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Abstract Group: 1.5. Diffuse Parenchymal Lung Disease

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Title: Lung function progression in Langerhans cell histiocytosis

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Body: LCH is a rare, multisystem dendritic cell disorder commonly involving the lungs. The natural history is variable with little lung function outcome data. In our database of 92 patients referred to AC for treatment, 51 patients were male; mean age at diagnosis was 31 (range 1-77) years (y). 17/92 patients, 10 male, mean age 31 y, had primary lung disease (PLLCH). 14/75 had systemic LCH with lung involvement (SLLCH). All PLLCH had smoked with mean 13 pack-y; 9/17 continued smoking after diagnosis. Initial lung function (n=15) showed mean %predicted FEV_1 76 (34-113)%, VC 81 (53-114)%, TLCO 67 (19-108)%. 2 patients had obstructive, 3 restrictive, and 3 mixed lung disease. 9/14 SLLCH patients were male, mean age 25y, 10 had smoked; mean 12 pack-y, 7 continued smoking after diagnosis. Initial lung function (n=9) showed mean FEV_1 83 (72-105)%pred, VC 89 (72-104)%, TLCO 76 (49-106)%. In PLLCH (n=14) at mean follow up of 8 (range 1-16) y mean FEV_1 was 70 (30-103)%pred, VC was 84 (51-109)%, TLCO was 63 (17-105)%. Mean change in FEV_1 was -7 (-39 to +12)%, in VC was 1 (-19 to +14)%, in TLCO was -10 (-49 to +29)%. 11/17 patients received treatment. To date 3 have died, 10 are in remission and 4 have active disease. In SLLCH (n=6) at mean follow up of 6 (2-22) y mean FEV_1 was 83 (66-101)%pred, VC was 92 (79-103)%, TLCO was 93 (82-94)%. Mean change in FEV_1 was -3 (-9 to +5)%, in VC was -1 (-11 to +12)% and in TLCO was 14 (-2 to +45)%. 12/14 patients received treatment. To date 1 has died, 8 are in remission and 5 have active disease. LCH is rare but in this relatively small, selected series lung function is worse and declines more in patients who present with primary lung involvement than in those with lung involvement in systemic LCH.