

Table 1S Complete Flow Cytometry Results at Diagnosis

Flow Cytometry Results: Mean absolute values, cells/mcL	Pediatric onset cohort (n=91)	Adult onset cohort (n=107)
Mean absolute CD3+ (All lymphocytes) (Normal adult range 550- 2202)	1737 (N=41)	1264 (N=65)
Mean absolute CD19+ (B cells) (Normal adult range 91-409)	254 (N=39)	210 (N=66)
Mean absolute CD21- (B cell subset) (Normal adult range 0.3-22)	27 (N=5)	18 (N=49)
Mean absolute switched memory B cells CD27+IgM- IgD- (B cell subset) (Normal adult range 7-61)	3 (N=4)	9 (N=46)
Mean absolute CD4+ (T helper cells) (Normal adult range 365- 1437)	394 (N=42)	796 (N=65)
Mean absolute CD8+ (Cytotoxic T cells) (Normal adult range 199-846)	733 (N=41)	440 (N=65)
Mean absolute CD16+ (Natural killer cells) (Normal adult range 59-513)	143 (N=36)	151 (N=22)

Note: Due to incomplete data sets for all laboratory parameters the number of patients with data is represented in parenthesis as above.

Table 2S Laboratory values at most recent clinic visit or hospitalization

Mean immunoglobulin levels in mg/dL (range)	Pediatric onset cohort (n=91)	Adult onset cohort (n=107)
IgG (Normal adult range 588 - 1573)	849 (47-1830)	914 (11-1900)
IgA (Normal adult range 46 - 287)	9 (0-150)	26 (0-319)
IgM (Normal adult range 57 - 237)	42 (0-631)	56 (0-653)

Fig S1

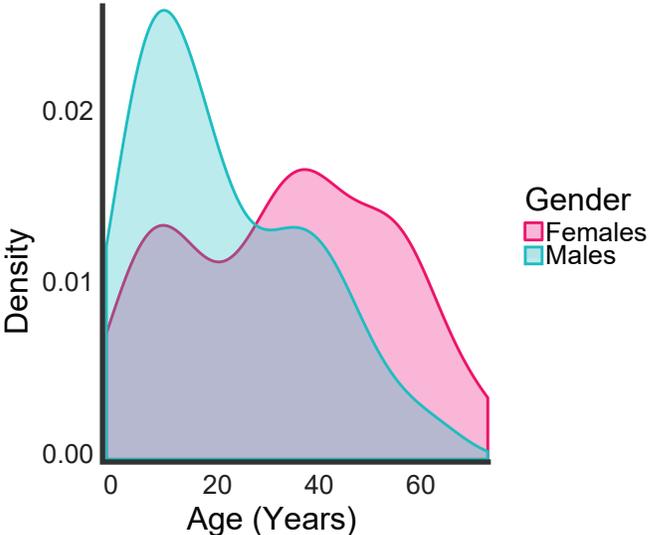
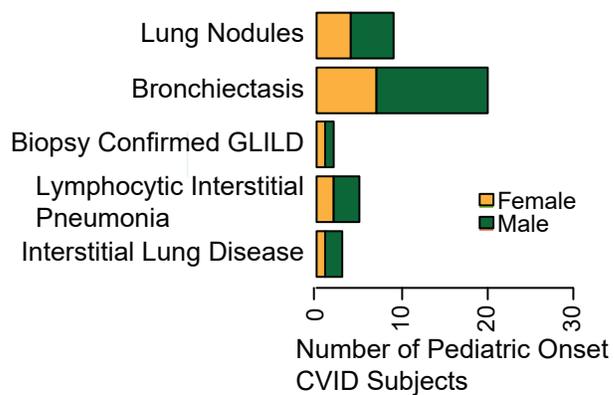


Fig 2S

a

Features of lung disease



b

Association of individual lung features with deceased status

