Supplementary appendix

Collection of baseline data

We used standardised forms to record the following information: demographic data; medical history, including age at sarcoidosis onset; physical findings; World Health Organisation functional class; routine blood test results; 6-minute walking distance(1); and most recent PFT results before lung transplantation including forced vital capacity (FVC), forced (2) expiratory volume in 1 s (FEV1), and diffusing capacity for carbon monoxide (DLCO), measured according to established protocols and reported as percent of predicted values.

PH was sought during right heart catheterisation and defined as mean pulmonary artery pressure >20 mm Hg, pulmonary capillary wedge pressure ≤15 mm Hg, and pulmonary vascular resistance >3WU in the absence of other known causes of PH (3). Severe PH (sPH) was defined as follow: mean pulmonary artery pressure ≥35 mm Hg or as mean pulmonary artery pressure ≥25 mm Hg with a cardiac index ≤ 2.0L/min/m^2(4).

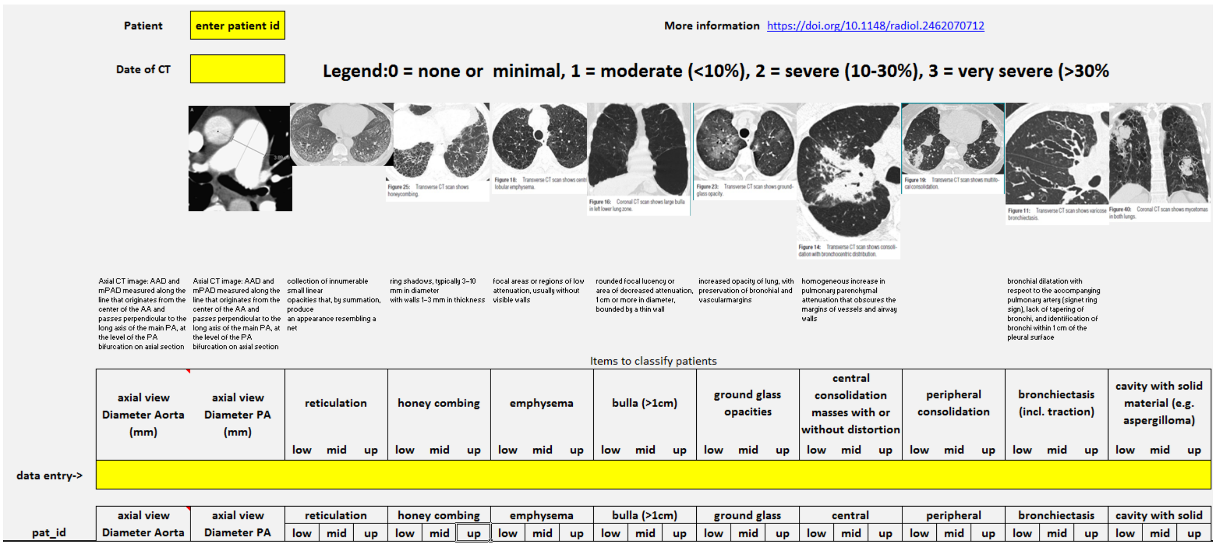
Thoracic CT protocol and review

All patients were scanned in the supine position from the lung apices to the lung bases at full suspended inspiration using standard exposure parameters (90 mA and 120kVp). All images were anonymised and reviewed independently by two thoracic radiologists or one thoracic radiologist and one pulmonologist working independently of each other and using standard window settings for lung parenchyma visualisation.

The readers had no knowledge of pulmonary function data or other clinical indicators of disease severity. The presence and extent of the following patterns, based on the Fleischner Society's glossary of terms for thoracic imaging (5) with some minor modifications were

evaluated for the entirety of both lungs: 1) fibrosis defined as reticular pattern associated with architectural distortion with or without honeycombing and/or traction bronchiectasis 2) ground glass opacity defined as hazy increased opacity of lung, with preservation of bronchial and vascular margins, 3) other, defined as patterns of disease not covered by (1) or (2) and including perihilar and peripheral pulmonary masses, emphysema, bullae, and cavity containing solid material (aspergilloma) (6). The lungs were divided into three zones (upper, middle, and lower), each of which was evaluated separately. The upper lung zone was defined as the part of the lung above the level of the tracheal carina, the lower lung zone as the part below the level of the inferior pulmonary vein, and the middle lung zone as the part between the upper and lower zones. When abnormal findings were present, the extent of lung involvement by each pattern was scored visually and independently for each of the three lung zones as follows: absent=0, <10%=1, 10 to 30%=2, and >30%=3. Involvement by each pattern was then obtained by summing the scores for the three zones. The main pulmonary artery diameter and ascending aorta diameter ratio were also assessed.

Supplemental Figure S1: CT scoring sheet.



