

Table 1S List of data contributors

Country	City	Centre	No of cases
Belgium	Leuven	Pediatric pulmonology, KUL UZ Gasthuisberg	2
the Czech Republic	Olomouc	Dept. of Pediatrics, University Hospital Olomouc	1
Denmark	Copenhagen	Paediatric Pulmonary Service, Dept of Paediatrics and Adolescent Medicine, Copenhagen University Hospital, Rigshospitalet	5
Denmark	Aarhus	Danish Center of Pediatric Pulmonology and Allergology, Department of Pediatrics and Adolescents Medicine, University Hospital of Aarhus,	1
England	London	Royal Brompton Hospital, London, United Kingdom	2
France	Paris	APHP-Sorbonne Université, Pediatric Pulmonology and Reference Center for rare lung diseases RespiRare, Inserm U933 Laboratory of childhood genetic diseases, Armand Trousseau Hospital	13
France	Lyon	Université de Lyon, Hôpital Femme Mère Enfant, Pediatric pulmonology department	1
France	Strasbourg	Strasbourg University, Hôpital de Hautepierre, Pediatric pulmonology department	1
France	Toulouse	Dept of Pediatric Pulmonology and Allergy, Children's University Hospital,	1
Germany	Hannover	Clinic for Paediatric Pneumology, Allergology and Neonatology, Hannover Medical School, German Center for Lung Research (DZL)	21
Germany	Essen	Clinic for pediatrics III, University hospital Essen	12

Greece	Athens	2nd Pulmonary Medicine Department General University Hospital, Athens Medical School National and Kapodistrian University of Athens	2
Ireland	Dublin	CHI at Tallaght University Hospital,	1
Palestine	East Jerusalem	Pediatric department, Makassed	6
Poland	Warsaw	Department of Pediatric Pneumology and Allergy, Medical University of Warsaw	9
Portugal	Lisbon	Department of Pediatrics, Respiratory Unit. Hospital de Santa Maria, Centro Hospitalar Lisboa Norte	2
Portugal	Porto	Departamento de Ginecologia Obstetrícia e Pediatria, Faculdade de Medicina, Universidade do Porto and Serviço de Pediatria, Centro Hospitalar Universitário de S. João	1
Spain	Barcelona	Pediatric Pulmonology and Allergology Department. Parc Taulí Hospital Universitari, Institut d'Investigació i Innovació Parc Taulí I3PT, Univesitat Autónoma de Barcelona	3
Spain	Palma de Mallorca	Pediatric pulmonology department, Son Espases University Hospital	3
Spain	Valencia	Pediatric pulmonology. Clinical Hospital of Valencia	1
Sweden	Gothenburg	Devison of Paediatrics, Drottning Silvias barn- och ungdomssjukhus, Univercity of Gothenburg	9
Turkey	Ankara	Gazi University Faculty of Medicine, Department of Pediatric Pulmonology	12
Turkey	Ankara	Hacettepe University Faculty of Medicine, Department of Pediatric Pulmonology	6
Turkey	Ankara	Department of Pediatrics, Pediatric Pulmonology Division, Ankara University Faculty of Medicine	4

Turkey	Istanbul	Division of Paediatric Pulmonology, Marmara University Faculty of Medicine	3
--------	----------	---	---

Data contributors are arranged by country in alphabetical order. The median (range) number of included patients per centre was 2 (1;21).

