Supplementary appendix

Index:

Supplementary table 1

Extensive description of case reports/series and clinical trials of humans with a surfactant-processing mutation involved in fibrotic IP

Gene mutation	Diagnosis	Drug	Outcome after treatment	Ref
Adult case reports and series				
SFTPC p.I73T	Adult with CPFE	Prednisone	No improvement, pulmonary function test and HRCT worsened	(1)
SFTPA2 p.G231V	1 Adult with HP	Prednisone and avoidance of birds	Remission of disease	(2)
	1 Adult with PF and broncho-alveolar carcinoma	Prednisone	No improvement	
ABCA3 p.G964D	Adult with PF	Antibiotics	Ineffective	(3)
		Prednisone and azithromycin	Minimal progression of lung function impairment. HRCT	

			scan: disappearance of ground-glass areas with persistence of fibrosis	
ABCA3 p.G964D	Adult with restrictive lung disease[#]	Steroids and azithromycin	Moderate improvement, transient, sick-same after 5 years	(4)
HPS[*]	Adult with HPS	Antibiotics and oxygen inhalation	Not effective, lung function deteriorated, patient died	(5)
HPS1 IVS5+5 G>A	Adult with HPS	Prednisolone, cyclosporine A	Vital capacity decreased and the areas of lung opacity deteriorated	(6)
		+ Pirfenidone	Respiratory symptoms stabilized and serum KL-6 level decreased for a few months, later progression, increase in areas of diffuse lung opacity and gradual decrease of lung volume. Oxygen therapy was administered, died of an acute exacerbation.	
HPS4 p.Q620X	Adult with HPS	Corticosteroids, pirfenidone	Breathing condition, clinical marker levels and radiological characteristics stabilized. Corticosteroid tapered, later gradual progression of bilateral diffuse ground-glass opacity with mild traction bronchiectasis, condition still considered stable	(7)

HPS1 p.L668P	Adult with HPS	Prednisolone, pirfenidone, azathioprine	Progressively deterioration of diffuse reticulations, later died	(8)
*HPS**	Adult with HPS	Oral corticosteroids	No symptomatic improvement, died	(9)
HPS4 p.P685delC	Adult with HPS	High dose steroids and azathioprine	No effect; still alive but progressively dyspnoeic	(10)
*HPS**	Adult with HPS and pulmonary sarcoidosis	Prednisone	Resolution of dyspnoea	(11)
*HPS**	Adult with HPS	Prednisolone and pirfenidone	Gradual progression, oxygen therapy initiated	(12)
Paediatric case reports/series with *SFTPC* mutations				
SFTPC p.I73T p.I38F, p.V39L	4 Children with DIP , 1 child with chronic interstitial pneumonitis (p.V39L)	1/5 Systemic steroids	No improvement	(13)
		5/5 Hydroxychloroquine	2 out of 5: improved lung function, later on reduced exercise capacity 3 out of 5 free of symptoms, improved pulmonary function test	
SFTPC p.A116D	Child with NSIP	Hydroxychloroquine and supplemental oxygen	Improvement on chest film, weight gain, no longer supplemental oxygen required	(14)
SFTPC p.I73T	Child with ILD	Corticosteroids and supplemental oxygen	Condition improved	(1)

SFTPC c.460+1 G → A	Child with cellular or NSIP	Corticosteroids and supplemental oxygen	Respiratory symptoms (tachypnea and cyanosis) improved somewhat	(15)
SFTPC p.I73T	5 Children with chronic ILD	5/5 Mehtylprednisolone, 4/5 Hydroxychloroquine, 5/5 Supplemental oxygen	Decrease in retractions, oxygen requirement and in breath rate	(16)
Different *SFTPC* BRICHOS/ non BRICHOS	17 Children with ILD[##](NSIP PAP, DIP)	14/17 Hydroxychloroquine	12/14 moderate improvement – good response, 2/14 no improvement	(17)
		15/17 Systemic steroids	14/15 moderate improvement – good response, 1/15 no improvement	
		7/17 surfactant	2 moderate improvement – good response, 5 no improvement	
		3/17 Colchicine	3 no improvement	
SFTPC 14 non BRICHOS, 6 BRICHOS	22 Children with chronic ILD at diagnosis	18/22 Methylprednisolone, 11/22 Hydroxychloroquine, 5/22 Azithromycin 20/22 Supplemental oxygen	4 still with oxygen (did not receive HCQ) 16 weaned off oxygen, of which 4 symptom free, 12 with progressive clinical improvement, moderate dyspnoea and exercise tolerance	(18)
SFTPC p.G97S	Child with CPI pattern with globular	Home ventilator support, oxygen, pulse methylprednisolone,	Resolution of ground glass opacities, thickening of interlobular septa persisted, ventilation support was	(19)

