

Supplemental Table 1: Diseases classification.

Disorders more prevalent in infancy
Diffuse developmental disorders
Acinar dysplasia
Congenital alveolar dysplasia
Alveolar capillary dysplasia with misalignment of pulmonary veins
Growth abnormalities
Chronic neonatal lung disease (not prematurity-related)
Structural pulmonary changes with chromosomal abnormalities ¶¶
Associated with congenital heart disease in chromosomally normal children
Specific conditions of undefined aetiology
Primary Pulmonary interstitial glycogenosis §
Pulmonary interstitial glycogenosis associated with other disorders §
Neuroendocrine cell hyperplasia of infancy
Surfactant dysfunction mutations and related disorders
Surfactant protein B mutations (<i>SFTPB</i> ; R)
Surfactant protein C mutations (<i>SFTPC</i> ; D)
ABCA3 mutations (<i>ABCA3</i> bi-allelic mutations)
Brain-lung-thyroid syndrome (<i>NKX2-1/TTF-1</i> mutations; D) ¶¶
Lysinuric protein intolerance (<i>SLC7A7</i> ; R) ¶¶
Surfactant protein A mutations (<i>SFTPA</i> ; D) ¶¶
Acquired pulmonary alveolar proteinosis / autoimmune ¶¶
Histology with surfactant dysfunction disorder without a recognised genetic aetiology
Chronic pneumonitis of infancy
Desquamative interstitial pneumonitis
Nonspecific interstitial pneumonia
Pulmonary fibrosis ¶¶
Congenital multisystemic disorders and other genetic diseases
STING-associated vasculopathy with onset in infancy (<i>TMEM173-STING</i> ; D) ¶¶
ILD associated to dyskeratosis congenita (<i>DKC1</i> ; D) ¶¶
Filamin A mutations (<i>FLNA</i> ; D) ¶¶
Multisystemic Smooth Muscle Dysfunction (<i>ACTA2</i> ; D) ¶¶
<i>FARSB</i> , <i>MARS</i> and other <i>ARS</i> mutations ¶ (a) (R and D)
Pulmonary alveolar proteinosis due to GMCSF receptor deficiency (<i>CSF2RA</i> , <i>CSF2RB</i> ; D) ¶¶
COPA syndrome (<i>COPA</i> ; D) ¶¶
ILD associated to Arthrogyrosis-renal dysfunction-cholestasis syndrome (<i>VPS33B</i> ; D) ¶¶(b)
Alveolar hemorrhage syndromes
Acute idiopathic pulmonary hemorrhage of infancy ¶¶

Adapted from Deutsch GH, et al. Pathology Cooperative Group; ChILD Research Co-operative. Diffuse lung disease in young children: application of a novel classification scheme. *Am J Respir Crit Care Med.* 2007;176:1122. ¶¶ This category refers to the presence of alveolar simplification in chromosomal diseases. § Adapted from the original classification. ¶ Added entity not included in the original classification. (a) The only case in this group corresponds to a *FARSB* mutation. (b) Characterized by arthrogyrosis, renal tubular dysfunction and neonatal cholestasis, and platelet abnormalities and may be associated to pulmonary haemorrhage (personal observation). (c) Niemann-Pick type B (*SMPD1* 5), Niemann-Pick type A/B (*SMPD1* 1), Niemann-Pick type C1 (*NPC1* 2). (d) Only cases of BOOP non-related to transplantation or infection are included, other types of obliterans bronchiolitis are not included. Excluded entities from the original classification: pulmonary hypoplasia, prematurity-related chronic lung disease, infectious and postinfectious processes, aspiration syndromes, malignant infiltrates, opportunistic infections, disorders related to transplantation and rejection syndromes, veno-occlusive disease, congestive changes related to cardiac dysfunction. R: recessive biallelic mutation; D: dominant mutation; *SFTPB*: surfactant protein B; *SFTPC*: surfactant protein C; *ABCA3*: ATP-binding cassette 3; *TTF1*: thyroid transcription factor 1; *SLC7A7*: Solute Carrier Family 7 Member 7; *SFTPA*: surfactant protein A; *STING*: stimulator of interferon; *DKC1*: Dyskerin Pseudouridine Synthase 1; *FLNA*: Filamin A; *ACTA2*: Actin alpha 2; *FARSB*: Phenylalanine-tRNA synthetase Betachain; *MARS*: methionyl transfer RNA synthetase; *GMCSF*: Granulocyte-macrophage colony-stimulating factor; *CSF2RA*: Colony Stimulating Factor 2 Receptor Subunit Alpha; *CSF2RB*: Colony Stimulating Factor 2 Receptor Subunit Beta; *COPA*: non-clathrin-coated vesicular coat protein A; *VPS33B*: Vacuolar protein sorting 33. ILD: Interstitial Lung Disease; MPS: Mucopolysaccharidosis; SCID: severe combined immunodeficiency; ADA: adenosine deaminase; STAT1: Signal transducer and activator of transcription 1; M: months; Y: years.