Patient management

Most of the patients with sPH were transplanted with intraoperative veno-arterial ECMO. ECMO was switched prophylactically from its central location to peripheral cannulation after implantation of the lungs. Patients were cannulated in the groin, and the ECMO device used intraoperatively was connected. A Swan-Ganz catheter and transthoracic echocardiography were used to measure pulmonary artery pressures, assess cardiac function, and monitor the adequacy of fluid management and adrenergic support. ECMO flow was kept at a low level, usually about half the normal cardiac output. ECMO remained in place until the patients were haemodynamically stable and had normal chest X-ray findings, adequate oxygenation (fraction of inspired oxygen <0.5), a low ventilation pattern and, most importantly, a normal fluid balance, i.e., removal of the excessive fluid load (no oedema or ascites).

Primary graft dysfunction (PGD) grade was assessed prospectively, using the International Society for Heart and Lung Transplantation criteria, defined by the Pa_{O2}/Fi_{O2} ratio and the presence of infiltrates within the allograft or allografts (7).

Post-transplantation immunosuppression and induction therapy were given according to local guidelines at each centre. All patients received life-long *Pneumocystis jirovecii* pneumonia prophylaxis with cotrimoxazole. Valganciclovir for cytomegalovirus prophylaxis was given according to local guidelines. Monitoring transbronchial biopsies were obtained routinely or as clinically indicated depending on the standard protocol at each centre. Patients with allograft dysfunction were investigated for acute cellular rejection, lymphocytic bronchiolitis/neutrophilic reversible allograft dysfunction, and airway injury caused by infection/colonisation. CLAD was diagnosed based on international criteria when FEV_1 and/or FVC declined to ≤80% of the best postoperative value (8) during routine outpatient assessments after a minimum of 3 months post-lung transplantation. Comprehensive PFTs

including spirometry and lung volume measurements, HRCT of the chest, and bronchoscopy with bronchoalveolar lavage and transbronchial biopsy were performed to look for causes of lung allograft dysfunction, including persistent acute rejection, azithromycin-responsive allograft dysfunction, infection, anastomotic stricture, and sarcoidosis recurrence.

Figure S3: Main features of the overall population according to inclusion/exclusion status

	Overall n = 166	Included n = 112	Excluded n =54	*P** value
Male, n (%)	102 (61)	71 (64)	31 (57)	0.57
Recipient age, med [IQR]	51 [44 - 57]	52 [46 - 59]	46 [40 - 53]	<0.01
Body mass index, kg/m² med [IQR]	22 [20 - 26]	23 [20 - 26]	22 [16 - 34]	0.51
Caucasian, n (%)	141 (85)	92 (82)	49 (91)	0.49
Smoker, med [IQR]	5 [0 - 18]	6 [0 - 19]	1.5 [0 - 16]	0.26
Extrathoracic sarcoidosis, 0/1/2/3, n (%) n/165	137 (83)/26 (15) /1 (1)/1 (1)	90 (80)/20 (18) /1 (1)/1 (1)	47 (89)/6 (11)/0/0	0.71
History of pulmonary aspergillosis, n (%), n / 155	25 (18)	18 (19)	7 (16)	0.87
Blood group, O/A/B/AB, %	40 / 43 / 14 / 3	39 / 44 / 16 / 1	43 / 43 / 11 / 3	0.54
Lung transplantation delay, d, med [IQR]	88 [25 - 283]	92 [27 - 299]	76 [19 - 245]	0.29
Lung transplantation procedure DLT/HLT/SLT, n (%)	139 (84) / 6 (4) / 21 (12)	101 (90) / 3 (3) / 8 (6)	38 (70) / 3 (6) / 13 (24)	0.04
High-emergency transplant programme, n (%)	27 (16)	17 (15)	10 (19)	0.74
Cardiopulmonary bypass, n (%), n / 161	71 (44)	54 (50)	17 (33)	0.07
Right ischaemic time, min, med [IQR], n / 86	288 [238 – 378]	300 [240 – 372]	285 [204 – 390]	0.69
Left ischaemic time, min, med [IQR], n / 65	360 [300 – 401]	360 [300 – 395]	376 [312 – 405]	0.61
Induction, n (%), n / 123	49 (38)	28 (34)	21 (48)	0.18
Dialysis during intensive care unit stay n (%), n / 144	21 (14)	14 (14)	7 (15)	0.99
Primary graft dysfunction score Grade 3 at 72 hours, n (%) , n / 159	33 (21)	24 (22)	9 (19)	0.84
Ventilation time during intensive care unit stay, med [IQR], n / 153	3 [1 - 17]	2 [1 - 19]	3 [1 - 11]	0.95
Haemothorax, n (%), n / 127	26 (20)	16 (18)	10 (26)	0.41
Pulmonary sarcoidosis recurrence, n (%), n / 119	11 (9)	11 (14)	0	0.03
Chronic lung allograft dysfunction at last follow-up, n (%), n / 159	61 (38)	33 (31)	28 (54)	0.01
In-hospital mortality, n(%)	22 (13)	18 (16)	4 (7)	0.35
Retransplantation, n (%)	4 (2)	3 (3)	1 (2)	0.67

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