Table 2S. Detailed data of diagnostic workup in each sub-group

	HRCT		BAL			Lung biopsy				Echocardiography		Genetic analysis	
	n (%)	GGO, n (% of tested)	n (%)	HLM, n (% of tested)	Fresh bleeding, n (% of tested)	n (%)	Hemochromatosis/HLM, n (% of tested)	Vasculitis/capillaritis, n (% of tested)	Fibrosis, n (% of tested)	n (%)	PAH n (% of tested)	Tested, n (%)	Mutation
All DAH patients n =117	110 (94)	78 (71)	100 (85)	74 (74)	28 (28)	49 (42)	25 (51)	3 (6)	8 (16)	94 (80)	11(12)	52 (44)	
IPH n=36	34 (94)	23 (68)	29 (81)	25 (86)	9 (31)	15 (42)	10 (67)	0	3 (20)	29 (81)	2(7)	15 (42)	Trisomy 21 (n=3) Other ^s (n=1) No mutation (n=11) Pending (n=1)
DAH associated with autoimmune features n=20	17 (85)	13 (76)	18 (90)	13 (72)	6 (33)	11 (55)	4 (36)	0	2 (18)	15 (75)	0	11 (55)	NKX2.1 (n=1) No mutation (n=9) Pending (n=1)
Systemic and collagen diseases n=18	18 (100)	12 (67)	14 (78)	9 (64)	6 (43)	4 (22)	3 (75)	1 (25)	0	15 (83)	1 (7)	3 (17)	HLA DQA1*05 positive, HLA DQB1*02 positive (n=1) 22q11 del (n=1) No mutation (n=1)
Immuno-allergic diseases n=10	9 (90)	7 (78)	9 (90)	7 (78)	1 (11)	2 (20)	2 (100)	0	0	10 (100)	1 (10)	1 (10)	No mutation (n=1)
Other chILD n=5	4 (80)	3 (75)	5 (100)	4 (80)	2 (40)	5 (100)	3 (60)	1 (20)	1 (20)	4 (80)	3 (75)	4 (80)	STPTC (n=1) NKX2.1 (n=1) TBX4 (n=1) Unknown (n=1)

Autoinflammatory n=3	3 (100)	3 (100)	3 (100)	3 (100)	0	3 (100)	0	0	1(33)	2 (66)	1 (50)	3(100)	COPA (n=2) STAT-3 (n=1)
Secondary to other conditions n=21	22 (88)	18 (82)	20 (80)	13 (65)	4 (20)	9 (36)	2 (22)	1 (11)	1 (11)	18 (72)	3 (17)	13 (62)	Trisomy 21 (n=2) ATP7B (n=1) PFIC 1 (n=1) WAS (n=1) FLT3-ITD (n=1) WT1 (n=1) No mutation (n=6)
Non-specified DAH diagnosis =5	3 (75)	2 (67)	2 (50)	NA	NA	0	0	0	0	3 (75)	0	2 (50)	CYBB (n=1) Trisomy 21 (n=1) No mutation (n=1)

HRCT; High-resolution CT, BAL; Bronchoalveolar lavage, Echo; echocardiography, DAH; Diffuse alveolar haemorrhage, IPH; Idiopathic pulmonary Hemosiderosis, chILD; Childhood Interstitial Lung Disease, HLA DQA1; human leucocyte antigen DQ alpha 1, HLA DQB1; human leucocyte antigen DQ beta 1, 22q11; DeGeorges syndrome, NKX2.1; NK Homeobox 1, STPTC; surfactant protein C, TBX4; T-box transcription factor 4, COPA; COPI coat complex subunit alpha, STAT-3; Signal transducer and activator of transcription 3, ATP7B; ATPase cobber transporting beta; PFIC-1; progressive familial intrahepatic cholestasis 1, WASp; Wiscott -Aldrich Syndrome, FLT3-ITD; fms-like tyrosine kinase 3 internal tandem duplication, WT1; Wilms' tumor 1; CYBB; cytochrome B-245 beta chain. [§]Heterozygote for CSF2RB (colony-stimulating factor 2 receptor subunit beta) (pulmonary alveolar proteinosis).