	alveolar proteinosis	azithromycin, hydroxychloroquine	discontinued. Supplemental oxygen requirement remained.	
SFTPC p.I73T, p.I38F	2 Children with CPI	Hydroxychloroquine, prednisone, ranitidine, TMP-SMX, two months after therapy began, pulse therapy of methylprednisolone, later hydroxychloroquine alone	1: minute ventilation (V_E) remained high later improved and became normal, respiratory system compliance (C_{rs}) continued to decline improved after methylprednisolone, 2: improved V_E, . Functional residual capacity declined, later on improved and became normal. C_{rs} continued to decline, later on improved reaching normal values	(20)
SFTPC p.I73T	Child with CPI, pneumatoceles after biopsy	Methylprednisolone, hydroxychloroquine, azithromycin	Pneumatoceles resorbed after 10 days. Weaned off oxygen, steroid treatment was stopped, radiological improvement	(21)
SFTPC p.I73T	Child with ILD	Hydroxychloroquine, oxygen supplementation	Improvement of cough and dyspnoea, three months completely asymptomatic, disappearance ground glass abnormalities, six months disappearance micronodules	(22)
	Child with ARDS/DIP	Supplemental oxygen and steroids	No improvement	

		Steroids pulse therapy	Initial improvement, but thereafter progression of ILD	
		Hydroxychloroquine	Improvement in saturation, discontinuation of oxygen therapy	
		Hydroxychloroquine replaced by azithromycin	Clinically and radiologically stable	
	Child with DIP	Hydroxychloroquine and steroids	No effect, died	
SFTPC p.I73T	Child with chILD	Bronchodilators, inhaled corticosteroids and antileukotrienes, azathioprine, hydroxychloroquine and intravenous immunoglobulins, exogenous surfactant	No effect	(23)
SFTPC p.I73T	Child with PAP and NSIP	Supplemental oxygen, whole lung lavages, systemic corticosteroids and azathioprine	Significant acute improvements of gas exchange with a reduced need for additional oxygen, later reduced effect	(24)
		Additional corticosteroid pulse therapy plus azathioprine	Still oxygen dependent, delay in growth	

SFTPC Δexon 4	Child with respiratory distress	Oral and intravenous corticosteroids, hydroxychloroquine, supplemental oxygen	Some improvement in respiratory status, still continuous supplemental oxygen requirement and persistent respiratory distress	(25)
SFTPC p.E66K, p.I73T, p.V102M, p.A155P	8 Children with idiopathic diffuse lung diseases	2 Children supplemental oxygen, pulse steroids and hydroxychloroquine	Little effect, died	(26)
		Supplemental oxygen, pulse steroids, hydroxychloroquine	Alive on oxygen, listed for transplant	
		Supplemental oxygen, hydroxychloroquine	Alive on room air	
		Supplemental oxygen, steroids, hydroxychloroquine, azithromycin	Alive on room air	
		Supplemental oxygen, pulse steroids, hydroxychloroquine	Alive on oxygen	
		Supplemental oxygen, pulse steroids, bronchodilators, antibiotics	Alive on oxygen	
		Steroids, supplemental oxygen	Alive on room air	