Supplemental Table 1 (cont.): Diseases classification

Disorders not specific to infancy
Disorders of the normal host
Hypersensitivity pneumonitis
Acute eosinophilic pneumonia
Chronic eosinophilic pneumonia
Disorders related to systemic disease processes
Rheumatoid arthritis
Mixed connective tissue disease
Scleroderma
Granulomatosis with polyangiitis
Systemic lupus erythematosus
Antiphospholipid antibody syndrome
Goodpasture Syndrome
ILD in inflammatory bowel disease (Crohn)
ILD in dermatomyositis
Behcet disease
Churg-Strauss syndrome
Chronic alveolar haemorrhage, microscopic polyangiitis ¶
ILD associated to leukocytoclastic vasculitis ¶
Pulmonary lymphomatoid granulomatosis ¶
Sarcoidosis
Langerhans cell histiocytosis
Tuberous sclerosis ¶
Storage diseases
Niemann-Pick disease
Hermansky-Pudlak syndrome
Gaucher disease
ILD associated to Mucopolidosis type II ¶
Hurler syndrome (MPS I) ¶
Hunter syndrome (MPS II) ¶
Other alveolar haemorrhage syndromes: Idiopathic pulmonary hemosiderosis ¶
Disorders of the immunocompromised host
ILD associated to Chronic granulomatous disease ¶
Granulomatous Lymphocytic Interstitial Lung Disease ¶
ILD associated to Severe combined immunodeficiency (SCID) due to ADA deficiency ¶
ILD associated to Medullary aplasia ¶
ILD in LRBA deficiency ¶
ILD associated to Autoimmune lymphoproliferative syndrome (ALPS) ¶
ILD related to Gain-of-function mutation in STAT1 ¶
Disorders related to therapeutic intervention: drugs
Disorders related to therapeutic intervention: radiotherapy
Lymphatic disorders (Disorders masquerading as ILD)
Primary lymphangiectasia
Diffuse pulmonary lymphangiomatosis
Other entities
Lymphocytic interstitial pneumonia ¶
Follicular bronchiolitis ¶
Bronchiolitis obliterans Organising pneumonia ¶ (d)
Alveolar microlithiasis ¶
Pleuroparenchymal fibroelastosis ¶
Lung disease with unknown cause

Adapted from Deutsch GH, et al. Pathology Cooperative Group; ChILD Research Co-operative. Diffuse lung disease in young children: application of a novel classification scheme. *Am J Respir Crit Care Med.* 2007;176:1122. ¶ This category refers to the presence of alveolar simplification in chromosomal diseases. § Adapted from the original classification. ¶ Added entity not included in the original classification. (a) The only case in this group corresponds to a FARSB mutation. (b) Characterized by arthrogyposis, renal tubular dysfunction and neonatal cholestasis, and platelet abnormalities and may be associated to pulmonary haemorrhage (personal observation). (c) Niemann-Pick type B (*SMPD1*) 5, Niemann-Pick type A/B (*SMPD1*) 1, Niemann-Pick type C1 (*NPC1*) 2. (d) Only cases of BOOP non-related to transplantation or infection are included, other types of obliterans bronchiolitis are not included. Excluded entities from the original classification: pulmonary hypoplasia, prematurity-related chronic lung disease, infectious and postinfectious processes, aspiration syndromes, malignant infiltrates, opportunistic infections, disorders related to transplantation and rejection syndromes, veno-occlusive disease, congestive changes related to cardiac dysfunction. R: recessive biallelic mutation; D: dominant mutation; STFPB: surfactant protein B; STFPC: surfactant protein C; ABCA3: ATP-binding cassette 3; TTF1: thyroid transcription factor 1; SLC7A7: Solute Carrier Family 7 Member 7; STFPA: surfactant protein A; STING: stimulator of interferon; DKC1: Dyskerin Pseudouridine Synthase 1; FLNA: Filamin A; ACTA 2: Actin alpha 2; FARSB: Phenylalanine-tRNA synthetase Betachain; MARS: methionyl transfer RNA synthetase; GMCSF: Granulocyte-macrophage colony-stimulating factor; CSF2RA: Colony Stimulating Factor 2 Receptor Subunit Alpha; CSF2RB: Colony Stimulating Factor 2 Receptor Subunit Beta; COPA: non-clathrin-coated vesicular coat protein A; VPS33B: Vacuolar protein sorting 33. ILD: Interstitial Lung Disease; MPS: Mucopolysaccharidosis; SCID: severe combined immunodeficiency; ADA: adenosine deaminase; STAT1: Signal transducer and activator of transcription 1; M: months; Y: years.