Table 3S Autoantibodies and specific immunoglobulins

	ATA, IgA	ATA, IgG	antigliadin antibodies (AGA)	ANCA	ANA	Anti-endomysium antibodies (AEA)	SMA	RF	Cow's milk, IgG
All DAH patients, n=117	12/69	4/55	9/56	17/100	17/96	5/51	7/74	10/66	5/37
IPH, n=36	0/22	1/16	1/20	0/28	0/28	0/18	0/18	0/21	0/14
DAH associated with autoimmune features, n=20	0/13	0/10	0/9	7/18	8/18	0/8	4/15	6/13	1/6
Systemic and collagen diseases, n=18	2/8	0/6	0/5	10/18	4/16	1/7	1/12	1/7	0/4
Immuno-allergic diseases, n=12	7/9	3/9	6/8	0/9	1/10	4/8	0/8	1/9	4/6
Other chILD condition, n=5	1/1	0/1	0/2	0/4	0/4	0/1	0/2	0/2	0/1
Autoinflammatory diseases, n=3	0/3	0/3	1/3	0/3	2/3	0/1	1/3	1/3	0/1
Secondary to other conditions, n=21	2/11	0/9	1/7	0/17	2/15	0/6	1/14	1/9	0/5
Non-specified DAH diagnosis, n=4	0/2	0/1	0/2	0/3	0/2	0/2	0/2	0/2	0

Results are presented as number tested positive / number tested. DAH; Diffuse alveolar haemorrhage, IPH; Idiopathic pulmonary Hemosiderosis, chILD; Childhood Interstitial Lung Disease, ATA; anti transglutaminase antibody, ANCA; antineutrophilic cytoplasmic antibodies, ANA; antinuclear antibodies, SMA; anti-double-stranded DNA and anti-smooth-muscle antibodies; RF; rheumatoid factor.

Table 4S. Results from first available pulmonary function test.

	FEV1, % pred.	FVC, % pred.	FEV1/FVC, % pred.	TLC, % pred.	DLCO, % pred.	KCO, % pred.
All DAH patients, n=117	66 (56;86) n=36	68 (56;86) n=36	96 (84;106) n=36	87 (72;106) n= 34	69 (58;91) n= 26	77 (67;92) n=10
IPH, n=36	74 (53;79) n=9	77 (70;81) n=9	97 (94;104) n=9	73 (65;91) n=5	61 (54;69) n=7	75 (69;80) n=2
DAH associated with autoimmune features, n=20	61 (59;61) n=5	59 (50;67) n=5	97 (90;102) n=5	91 (68;99) n=6	54 (43;83) n=4	67 n=1
Systemic and collagen diseases, n=18	78 (66;85) n=11	78 (60;86) n=11	97 (84;106) n=11	89 (79;104) n=10	75 (68;94) n=9	77 (71;85) n=3
Immuno-allergic diseases, n=12	86 (44;98) n=3	90 (66;98) n= 3	91 (66;106) n=3	106 (105;106) n=2	94 (90;97) n=2	87 (76;98) n=2
Other chILD condition, n=5	51 (45;57) n=2	59 (50;68) n=2	65 n=2	114 n=1	69 n=1	NA
Autoinflammatory diseases, n=3	27 n=1	24 n=1	88 n=1	73 n=1	72 n=1	NA
Secondary to other conditions, n=21	71 (63;95) n= 6	66 (60;88) n=6	103 (93;107) n=6	81 (77;85) n= 6	100 (55;113) n=3	91 (83;98) n=2
Non-specified DAH diagnosis, n=4	NA	NA	NA	NA	NA	NA

Data is presented as median (IQ-range) unless n <5 in which case median (range) is presented. DAH; Diffuse alveolar haemorrhage, IPH; Idiopathic pulmonary Hemosiderosis, chILD; Childhood Interstitial Lung Disease, FEV1; Forced Expiratory Volume in the first second, FVC; Forced Vital Capacity, TLC; Total Lung Capacity, DLCO; the diffusion capacity for Carbon Monoxide, KCO; Carbon monoxide transfer coefficient n = the number of patients in which the data was available. NA; no data available.