SFTPC p.L188Q	2 Children with respiratory distress (NSIP- like pattern)	Methylprednisolone and hydroxychloroquine	No effect	(27)
*SFTPC**	Child with CPI	Corticosteroids, hydroxychloroquine and continuous oxygen	Child gained weight, discharged, later respiratory function slowly and progressively declined, underwent lung transplantation	(28)
SFTPC p.I73T	Child with NSIP/PAP	Supplemental oxygen, antibiotics and oral corticosteroids	No effect, died	(29)
SFTPC p.I73T	Child with PAP/ILD	Whole lung lavages, systemic corticosteroids and azathioprine	Restored gas exchange and growth velocity, normal psychomotor development.	(30)
SFTPC p.G182R, p.L188Q, p.C189W	1 Child with PAP/NSIP	Clearance, steroids, hydroxychloroquine, mechanical ventilation	Initially could not be weaned of mechanical ventilation, age of 6 decannulated	(31)
	1 Child with respiratory failure	Clearance, steroids, azathioprine, mechanical ventilation	Age of 5 successfully decannulated	
	1 Child with respiratory failure	Steroids, azithromycin, hydroxychloroquine, mechanical ventilation	Decannulated age of 3, no pulmonary-related hospitalizations anymore	

SFTPC p.L81V	Child with surfactant protein C deficiency	Hydroxychloroquine, oxygen therapy	Respiratory improvement, stable with minimal symptoms	(32)
*SFTPC**	Child with NSIP	Systemic steroids, azathioprine, hydroxychloroquine	Good response	(33)
Different *SFTPC* mutations	15 Children with interstitial chronic lung disease	All methylprednisolone 5/15 Azithromycin 8/15 Hydroxychloroquine	All alive, 11 patients still symptomatic with cough, dyspnea, or exercise intolerance.	(34)
Paediatric case reports/series with *ABCA3* mutations				
*ABCA3**/*SFTPC*	Child with DPLD/surfactant dysfunction	Surfactant	Good	(33)
		Systemic steroids	None, later on died	
*ABCA3**	Child with CPI	Systemic steroids, Surfactant	Good,	
		Chloroquine	None, later on died	
	Child with DIP	Systemic steroids, hydroxychloroquine	None, later on died	
ABCA3 c.358_359del	Child with ABCA3 deficiency	Methylprednisolone, oxygen therapy	Minimal improvement, died from progressive respiratory failure	(32)

ABCA3 p.W1148X and p.T1114A	Child with PAP-like features	Methylprednisolone, antibiotics, antivirals and antifungals, oxygen, mechanical ventilation	Ineffective, no improvement in respiratory function and chest X-ray findings	(35)
		BAL with bovine surfactant	Improved radiographic findings	
		Hydroxychloroquine	Decreased oxygen administration	
ABCA3 p.G964D	Child with (possible) IPF	Prednisone and macrolides	Stable condition	(3)
ABCA3 p.A307V	Child with respiratory distress	Dexamethasone and surfactant, CPAP	Some effect, no CPAP required, later reduced effect again CPAP required	(36)
		Methylprednisolone, azithromycin, hydroxychloroquine	No CPAP required, only nasal cannula	
ABCA3 p.Y1515X	Child with RDS	Pulse steroids, antibiotics	No improvement, died	(37)
ABCA3 p.R194G and V1615GfsX15	2 Children with IRDS	CPAP, corticosteroids and hydroxychloroquine	Little effect, died	(38)
ABCA3 p.D253H	Child with DPLD	Methylprednisolone, oral prednisone, oxygen therapy	No improvement	(39)

		Azithromycin	No steroids required, weaned off oxygen	
ABCA3 p.R280C and p.E690G	Child with DIP	Oxygen supplementation, surfactant therapy, corticosteroids	No improvement	(40)
		Hydroxychloroquine	Improvement FEV1	
ABCA3 p.D507del CA Ter 508, p.D696N	Child with DIP	Dexamethasone, supplemental oxygen, surfactant therapy	Some improvement, later further deterioration by lowering dose dexamethasone	(41)
		Methylprednisolone, azithromycin, hydroxychloroquine	Slow, but notable improvement, still oxygen requirement, remained tachypneic	
ABCA3 p.K914R, p.L1238_E1239insGG	Child with ILD	Methylprednisolone, antibiotics	Only transient and very limited effects	(42)
		+ Hydroxychloroquine	Reduced oxygen and corticosteroids required. Improvement on radiological and clinical aspects	
ABCA3 c.59G>T and c.2646_2647insC	Child with severe respiratory distress syndrome	Home ventilator, methylprednisolone, hydroxychloroquine, azithromycin	Respiratory mechanics and gas exchange noticeably improved, less hypercapnia	(43)
ABCA3 p.H778R, p.L1252P	Child with DIP like pattern	Methylprednisolone, prednisone and	Respiratory symptoms completely disappeared , complete resolution of ground	(44)

