



Clinical problems in rare interstitial lung diseases

Rare ILD/DPLD

PA-67373 - Epidemiology of diffuse interstitial lung disease in children in France: The RespiRare cohort

CAMILLE FLETCHER¹, Alice Hadchouel Duvergne², Caroline Thumerelle³, Manon Fleury⁴, Nouha Jedidi⁴, Myriam Benhamida⁵, Katia Bessaci⁶, Raphael Borie⁷, Tiphaine Billhouee⁸, Jacques Brouard⁹, Aurelie Cantais¹⁰, Annick Clément¹¹, Laurianne Coutier¹², Pierrick Cros¹³, Marie Laure Dalphin¹⁴, Christophe Delacourt², Ralph Epaud¹⁵, Eric Deneuille¹⁶, Antoine Deschildre¹⁷, Jean-Christophe Dubus¹⁸, Carole Egron¹⁹, Michael Fayon²⁰, Agnes Fina²¹, Aude Forgeron²², Elsa Gachelin²³, François Galode²⁴, Isabelle Gibertini²⁵, Lisa Giovannini-Chami²⁶, Tamazoust Guiddir²⁷, Audrey Herzog²⁸, Veronique Houdouin²⁹, Eglantine Hullo³⁰, Pierre-Henri Jarreau³¹, Geraldine Labouret³², Alice Ladaurade³³, Laurence Le Clainche Viala³⁴, Christophe Marguet³⁵, Alexandra Masson-Rouchaud³⁶, Julie Mazenq¹⁸, Caroline Perisson³⁷, Cinthia Rames³⁸, Philippe Reix³⁹, Marie-Catherine Renoux⁴⁰, Léa Roditis³², Cyril Schweitzer⁴¹, Aurélie Tatopoulos⁴², Pascale Trioche-Eberschweiler⁴³, Françoise Troussier⁴⁴, Clementine Vigier⁴⁵, Laurence Weiss²⁸, Marie Legendre⁴⁶, Camille Louvrier⁴⁶, Alix De Becdelievre⁴⁷, Aurore Coulomb⁴⁸, Chiara Siléo⁴⁹, Hubert Ducou Le Pointe⁴⁹, Laureline Bertheloot⁵⁰, Celine Delestrain⁵¹, Nadia Nathan¹¹

(1) Hopital Armand Trousseau, Paris, France; (2) "1 AP-HP, Hôpital Universitaire Necker-Enfants Malades, Service de Pneumologie Pédiatrique, Centre de Référence pour les Maladies Respiratoires Rares de l'Enfant, Paris, France 2 Université Paris Cité, Paris, France 3 INSERM U1151, Institut Necker Enfants Malades, Paris, France", Paris, France; (3) France, Lille, C.H.U de Lille, Pneumologie pédiatrique, Lille, France; (4) France, Paris, Hôpital Armand Trousseau AP-HP, Pneumologie pédiatrique, Paris, France; (5) France, Nantes, Centre hospitalier universitaire de Nantes, Pneumologie pédiatrique, Nantes, France; (6) France, Reims, Hôpital Robert Debré (CHU de Reims), Pneumologie pediatrique, Reims, France; (7) France, Paris, Hôpital Bichat - Claude Bernard, Service de Pneumologie A Centre constitutif du centre de référence des maladies pulmonaires rare, Paris, France; (8) France, Nantes, Centre hospitalier universitaire de Nantes, Pneumologie pediatrique, Nantes, France; (9) France, Caen, C.H.U de Caen Normandie, Pneumologie pediatrique, Caen, France; (10) France, Saint-Étienne, CHU Hôpital de Bellevue, Pneumologie pediatrique, St Etienne, France; (11) France, Paris, Hôpital Armand Trousseau AP-HP, Pneumologie pediatrique, Paris, France; (12) France, Saint-Genis-Laval, Hôpital Lyon Sud secteur 3, Pneumologie pediatrique, Lyon, France; (13) France, Brest, C.H. Régional Et Universitaire CHRU, Pneumologie, Brest, France; (14) France, Besançon, Centre hospitalier universitaire de Besançon, Pneumologie pediatrique, Besançon, France; (15) France, Créteil, Centre des Maladies Respiratoires Rares (RESPIRARE®), CRCM, Créteil, France, Pédiatrie générale, Créteil, France; (16) France, Rennes, CHU Rennes - Hôpital Sud - Maternité, Pneumologie pediatrique, Rennes, France; (17) France, Rennes, CHU Rennes - Hôpital Sud - Maternité, Pneumologie pediatrique, Lille, France; (18) France, Marseille, Hôpitaux Universitaires de Marseille Timone, Pneumologie pédiatrique, Marseille, France; (19) France, Clermont-Ferrand, CHU Clermont-Ferrand Site Estaing, Pneumologie pédiatrique, Clermont-Ferrand, France; (20) France, Bordeaux, Hôpital des Enfants - Groupe hospitalier Pellegrin - CHU de Bordeaux, Pneumologie pédiatrique et CRCM pédiatrique, Bordeaux, France; (21) France, Nice, Hopital Lenval, Pneumologie pediatrique, Nice, France; (22) France, Le Mans, C.H. - Le Mans, Pneumologie pediatrique, Le Mans, France; (23) France, Saint-Denis, CHU saint denis de la reunion, Pediatrie generale, La Reunion, France; (24) France, Bordeaux, C.H.? Universitaire de Bordeaux, Pneumologie pediatrique, Bordeaux, France; (25) France, Tours, CHRU de Tours • Hôpital pédiatrique Clocheville, Pneumologie pediatrique, Tours, France; (26) France, Nice, CHU lenval nice, Pneumologie pediatrique, Nice, France; (27) France, Le Kremlin-Bicêtre, Hôpital Bicêtre AP-HP, Pédiatrie générale, Kremlin Bicêtre, France; (28) France, Strasbourg, CHRU Strasbourg - Prévention, Pneumologie pediatrique, Strasbourg, France; (29) France, Paris, Hôpital Robert Debré AP-HP, Pneumologie pédiatrique, Paris, France; (30) France, La Tronche, CHU Grenoble Alpes, Pneumologie pediatrique, Grenoble, France; (31) France, Paris, Hôpital Cochin-Port Royal - Centre de Planification et d'Education Familiale, Réanimation néonatale, Paris, France; (32) France, Toulouse, C.H.U de Toulouse, Pneumologie pediatrique, Toulouse, France; (33) France, Besançon, CHU de Besançon - Urgences, Pneumologie pediatrique, Besançon, France; (34) France, Paris, Hôpital Robert Debré AP-HP, Pneumologie pediatrique, Paris, France; (35) France, Rouen, C.H.U de Rouen, Pédiatrie médicale, Rouen, France; (36) France, Limoges, Hôpital de la mère et de l'enfant, Pediatrie generale, Limoges, France; (37) France, La Réunion, France, CHU Sud Réunion, Pneumologie pédiatrique, La Reunion, France; (38) France, Amiens, CHU Amiens-Picardie Site Nord, Pneumologie pediatrique,