Table 5S. Most frequent medical treatments besides corticosteroids, hydroxychloroquine, and azathioprine

	Non-specific immuno-suppressive drugs	Immuno-globulins	cytostatic	Biological treatment	plasmapheresis
All DAH patients, n=117	16 (14%)	3 (3%)	11 (9%)	11 (9%)	6 (5%)
IPH, n=36	3 (8%) Mycophenolate mofetil, 2 Cyclosporine, 1	0	4 (11%) 6-mercaptopurine, 2 Cyclophosphamide, 2	1 (3%) Rituximab, 1	0
DAH associated with autoimmune features n=20	3 (15%) Mycophenolate mofetil, 3	1 (5%)	2 (10%) Cyclophosphamide, 2	2 (10%) Rituximab, 1 Baricitinib, 1	0
Systemic and collagen diseases, n=18	8 (44%) Mycophenolate mofetil, 7 Cyclosporine, 3		5 (28%) Methotrexate, 2 Cyclophosphamide, 3	5 (28%) Rituximab, 4 Mepolizumab, 1	6 (33%)
Immuno-allergic diseases, n=10	0	0	0	0	0
Other ILD condition, n=5	0	1 (20%)	0	1 (20%) Tocilizumab	0
Autoinflammatory diseases, n=3	0	0	0	2 (33%) Baricitinib, 2 Canakinumab, 1 Rituximab, 1	0
Secondary to other conditions, n=21	2 (10%) Mycophenolate mofetil, 2	1 (5%)	0	0	0
Non-specified DAH diagnosis, n=4	0	0	0	0	0

DAH; Diffuse alveolar haemorrhage, IPH; Idiopathic pulmonary Hemosiderosis, chILD; Childhood Interstitial Lung Disease.

Table 6S. Follow-up data: time of follow-up, chest imagining, and pulmonary function.

	Time to follow-up		Follow-up HRCT or chest x-ray	Follow-up pulmonary function					
	from debut, yrs	from diagnosis, yrs		follow-up HRCT or x-ray with abnormalities Assoc. with DAH	FEV1, % pred.	FVC, % pred.	FEV1/FVC, % pred.	TLC, % pred.	DLCO, % pred.
All DAH patients, n=117	4 (1.5;7.8)	3.2 (1.2;7.0)	60% (61/101)	84 (66;97) n=20	87 (69;96) n=63	96 (86;101) n=63	100 (85;112) n=42	87 (66;99) n=47	91 (83;94) n=21
IPH, n=36	8.8 (5.2;16.0)	7.4 (4.8;14.6)	62% (18/29)	74 (66;93) n=17	79 (68;87) n=17	91 (86;97) n=17	99 (95;112) n=9	83 (70;93) n=14	87 (73;99) n=7
DAH associated with autoimmune features, n=20	4.4 (3.1;7.0)	3.9 (1.7;9.4)	68% (13/19)	61 (60;73) n=11	70 (60;78) n=11	86 (82;98) n=11	85 (74;94) n=9	71 (53;83) n=8	89 n=2
Systemic and collagen disorders, n=18	3.7 (1.8;5.5)	3.4 (1.7;5.0)	56% (9/16)	99 (94;112) n=16	101 (95;116) n=16	97 (95;99) n=16	108 (103;120) n=11	100 (85;113) n=14	98 (86;122) n=4
Immuno-allergic disorders, n=10	5.0 (2.6;7.0)	3.8 (1.3;6.3)	55% (5/9)	87 (71;103) n=8	88 (86;98) n= 8	85 (77;100) n=8	114 (105;129) n=4	92 (60;120) n=4	88 (66;114) n=4
Other chILD, n=5	1.5 (0.9;6.9)	1.5 (0.9;7.3)	60% (3/5)	83 (30;95) n=3	92 (30;93) n=3	107 (88;108) n=3	124 (108;140) n=2	93 (82;103) n=2	78 n=1
Autoimmune disorders, n=3	13.0 (12.8;17.8)	11.0 (5.8;14.9)	100% (2/2)	44 (23;64) n=2	42 (21;62) n=2	100 (61;106) n=2	58 (39;76) n=2	NA	NA
Secondary to other conditions, n=21	2.4 (1.8;3.0)	2.3 (1.7;3.4)	44% (8/18)	93 (81;96) n= 11	87 (77;95) n=11	101 (93;108) n=11	90 (81;100) n= 4	87 (55;97) n=2	91 (74;95) n=3
Non-specified DAH diagnosis, n=4	4.5 (0.7;32.4)	4.4 (0.6;32.4)	50% (2/4)	72 n=1	69 n=1	104 n=1	90 n=1	NA	NA