		hydroxychloroquine, clarithromycin	glass opacities, improved lung function	
Different *ABCA3* mutations	9 Children with PAP pattern, DIP pattern and NSIP pattern	All corticosteroids, 7/9 Hydroxychloroquine	1 died, 3 transplanted – 2 died 5 without transplantation, 3/5 improvement of hypoxaemia, 2/5 no improvement 4/5 stable lung function 1/5 slow decline FEV1	(45)
ABCA3 p.L798P p.R1612P	Child with DIP	Antibiotics, supplemental oxygen, exogenous surfactant, methylprednisolone, hydroxychloroquine	No clinical improvement, increased oxygen requirement	(46)
ABCA3 p.R1561Stop	Child with respiratory distress with cyanosis	Antibiotics, surfactant, dexamethasone, inhaled nitric oxide, methylprednisolone, hydroxychloroquine	Short term improvement of respiration, increasing reticular pattern on HRCT, later increased hypoxemia	(47)
ABCA3 large deletion exon 2-5	Child with IRDS	N-CPAP, surfactant therapy, dexamethasone	No effect, died of respiratory failure	(48)
ABCA3 ΔF1203 and c.1375ins15	Child with IRDS	Supplemental oxygen and systemic corticosteroids and diuretics	No effect, died	(49)
ABCA3 p.R20L and c.4483del25	Child with ILD	Prednisolone and supplemental oxygen	No improvement, patient died	(50)

ABCA3 heterozygous p.E292V	Child with cerebropulmonary dysgenetic syndrome	CPAP, mechanical ventilation and antibiotics	Worsening of disease, died	(51)
ABCA3 p.S1116F	Child with RDS	Supplemental oxygen, mechanical ventilation, exogenous surfactant, antibiotics, and inhaled nitric oxide	Despite treatment high oxygen and mechanical ventilation required, died of respiratory failure	(52)
ABCA3 mutation	19 Children### (14 RDS, 4 RDS/PAP, 1 PAP)	16/19 Surfactant	7 No effect, 9 moderate improvement	(4)
		19/19 Systemic steroids	14 No effect, 4 moderate improvement, 1 good improvement	
		9/19Hydroxychloroquine	5 No effect, 3 moderate improvement, 1 good improvement	
		2/19 Azithromycin	2 No effect	
Heterozygous *ABCA3* mutation	16 children#### (9 with RDS, 4 with RDS/PAP, 1 PAP, 2 chILD)	12/16 Surfactant	8 No effect, 4 moderate, transient improvement	
		12/16 Systemic steroids	8 No effect, 3 moderate, transient improvement, 1 good improvement	
		8 /16Hydroxychloroquine	3 No effect, 3 moderate, transient improvement, 2 good improvement	

ABCA3 p.M1227R and Ins1510fs/ter1519	Child with DIP	Macrolides, dexamethasone, mechanical ventilation	no effect	(53)
		Surfactant	Transient effect	
ABCA3 Heterozygous R288K (7 patients) R288K, P766S (heterozygous, 1 patient), R288K, S693L (heterozygous, 1 patient), R288K, Q215K (1 patient)	11 Children with ILD	5 Prednisolone, surfactant, oxygen or corticosteroids	Dead	(54)
		6 Oxygen, aspirin, surfactant, dexamethasone, montelukast, salbutamol, steroids, hydroxychloroquine, azathioprine, azithromycin or antibiotics	Sick-better	
Paediatric case report with *AP3B1* mutation				
AP3B1 p.R509X and p.E659X	Child with HPS2	Oxygen, systemic corticosteroids, G-CSF	Clinically stable PF, age 14 dyspnoea on mild exertion	(55)
Clinical trials				

