



Familial pulmonary fibrosis: a heterogeneous spectrum of presentations

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Chart S1. Radiological and pathological features of index cases and their affected relatives.

Index case number	Index case		HRCT pattern ^a	Relative	
	HRCT pattern ^a	Histological pattern		Histological pattern	Chest X-ray
1	NSIP	DAD progressing to OP	Indeterminate: 1 Incipient ILD: 4 Other than IPF: 1	NA	Fibrosing ILD: 1
2	UIP	NA	NSIP: 2 Probable: 1 Indeterminate: 1	uILD: 1	NA
3	Inconsistent with ILD	NA	NA	NA	Fibrosing ILD: 1
4	NSIP	IPBA	Inconsistent with ILD: 2	NA	NA
5	UIP	NA	NA	NA	Fibrosing ILD: 1
6	NSIP	NA	Inconsistent with ILD: 2	NA	NA
7	Other than IPF	NA	Inconsistent with ILD: 1 UIP: 1	NA	NA
8	Inconsistent with ILD	NA	Inconsistent with ILD: 1	NA	NA
9	Inconsistent with ILD	NA	Inconsistent with ILD: 1	NA	NA
10	NSIP	NA	UIP: 1 Incipient ILD: 1	IPBA	NA
11	Inconsistent with ILD	NA	Inconsistent with ILD: 1	NA	NA
12	NSIP	NA	NA	NA	Fibrosing ILD: 1
13	Other than IPF	NA	UIP: 1	NA	NA
14	NSIP	IPBA	UIP: 1	NA	NA
15	Other than IPF	NA	UIP: 1	NA	NA
16	UIP	NA	HP	NA	NA
17	Other than IPF	NA	Inconsistent with ILD: 1	NA	NA
18	Indeterminate	NA	NA	NA	Fibrosing ILD: 1
19	Inconsistent with ILD	NA	NA	NA	Fibrosing ILD: 1
20	Inconsistent with ILD	NA	NSIP: 1 Indeterminate: 1	NA	NA
21	HP	IPBA	NSIP: 1 Indeterminate: 1	NA	NA
22	HP	NA	Inconsistent with ILD: 1	NA	NA
23	UIP	NA	UIP: 1	NA	NA
24	UIP	NA	UIP: 1	NA	NA
25	NSIP	NA	Incipient ILD: 1	NA	NA
26	NSIP	NA	Inconsistent with ILD: 1	NA	NA
27	NSIP	NA	NA	NA	Fibrosing ILD: 1
28	Indeterminate	NA	Incipient ILD: 1	NA	NA
29	UIP	NA	NA	NA	Fibrosing ILD: 1
30	Inconsistent with ILD	NA	HP: 1	IPBA	NA
31	Inconsistent with ILD	IPBA	Inconsistent with ILD	NA	NA
32	Inconsistent with ILD	NA	Probable	NA	NA
33	Inconsistent with ILD	NA	NA	NA	Fibrosing ILD: 1
34	Indeterminate	uILD	NA	NA	Fibrosing ILD: 1
35	Inconsistent with ILD	NA	NA	NA	Fibrosing ILD: 1

NSIP: nonspecific interstitial pneumonia; DAD: diffuse alveolar damage; OP: organizing pneumonia; ILD: interstitial lung disease; NA: not available; UIP: usual interstitial pneumonia; uILD: unclassifiable interstitial lung disease; IPBA: interstitial pneumonia with bronchiolocentric accentuation; IPF: idiopathic pulmonary fibrosis; and HP: hypersensitivity pneumonitis. ^aNSIP: findings consistent with a diagnosis other than IPF and suggestive of NSIP; UIP: typical UIP; Probable: probable UIP; Indeterminate: indeterminate UIP; Other than IPF: CT findings consistent with a diagnosis other than IPF; HP: findings consistent with a diagnosis other than IPF and suggestive of HP.