Supplementary table 3: Highlights on treatment changes after the diagnostic evaluation.

Subject	Treatment after clinical evaluation & NIT*	Final opinion after Invasive tests/ specific immune tests	Treatment after diagnostic evaluation*	Additional notes
1	Systemic steroids (IV Pulse steroid therapy)	Specific Dx **(by LBx)**: chronic HP	Exposure elimination +oral steroids (tapered over 9 months)	Complete resolution at the 9 months follow-up†
2	Antibiotics only	Specific Dx (**by LBx**): chronic HP	Exposure elimination +oral steroids (tapered over 6 months)	Complete resolution at the 6 months follow-up†
3	Antibiotics only	Specific Dx (**by LBx**): subacute HP	Exposure elimination + IV pulse steroids followed by oral steroids (tapered over 12 months)	Complete resolution at the 12 months follow-up†
4	Antibiotics + Systemic steroids (IV Pulse steroid therapy)	Specific Dx (**by LBx +DHR**): CGD	*Prophylactic antimicrobials (trimethoprim sulfamethoxazole & voriconazole) +continued low dose systemic steroids *Mycophenolate mofetil was added#	Frequent exacerbations requiring parenteral antibiotics, antifungals & systemic steroids for stabilization
5	Antibiotics + Systemic steroids (IV Pulse steroid therapy)	Specific Dx **(by DHR)**: CGD	Prophylactic antimicrobials (trimethoprim sulfamethoxazole and itraconazole) + continued low dose systemic steroids	
6	Antibiotics + Systemic steroids (IV Pulse steroid therapy)	Specific Dx (**by LBx+ DHR**): CGD		
7	Systemic steroids (IV Pulse steroid therapy)	Specific Dx (**by LBx**): non-CVID related GLILD	Systemic steroids Mycophenolate mofetil was added	Frequent exacerbations requiring pulse steroids
8	Systemic steroids (IV Pulse steroid therapy)	Specific Dx (**by LBx**): IP (fibrotic NSIP-honey comb lung)	No change (continued on systemic steroids, however, they were shifted to the oral route)	Hydroxychloroquine and azithromycin were added on follow-up due to frequent exacerbations
9	Combination therapy (pulse steroids+ Azithromycin+ hydroxychloroquine)	Specific Dx (**by LBx**): IP (NSIP/DIP)	No change (same treatment lines were continued after the diagnostic evaluation; However, the steroids were shifted to the oral route)	Oral steroids dose was adjusted (increased or decreased) according to the clinical condition
10	No specific treatment	Specific Dx (**by LBx**): IP (NSIP)	Monotherapy with Azithromycin	Improved clinically: treatment was stopped at the 3 months follow-up
11	Combination therapy (Oral steroids+ Azithromycin)	Specific Dx (**by LBx**): IP (BPIP)	No change (same treatment lines were continued after the diagnostic evaluation)	Improved clinically: treatment was gradually tapered and stopped
12	Systemic steroids (IV Pulse steroid therapy)	Suggestive Dx (clinical+ NIT): familial	Oral & inhaled steroids +Azithromycin	Frequent exacerbations requiring escalation in

Subject	Treatment after clinical evaluation & NIT*	Final opinion after Invasive tests/ specific immune tests	Treatment after diagnostic evaluation*	Additional notes
		ILD of unidentified aetiology		oral steroid dose
13	Combination therapy (inhaled and systemic steroids, hydroxychloroquine and Azithromycin)	Specific Dx (**by LBx**): SAD with background IP	No change (same treatment lines were continued after the diagnostic evaluation)	Frequent exacerbations requiring pulse steroids
14		Specific Dx (**by LBx**): SAD with background IP		Improved clinically: Treatment was gradually tapered and stopped after 12 months
15		Specific Dx (**by LBx**): SAD with background IP		Improved clinically: Treatment was gradually tapered and stopped after 3 months
16	Systemic steroids (IV Pulse steroid therapy followed by oral steroids)	Specific Dx (**by LBx+ further tests**): Systemic sclerosis	Steroids were tapered and Mycophenolate mofetil was added	
17	Frequent blood transfusion	Specific Dx **(by BAL)**: IPH	Oral steroids (tapered till remission)	
18	Frequent blood transfusion	Specific Dx **(by BAL)**: IPH	Oral steroids (tapered till remission)	
19	No specific treatment	Suggestive Dx **(by radiology)**: LCH	Death shortly after presentation	
20	No specific treatment	Suggestive Dx **(by radiology)**: LCH	Dropped out	
21	Antibiotics + Antituberculous drugs	Specific Dx **(clinical+ NIT)**: post tuberculous chILD	Oral steroids (tapered till remission) ^	
22	Antibiotics	Specific Dx **(by LBx)**: post infectious chILD (OP)	Oral steroids (tapered till remission)	

*We used the European protocols for diagnosis and initial management of chILD [2] as a guide for treatment, however, treatment plans were adjusted on case by case basis according to the clinical presentation, Fan score and the specific cause of chILD identified following the diagnostic evaluation. After the completion of diagnostic evaluation, we followed up the patients every 1-3 months at the pediatric pulmonology special clinic (follow up parameters included symptoms, signs, hypoxemia at rest and after exercise). Also, follow up echocardiography was done if the patient was originally presented with pulmonary hypertension, or if there was a clinical deterioration on follow up. Pulmonary function tests were also performed once yearly to evaluate disease progression. We are still following up enrolled subjects till the present time. † Complete resolution of clinical symptoms, signs with resolution of the radiological signs and normalization of pulmonary function tests on follow up. NIT: non-invasive tests; IV: intravenous; Dx: diagnosis; LBx: lung biopsy; BAL: bronchoalveolar lavage; HP: hypersensitivity pneumonitis; DHR: dihydrorhodamine test; TB: tuberculosis; CGD: chronic granulomatous disease; GLILD: granulomatous lymphocytic interstitial lung disease; NSIP: non-specific interstitial pneumonia; DIP: desquamative interstitial pneumonia; BPIP: bronchiolocentric pattern of interstitial pneumonias; ILD: interstitial lung disease; SAD: small airway disease; IP: interstitial pneumonia; IPH: idiopathic pulmonary hemosiderosis; LCH: Langerhans cell histiocytosis; Op: organising pneumonia. #Case 4: immunosuppression was started despite diagnosis of primary immune deficiency due to admission with frequent non-infective exacerbations refractory to pulse steroid therapy. ^ Subject 21: presented initially with a 6 months history of night fever and sweats. Initial CT showed bilateral upper lobar GGO, air trapping and mediastinal lymphadenopathy. Further investigations revealed positive tuberculin test (20 mm) and BALF culture showed positive M. Tuberculosis. Diagnosis of pulmonary TB was confirmed and she received antituberculous therapy. On further assessment she continued to have progressive dyspnea and dry cough one year post anti-TB treatment. Chest imaging was repeated and showed GGO, reticulations, atelectatic bands and calcified nodule. She continued to have progressive dyspnea and developed hypoxemia at rest. She fulfilled diagnostic criteria of chILD (persistent symptoms, signs, radiological changes and hypoxemia) after resolution of infection. Diagnosis of postinfectious chILD was made and she was started on systemic steroids with marked clinical improvement.