HPS1†	21 Adults with HPS	11 treated with pirfenidone 10 placebo	Pirfenidone superior to placebo: ΔFVC of 0.46%/month (p=0.587) Restricted group including only patients with initial FVC values >50% of predicted: difference in pulmonary function: ~0.7%/month (p=0.02)	(56)
HPS1 or 4 ‡	35 Adults with HPS	23 treated with pirfenidone, 12 placebo	No statistically significant difference in lung function	(57)

HP: hypersensitivity pneumonitis; PF: pulmonary fibrosis; CPFE: combined pulmonary fibrosis and emphysema; HPS: Hermansky Pudlak syndrome; DIP: desquamative interstitial pneumonia; NSIP: nonspecific interstitial pneumonia; ILD: interstitial lung disease; PAP: pulmonary alveolar proteinosis; CPI: chronic pneumonitis of infancy; IRDS: infant respiratory distress syndrome; ARDS: Acute respiratory distress syndrome; chILD: childhood interstitial lung disease; BAL: bronchoalveolar lavage; (N-) CPAP: (Nasal-) continuous positive airway pressure;

ABCA3 mutations were compound heterozygous or homozygous mutations, unless otherwise stated

*specific mutation not mentioned in the article, diagnosis based on absence of dense bodies platelets under electron microscopy or genetic testing

case also reported in (3)

1 case also reported in (24)

1 case also reported in (3)

1 case also reported in (46)

† 20 of these patients were Puerto Ricans homozygous for a 16-bp duplication in exon 15 of the *HPS1* gene, which leads to a frameshift. The other patient was a Puerto Rican with a 3904-bp deletion in the *HPS3* gene

‡ 33 of these patients were Puerto Ricans homozygous for the known 16-bp duplication in exon 15 of the HPS1 gene. Two patients were non-Puerto Ricans, and the mutations in these patients are not reported

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Search strategy with three search items in Pub Med

(("Idiopathic Interstitial Pneumonias"[Mesh] OR "Lung Diseases, Interstitial"[Mesh:NoExp] OR "Pulmonary Fibrosis"[Mesh] OR interstitial pneumon*[tiab] OR interstitial lung diseas*[tiab] OR pulmonary fibros*[tiab] OR lung fibros*[tiab] OR "Pneumocytes"[Mesh] OR pneumocyte*[tiab] OR alveolar epithelial cell*[tiab]) AND ("Pulmonary Surfactant-Associated Proteins"[Mesh] OR surfactant associated protein*[tiab] OR lung-surfactant protein*[tiab] OR SFTP*[tiab] OR SP-C[tiab] OR SP-A[tiab] OR SPC[tiab] OR SPA[tiab] OR "Pulmonary Surfactant-Associated Protein C"[Mesh] OR "Pulmonary Surfactant-Associated Protein A"[Mesh] OR "ATP-Binding Cassette Transporters"[Mesh] OR ATP binding cassette transporter*[tiab] OR ABC transporter*[tiab] OR ABCA3[tiab] OR "Hermanski-Pudlak Syndrome"[Mesh] OR hermansky pudlak syndrom*[tiab] OR hermanski pudlak syndrom*[tiab] OR HPS[tiab] OR "HPS1 protein, human" [Supplementary Concept] OR "HPS4 protein, human" [Supplementary Concept] OR "Hps1 protein, rat" [Supplementary Concept] OR "Hps1 protein, mouse" [Supplementary Concept] OR "Hps4 protein, mouse" [Supplementary Concept]) AND ("Pharmaceutical Preparations"[Mesh] OR "Physiological Effects of Drugs"[Mesh] OR "Drug Therapy"[Mesh] OR "drug therapy" [Subheading] OR drug*[tiab] OR therap*[tiab] OR pharmaceutic*[tiab] OR medication*[tiab] OR treat*[tiab] OR "drug effects" [Subheading] OR intervention*[tiab]) AND (english[la]))

