

Value of serial CT measurements in a small series of patients with shrinking lung syndrome (SLS)

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Background

- SLS - rare pulmonary manifestation of SLE – up to 1.53% of patients^{1,2,3}
- SLS - presenting with gradual decrease in lung volumes³ and breathlessness
- Lung height corrected for body habitus (LH) is a CT marker of lung volume⁴
- LH has shown good correlation with pulmonary function tests (PFTs) in idiopathic pulmonary fibrosis (IPF)⁴

Aim

- Case series of patients with SLS
- Concurrent serial PFTs and CT parameters
- **Assess the utility of LH in SLS**

Method

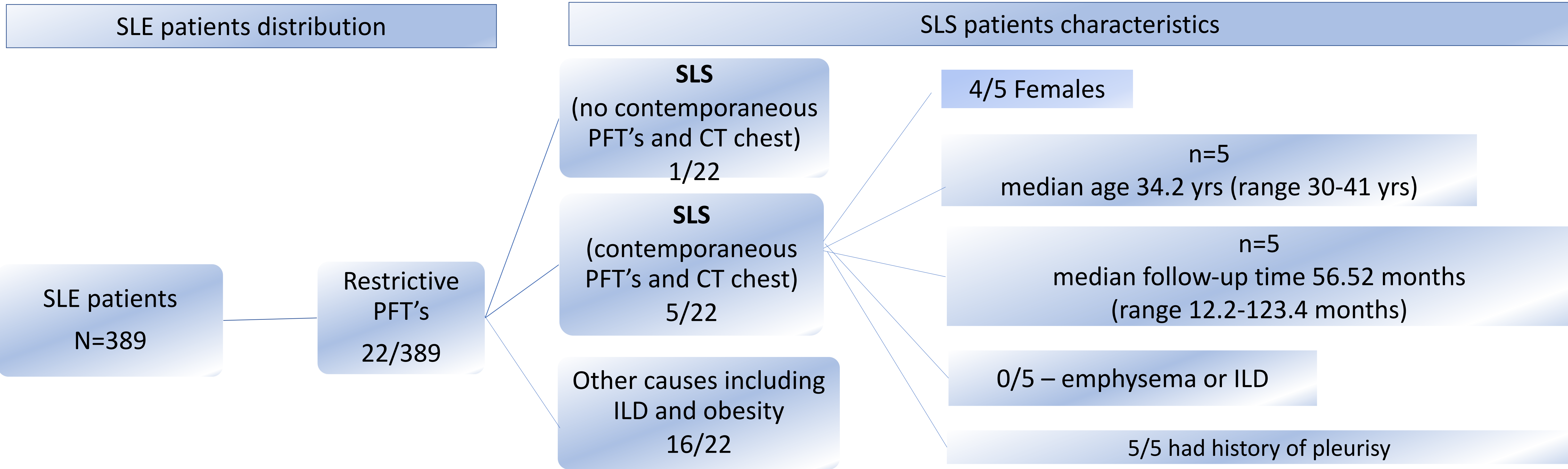
- Retrospective analysis - all SLE patients
- Between 2007-2020
- King's College Hospital London - tertiary centre
- Identified all patients - multidisciplinary diagnosis of SLS

- Contemporaneous CT and PFTs - evaluated longitudinally

Experienced thoracic radiologist - blinded to all clinical data:

- LH Measurements made on sagittal reformats and standardised at mid clavicular line , in inspiration (round trachea)
- LH - both lungs and averaged - single value for each patient
- LH - corrected for body habits = dividing LH by vertebral height (tallest vertebrae in the lower thoracic/upper lumbar spine)
- Scans scored for co-existing emphysema and ILD