Data are presented as median (IQ-range) unless other is specified. DAH; Diffuse alveolar haemorrhage, IPH; Idiopathic pulmonary Hemosiderosis, chILD; Childhood Interstitial Lung Disease, FEV1; Forced Expiratory Volume in the first second, FVC; Forced Vital Capacity, TLC; Total Lung Capacity, DLCO; the diffusion capacity for Carbon Monoxide, VA; Alveolar Volume. n = the number of patients in which the data was available, yrs; years.

Table 7S. Description of patients who have passed away

Diagnoses	Age at debut, years	Gender	Cause of death	Age at time of death, years	Period from debut to time of death	Medical treatment	
						Type of medication	Months of treatment
IPH	7.0 years	Female	Acute bleeding	7.4 years	4.0 months	Methylprednisolone, iv Prednisolone, po Azathioprine, po	NA NA NA
IPH	7.5 years	Female	Acute bleeding	9 years	18 months	Prednisolone, po	9 (8030 mg)
IPH	2.8 years	Female	Acute bleeding	4.9 years	1.9 years	Prednisolone, po	12 (8800 mg)
IPH	0.3	Female	Acute rejection after lung transplantation	NA	NA	Methylprednisolone, iv Prednisolone, po Hydroxychloroquine, po Cyclosporine	NA NA NA NA
IPH	9.3	Female	Chronic respiratory failure	13.3 years	4.0 years	Methylprednisolone, iv Prednisolone, po Hydroxychloroquine, po Azathioprine, po Cyclophosphamide	NA 46 39 6 NA
Anti-glomerular basement membrane disease	12.8	Female	Chronic respiratory failure	12.9 years	1 month	Methylprednisolone, iv Hydroxychloroquine, po Cyclophosphamide	1 155 NA
STAT-3 mutation	2.0	Male	Died after lung transplantation	14.8 years	12.8 years	Methylprednisolone, iv Prednisolone, po Hydroxychloroquine, po Etanercept Mycophenolate mofetil	NA NA NA NA NA
Pulmonary interstitial glycogenosis (PIG)	0.0	Male	Multiorgan failure	0.05	0.5 months	Methylprednisolone, iv	1
Lung fibrosis	5.8 years	Male	Chronic respiratory failure	14.9 years	9.1 years	Methylprednisolone, iv Prednisolone, po Immunoglobulins Azathioprine, po Tocilizumab	NA 109 NA 3 NA

Pulmonary hemorrhage with cow's milk antibody (IgG)	8 months	Female	Acute bleeding	11 months	3 months	Other	
Cantu-syndrome	9 months	Male	Pulmonary infection	3.3 years	2.4 years	methylprednisolone, iv Prednisolone, po Other	28 months NA
DAH associated with autoimmune features (ANCA positive)	15.2 years	Male	Pulmonary aspergillosis	16.7 years	18 months	methylprednisolone, iv Cyclophosphamide	1 NA
DAH developed secondary to infection	9 months	Male	Pulmonary infection	2.5 years	22 months	methylprednisolone, iv	1
Bone marrow transplant-related lung injury	14.3 years	Female	DAH/idiopathic pneumonia syndrome after allogeneic bone marrow transplantation	15.6 years	16 months	Other	NA
non-specified DAH diagnosis	17 years	Male	Pulmonary aspergillosis	17.4 years	3.6 months	NA	NA

DAH; Diffuse alveolar haemorrhage, IPH; Idiopathic pulmonary Hemosiderosis, ANCA; Anti-neutrophil cytoplasm antibodies.