Search strategy with three search items in Embase

('interstitial pneumonia'/exp OR 'interstitial lung disease'/de OR 'lung fibrosis'/exp OR (interstitial NEXT/1 pneumon*):ab,ti OR (“interstitial lung” NEXT/1 diseas*):ab,ti OR ((pulmonary OR lung) NEXT/1 fibros*):ab,ti OR 'lung alveolus cell'/exp OR pneumocyte*:ab,ti OR (“alveolar epithelial” NEXT/1 cell*):ab,ti) AND ('surfactant associated protein'/exp OR ((‘surfactant associated’ OR ‘lung-surfactant’) NEXT/1 protein*):ab,ti OR SFTP*:ab,ti OR ‘SP-C’:ab,ti OR ‘SP-A’:ab,ti OR SPC:ab,ti OR SPA:ab,ti OR 'surfactant protein C'/exp OR 'surfactant protein A'/exp OR 'ABC transporter'/exp OR ((‘ATP binding cassette’ OR ABC) NEXT/1 transporter*):ab,ti OR ABCA3:ab,ti OR ((‘hermansky pudlak’ OR ‘hermanski pudlak’) NEXT/1 syndrom*):ab,ti OR HPS:ab,ti) AND ('pharmaceutics'/exp OR 'drug effect'/exp OR 'drug therapy'/exp OR 'drug therapy'/lnk OR 'pharmaceutics'/lnk OR drug*:ab,ti OR therap*:ab,ti OR pharmaceutic*:ab,ti OR medication*:ab,ti OR treat*:ab,ti OR intervention*:ab,ti) NOT 'conference abstract'/it AND [english]/lim

Search strategy with two search items Pubmed.

(("Idiopathic Interstitial Pneumonias"[Mesh] OR "Lung Diseases, Interstitial"[Mesh:NoExp] OR "Pulmonary Fibrosis"[Mesh] OR interstitial pneumon*[tiab] OR interstitial lung diseas*[tiab] OR pulmonary fibros*[tiab] OR lung fibros*[tiab] OR "Pneumocytes"[Mesh] OR pneumocyte*[tiab] OR alveolar epithelial cell*[tiab]) AND ("Pulmonary Surfactant-Associated Proteins"[Mesh] OR surfactant associated protein*[tiab] OR lung-surfactant protein*[tiab] OR SFTP*[tiab] OR SP-C[tiab] OR SP-A[tiab] OR SPC[tiab] OR SPA[tiab] OR "Pulmonary Surfactant-Associated Protein C"[Mesh] OR "Pulmonary Surfactant-Associated Protein A"[Mesh] OR "ATP-Binding Cassette Transporters"[Mesh] OR ATP binding cassette transporter*[tiab] OR ABC transporter*[tiab] OR ABCA3[tiab] OR "Hermanski-Pudlak Syndrome"[Mesh] OR hermansky pudlak syndrom*[tiab] OR hermanski pudlak syndrom*[tiab] OR HPS[tiab] OR "HPS1 protein, human" [Supplementary Concept] OR "HPS4 protein, human" [Supplementary Concept] OR "Hps1 protein, rat" [Supplementary Concept] OR "Hps1 protein, mouse" [Supplementary Concept] OR "Hps4 protein, mouse" [Supplementary Concept]) AND (english[la])) NOT (((("Idiopathic Interstitial Pneumonias"[Mesh] OR "Lung Diseases, Interstitial"[Mesh:NoExp] OR "Pulmonary Fibrosis"[Mesh] OR interstitial pneumon*[tiab] OR interstitial lung diseas*[tiab] OR pulmonary fibros*[tiab] OR lung fibros*[tiab] OR "Pneumocytes"[Mesh] OR pneumocyte*[tiab] OR alveolar epithelial cell*[tiab]) AND ("Pulmonary Surfactant-Associated Proteins"[Mesh] OR surfactant associated protein*[tiab] OR lung-surfactant protein*[tiab] OR SFTP*[tiab] OR SP-C[tiab] OR SP-A[tiab] OR SPC[tiab] OR SPA[tiab] OR "Pulmonary Surfactant-Associated Protein C"[Mesh] OR "Pulmonary Surfactant-Associated Protein A"[Mesh] OR "ATP-Binding Cassette Transporters"[Mesh] OR ATP binding cassette

