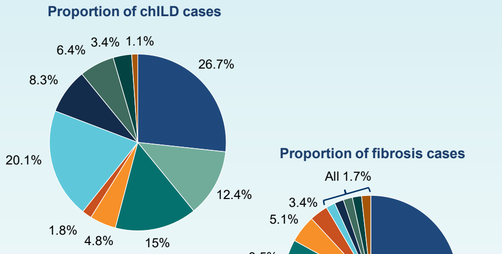
172 CT scans had signs of fibrosis

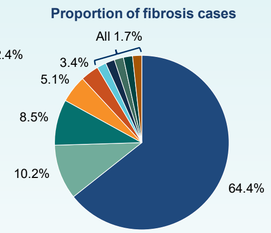
- Mean age at biopsy: 6.4 years (range 0–26 years)
- 40% (60/149) of biopsied patients had signs of fibrosis on their histology

DISEASE CATEGORIES AND FIBROSIS IN BIOPSY

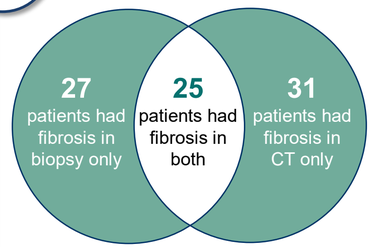
- Approximately half of the patients had conditions that primarily manifested during infancy, either surfactant dysfunction disorders or persistent tachypnoea of infancy (NEHI, an infant condition of undefined aetiology)
- Fibrosis was predominantly observed in disorders of the alveolar surfactant region, accounting for more than half of cases

Disease category	Proportion of chILD cases (%)	Proportion of fibrosis cases (%)
Alveolar surfactant region	26.7	64.4
Systemic disease processes	12.4	10.2
Exposures	15	8.5
Immunocompromised	4.8	5.1
RDS in mature neonate	1.8	3.4
Infant conditions of undefined aetiology	20.1	1.7
Lung vessels	8.3	1.7
Lung growth abnormalities	6.4	1.7
Diffuse developmental disorders	3.4	1.7
Lymphoid lesions	1.1	1.7





HISTOLOGY AND THE CORRESPONDING CT SCAN



83 pairs of biopsies and CT scans

- Time difference between CT and biopsy (all biopsy pairs): –9 days
  - Ten patients had >1 year difference (5/10 patients had fibrosis)
- Time difference between CT and biopsy (only patients with <1 year difference): –14 days

30% had fibrosis features on both CT and biopsy (25/83)

- Correlation between fibrosis in biopsy and CT was lower than expected
- Possible reasons include:
  - additional pairs of biopsy/CT required (data will become available)
  - grade of severity of fibrosis not considered
  - localisation of biopsy on CT scan not known

CONCLUSIONS

- Features of fibrosis were present in about 40% of chILD cases with a diagnostic lung biopsy
- Concordance between biopsy- and CT-diagnosed fibrosis was lower than expected
- Further studies may help to evaluate the use of biopsy in the diagnosis of fibrosis in chILD

INTERACTIVE

https://www.globalmedcomms.com/respiratory/ERS2020

https://www.globalmedcomms.com/respiratory/ERS2020/Griese

References

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Abbreviations

chILD, childhood interstitial lung disease; chILD-EU, European chILD registry; CPI, chronic pneumonitis of infancy; CT, computed tomography; NEHI, neuroendocrine cell hyperplasia of infancy; RDS, respiratory distress syndrome.

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