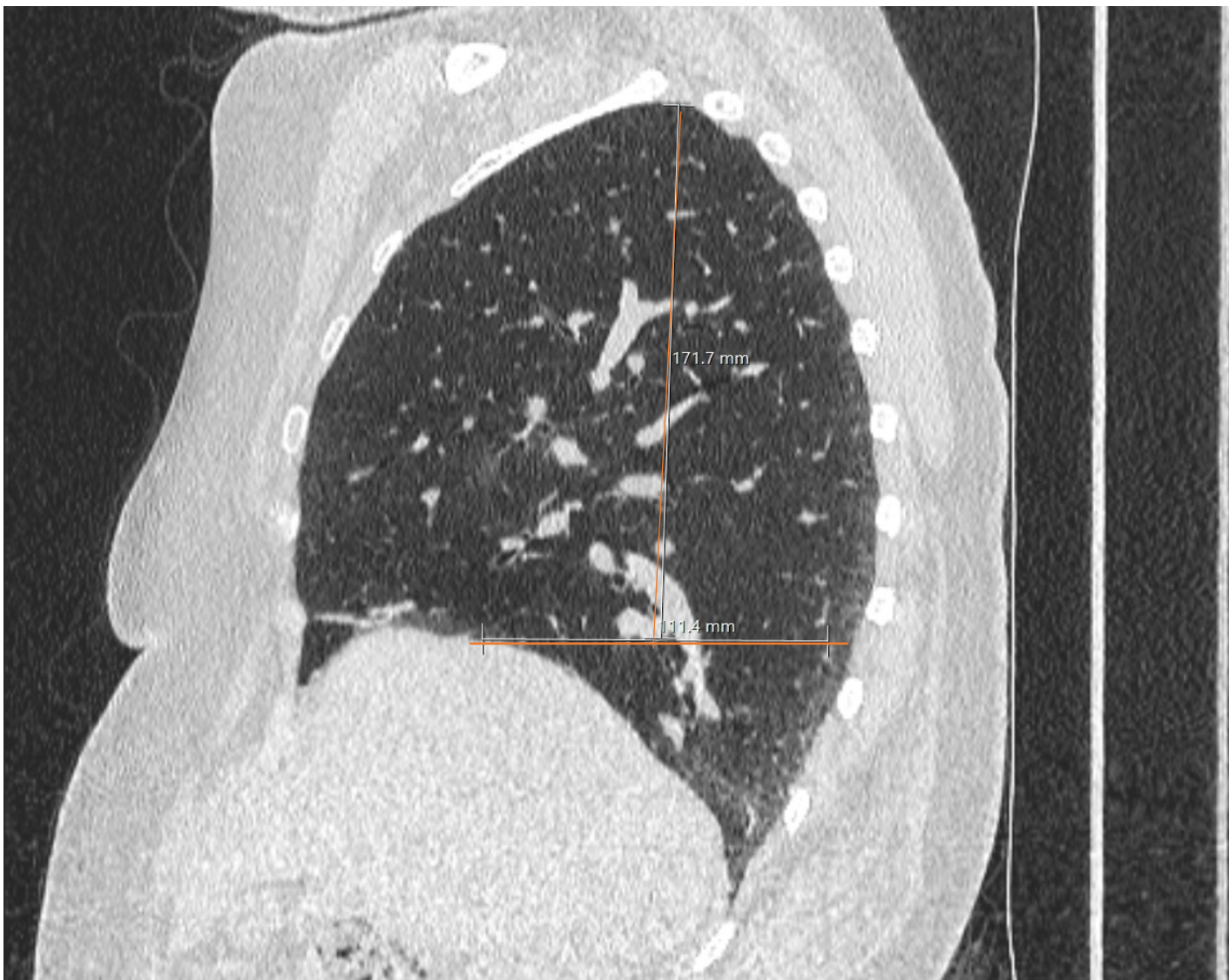
Results



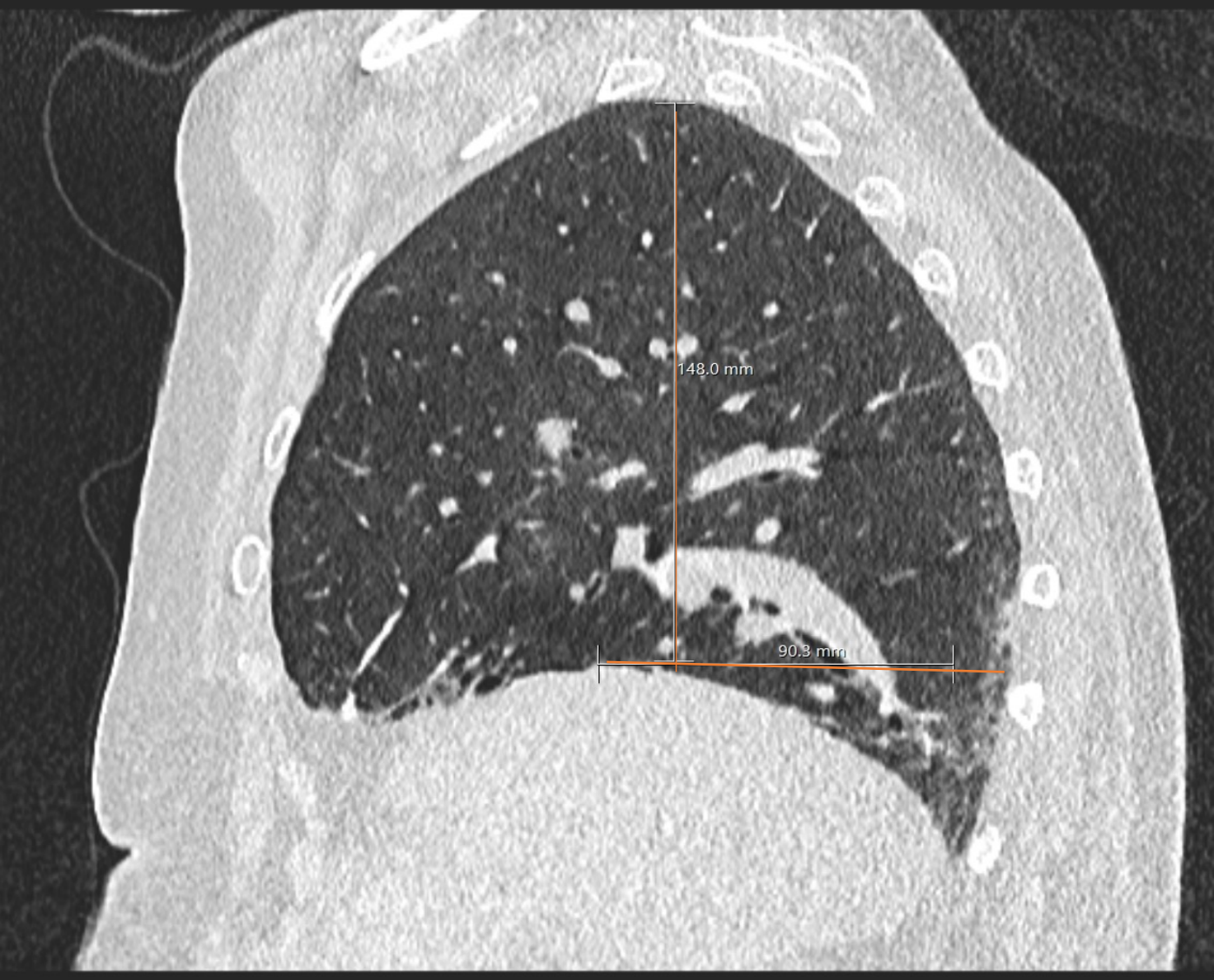
Lung height corrected for body habitus (LH) and Pulmonary Function Tests (PFTs) measurements and longitudinal difference