transporter*[tiab] OR ABC transporter*[tiab] OR ABCA3[tiab] OR "Hermanski-Pudlak Syndrome"[Mesh] OR hermansky pudlak syndrom*[tiab] OR hermanski pudlak syndrom*[tiab] OR HPS[tiab] OR "HPS1 protein, human" [Supplementary Concept] OR "HPS4 protein, human" [Supplementary Concept] OR "Hps1 protein, rat" [Supplementary Concept] OR "Hps1 protein, mouse" [Supplementary Concept] OR "Hps4 protein, mouse" [Supplementary Concept]) AND ("Pharmaceutical Preparations"[Mesh] OR "Physiological Effects of Drugs"[Mesh] OR "Drug Therapy"[Mesh] OR "drug therapy" [Subheading] OR drug*[tiab] OR therap*[tiab] OR pharmaceutic*[tiab] OR medication*[tiab] OR treat*[tiab] OR "drug effects" [Subheading] OR intervention*[tiab]) AND (english[la]))

Search strategy with two search items Embase

'interstitial pneumonia'/exp OR 'interstitial lung disease'/de OR 'lung fibrosis'/exp OR (interstitial NEXT/1 pneumon*):ab,ti OR ('interstitial lung' NEXT/1 diseas*):ab,ti OR ((pulmonary OR lung) NEXT/1 fibros*):ab,ti OR 'lung alveolus cell'/exp OR pneumocyte*:ab,ti OR ('alveolar epithelial' NEXT/1 cell*):ab,ti AND ('surfactant associated protein'/exp OR (('surfactant associated' OR 'lung-surfactant') NEXT/1 protein*):ab,ti OR sftp*:ab,ti OR 'sp-c':ab,ti OR 'sp-a':ab,ti OR spc:ab,ti OR spa:ab,ti OR 'surfactant protein c'/exp OR 'surfactant protein a'/exp OR 'abc transporter'/exp OR (('atp binding cassette' OR abc) NEXT/1 transporter*):ab,ti OR abca3:ab,ti OR (('hermansky pudlak' OR 'hermanski pudlak') NEXT/1 syndrom*):ab,ti OR hps:ab,ti) NOT 'conference abstract'/it AND [english]/lim AND ('case report'/de OR 'clinical article'/de OR 'clinical trial'/de OR 'cohort analysis'/de OR 'comparative study'/de OR 'controlled study'/de OR 'human'/de OR 'human tissue'/de OR 'major clinical

study'/de OR 'retrospective study'/de) AND 'article'/it NOT ('interstitial pneumonia'/exp OR 'interstitial lung disease'/de OR 'lung fibrosis'/exp OR (interstitial NEXT/1 pneumon*):ab,ti OR ('interstitial lung'NEXT/1 diseas*):ab,ti OR ((pulmonary OR lung) NEXT/1 fibros*):ab,ti OR 'lung alveolus cell'/exp OR pneumocyte*:ab,ti OR ('alveolar epithelial' NEXT/1 cell*):ab,ti AND ('surfactant associated protein'/exp OR (('surfactant associated' OR 'lung-surfactant') NEXT/1 protein*):ab,ti OR sftp*:ab,ti OR 'sp-c':ab,ti OR 'sp-a':ab,ti OR spc:ab,ti OR spa:ab,ti OR 'surfactant protein c'/exp OR 'surfactant protein a'/exp OR 'abc transporter'/exp OR (('atp binding cassette' OR abc) NEXT/1 transporter*):ab,ti OR abca3:ab,ti OR (('hermansky pudlak' OR 'hermanski pudlak') NEXT/1 syndrom*):ab,ti OR hps:ab,ti) AND ('pharmaceutics'/exp OR 'drug effect'/exp OR 'drug therapy'/exp OR 'drug therapy'/lnk OR 'pharmaceutics'/lnk OR drug*:ab,ti OR therap*:ab,ti OR pharmaceutic*:ab,ti OR medication*:ab,ti OR treat*:ab,ti OR intervention*:ab,ti) NOT 'conference abstract'/it AND [english]/lim)

Include: antibody and cells used for ILD research

Exclude: siRNA/shRNA, overexpression gene, acute respiratory distress syndrome, cell therapy, only ventilation/oxygen therapy, bronchoalveolar lavage or lung transplantation