Supplemental Table 2. Participants and data collection

Region	Province	Population <18y (2018)	Population <18y (2019)	Hospital	New cases 2018	New cases 2019	Prevalent cases 2018	Prevalent cases 2019	
Andalucía	Almería	144.867	145.658	Hospital Universitario Torrecárdenas	2	0	4	4	
	Cádiz	242.914	240.335	Hospital Universitario de Jerez	0	0	0	0	
	Córdoba	139.888	138.412	Hospital Universitario Reina Sofia	0	0	14	14	
	Granada		171.049	170.133	Hospital Universitario Clínico San Cecilio	0	4	0	4
					Hospital Universitario Virgen de las Nieves	0	5	5	10
	Málaga	313.603	314.191	Hospital Regional Universitario de Málaga	3	0	11	11	
Sevilla		388.842	385.635	Hospital Universitario Virgen del Rocío	11	10	58	68	
				Hospital Universitario Virgen Macarena	0	0	2	2	
Aragón	Zaragoza Huesca, Teruel*	224.054	224.324	Hospital Universitario Miguel Servet	1	1	7	8	
Principado Asturias	Asturias	135.285	133.937	Hospital Universitario Central de Asturias	0	0	1	1	
Islas Baleares	Baleares	211.779	212.954	Hospital Universitari Son Espases	3	3	12	15	
Canarias	Las Palmas	190.301	187.775	Hospital Universitario Materno Infantil Canarias	2	2	3	5	
	Santa Cruz de Tenerife	168.567	168.023	Hospital Universitario de Canarias	3	1	6	7	
				Hospital Universitario Nuestra Sra. Candelaria	0	0	2	2	
Cantabria	Cantabria	93.079	92.344	Hospital Universitario Marqués de Valdecilla	0	0	1	1	
Castilla La Mancha	Toledo	134.863	135.878	Hospital Universitario de Toledo	0	0	0	0	
Castilla y León	León	60.037	59.189	Complejo Asistencial Universitario de León	0	0	0	0	
	Salamanca	47.115	46.619	Hospital Universitario de Salamanca	1	1	1	1	
	Valladolid	82.657	82.182	Hospital Clínico Universitario de Valladolid	0	0	0	0	
Cataluña	Barcelona	1.029.161	1.030.918	Hospital Universitari Vall d'Hebron	4	8	40	48	
				Hospital Sant Joan de Déu	4	2	15	17	
				Consorci Corporació Sanitària Parc Taulí Sabadell	3	2	6	8	
				Hospital Universitari Germans Trias i Pujol	0	0	2	2	
	Girona	147.321	147.996	Hospital de la Santa Creu i Sant Pau	0	0	0	0	
				Hospital Universitari Dr. Josep Trueta	0	0	0	0	
	Lleida	77.922	78.212	Hospital Universitari Arnau de Vilanova	0	0	0	0	
				Hospital Universitari Joan XXIII	0	0	0	0	
Extremadura	Badajoz	117.238	115.680	Hospital Universitario de Badajoz	0	1	1	2	
	Cáceres	60.224	59.236	Hospital Universitario de Cáceres	0	0	0	0	
Galicia	La Coruña	349.600	347.614	Complejo Hospitalario Universitario A Coruña	0	0	0	0	
	Lugo, Pontevedra*			Hospital Clínico Universitario de Santiago	2	3	6	7	
	Orense	36.516	36.435	Complejo Hospitalario Universitario de Ourense	0	0	2	2	
Comunidad de Madrid	Madrid	1.228.148	1.231.516	Hospital Universitario La Paz	8	1	12	13	
				Hospital General Universitario Gregorio Marañón	0	0	12	12	
				Hospital Universitario 12 de Octubre	0	1	10	11	
				Hospital Universitario Puerta De Hierro	1	1	1	1	
				Hospital Infantil Universitario Niño Jesús	2	2	23	21	
				Hospital Clínico San Carlos	0	0	0	0	
				Hospital Universitario Ramón y Cajal	0	1	0	1	
Clínica Universidad de Navarra de Madrid	0	0	0	0					
Región de Murcia	Murcia	306.833	306.798	Hospital Clínico Universitario Virgen de la Arrixaca	4	4	24	28	
Comunidad Navarra	Navarra	121.250	121.715	Complejo Hospitalario de Navarra	0	0	1	1	
País Vasco	Guipuzcoa	124.318	123.754	Hospital Universitario Donostia	1	0	4	4	
				Hospital de Zumárraga	0	0	0	0	
	Vizcaya	184.224	183.912	Hospital Universitario Cruces	3	1	4	5	
				Hospital Universitario Basurto	1	2	3	5	
Comunidad Valenciana	Alicante	328.366	329.900	Hospital General de Alicante	2	0	3	3	
	Valencia	631.037	630.564	Hospital Clínico Universitario de Valencia	1	5	29	32	
	Castellón, Albacete*			Hospital de Manises	1	0	1	1	
				Hospital Universitario y Politécnico de La Fe	1	0	4	4	
Total		7.644.155	7.636.093		64	61	330	381	

*Population of areas with a referral hospital in a province which is not their place of residence has been added. Y:years