SLS patients (n=5) Sex/Age	LH corrected for body habitus				FVC (L)				BMI		Treatment
	1 st measurement LH1	2 nd measurement LH2	LH difference (LH1-LH2)	LH percentage change	1 st measurement FVC1 (L)	2 nd measurement FVC2 (L)	FVC difference (FVC1-FVC2)	FVC percentage change	1 st measurement	2 nd measurement	
F/41	7.20	5.91	-1.30	-17.98%	1.79	1.55	-0.24	-13.41%	28.2	31.2	Ritux,Cyclo MMF
F/35	6.50	6.36	-0.14	-2.20%	1.55	2.01	+0.46	+29.67%	21.8	28.7	MMF, HQ
M/30	5.68	5.42	-0.26	-4.58%	3.20	3.17	-0.03	-0.94%	33.6	45.5	Mtx; HQ
F/33	6.87	6.46	-0.41	-6.01%	3.00	2.33	-0.67	-22.33%	20.65	22.6	Aza
F/32	7.50	6.55	-0.95	-12.67%	3.43	1.78	-1.65	-48.10%	33.5	33.27	HQ, MMF

FVC- forced vital capacity; Ritux- Rituximab; Cyclo- Cyclophosphamide; MMF- Mycophenolate mofetil; HQ- Hydroxychloroquine; Mtx – Methotrexate; Aza- Azathioprine



LH in SLS - Baseline CT - 2017



LH in SLS - Interval CT - 2020

Conclusion

- Similar decreasing trend for LH and PFTs in 80% (4/5 patients)
- Serial CT LH measurement may be a useful diagnostic and monitoring tool in SLS
- Further validation in a larger cohort is needed
- Limitations: - Small retrospective study
 - No standardized CT protocol
 - Confounding factors

References:

1. Borrell H et al. Shrinking lung syndrome in systemic lupus erythematosus: A case series and review of the literature. *Medicine (Baltimore)*. 2016;95(33):e4626. doi:10.1097/MD.00000000000004626
2. Deeb M et al. Shrinking lung syndrome in systemic lupus erythematosus: a single-centre experience. *Lupus*. 2018 Mar;27(3):365-371. doi: 10.1177/0961203317722411. Epub 2017 Jul 31. PMID: 28758573.
3. Karim MY et al. Presentation and prognosis of the shrinking lung syndrome in systemic lupus erythematosus. *Semin Arthritis Rheum*. 2002 Apr;31(5):289-98. doi: 10.1053/sarh.2002.32555. PMID: 11965593
4. Robbie H et al. Visual and Automated CT Measurements of Lung Volume Loss in Idiopathic Pulmonary Fibrosis. *AJR Am J Roentgenol*. 2019 Aug;213(2):318-324. doi: 10.2214/AJR.18.20884. Epub 2019 May 7. PMID: 31063425.