Amiens, France; (39) France, Lyon, chu LYON hcl, Pneumologie pediatrique, Lyon, France; (40) France, Montpellier, C.H.U de Montpellier, Pneumologie pediatrique, Montpellier, France; (41) France, Nancy, C.H. Régional et Universitaire de Nancy Hôpital Central, Pneumologie pediatrique, Nancy, France; (42) France, Vandœuvre-lès-Nancy, CHRU de Nancy - Hôpitaux de Brabois, Pneumologie pediatrique, Nancy, France; (43) France, Vandœuvre-lès-Nancy, CHRU de Nancy - Hôpitaux de Brabois, Pneumologie pediatrique, Paris, France; (44) France, Angers, C.H.U Angers, Pneumologie pediatrique, Angers, France; (45) France, Rennes, CHU Rennes - Hôpital Sud - Maternité, Pneumologie pédiatrique, Rennes, France; (46) France, Paris, Hôpital Armand-Trousseau AP-HP, U.f. de génétique moléculaire (ap-hp), Paris, France; (47) France, Créteil, Hôpital Henri-Mondor AP-HP, Genetique, Creteil, France; (48) France, Paris, Hôpital Armand Trousseau AP-HP, ANATOMOPATHOLOGIE, Paris, France; (49) France, Paris, Hôpital Armand Trousseau AP-HP, Radiologie, Paris, France; (50) France, Paris, Hopital necker, Radiologie, Paris, France; (51) France, Créteil, C.H. Intercommunal Créteil, Pediatrie generale France, Créteil, Centre des Maladies Respiratoires Rares (RESPIRARE®), CRCM, Créteil, France, Pediatrie generale, Creteil, France

Introduction: Interstitial lung disease (ILD) in children (chILD) is rare and often severe. This study aims at analyzing the epidemiology of chILD in France from 2000 to 2022.

Methods: This study was retrospective and multicentric. A questionnaire was sent to all the RespiRare centers to collect the clinical, radiological, biological, histological and genetic data of the patients.

Results: 617 patients (0-18 years) were included in 42 centers. 84 patients were excluded. The median age at diagnosis was 0.3 years with 17% of familial forms. The main investigations performed were: chest CT scan (92%), bronchoalveolar lavage (52%), genetic analysis (78%), lung biopsy (23%). The main treatments were: corticosteroids (93%), oxygen therapy (52.2%), enteral nutrition (29%), hydroxychloroquine (16%), azithromycin (26%), immunosuppressive drugs (21%). The follow-up time was from 0 to 18,9 years (median duration 3,5years). The survival rate at 5 years was 68%. The overall incidence and prevalence were estimated at 38/million and 35/million children respectively.

Conclusion: This large chILD epidemiological study confirms the Spanish data with a higher incidence and prevalence than previously described. The large amount of phenotypic data collected will allow better understanding chILD and harmonizing their management.

Keywords: Chronic diseases, Diagnosis, Children