

SUPPLEMENTARY INFORMATION

CLINICAL OUTCOMES AND SURVIVAL FOLLOWING LUNG TRANSPLANTATION IN PATIENTS WITH PULMONARY LANGERHANS CELL HISTIOCYTOSIS

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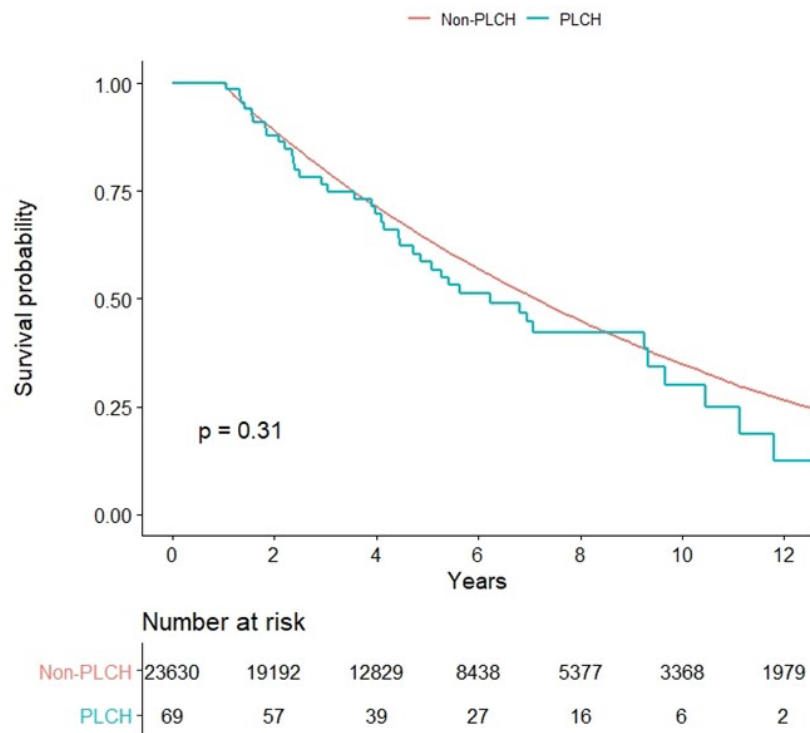
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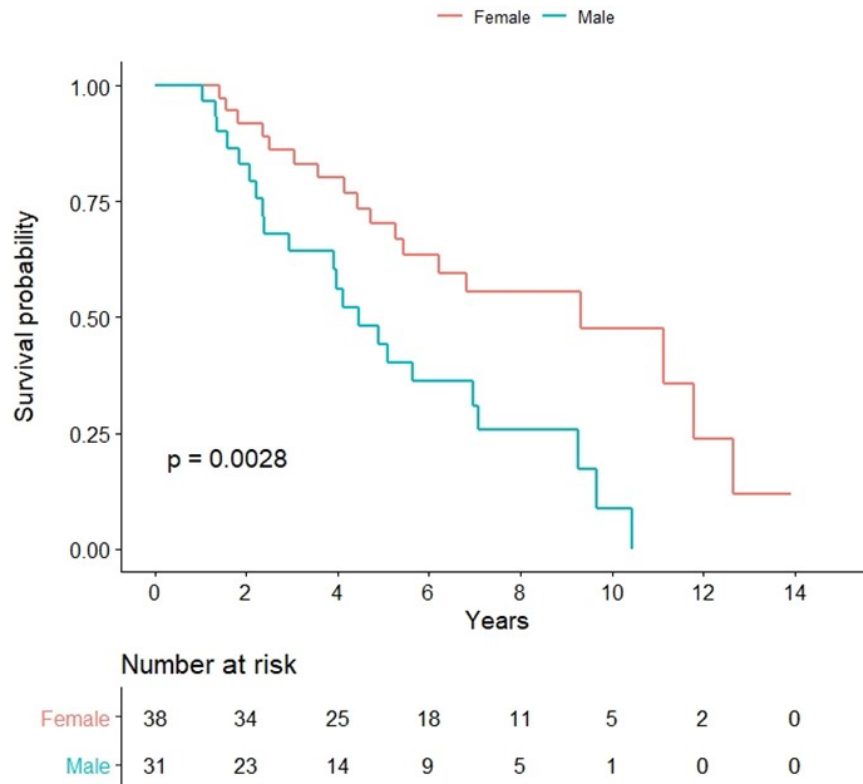
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Figure S1: Kaplan-Meier curve demonstrating probability of conditional survival, assessed after excluding patients who had died in the first 12 months following LT, in PLCH patients compared with non-PLCH patients.



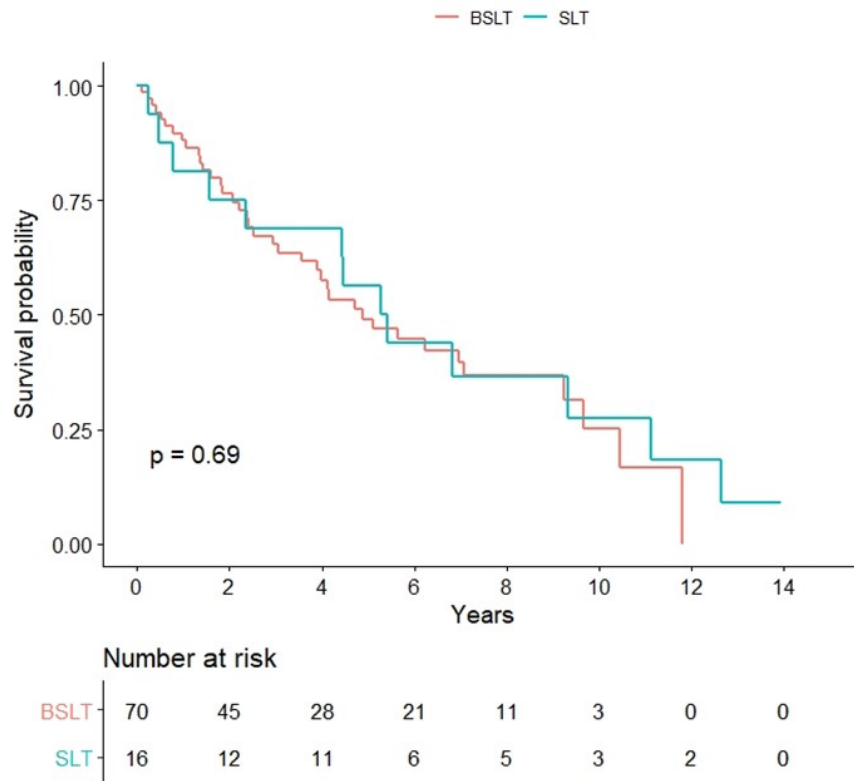
The comparisons were performed using log-rank test. There was no statistically significant difference between the two groups: median conditional survival 6.2 years in PLCH vs. 7.1 years in non-PLCH patients ($p=0.31$).

Figure S2: Kaplan-Meier curve demonstrating probability of conditional survival, assessed after excluding patients who had died in the first 12 months following LT, in female PLCH patients compared with male patients.



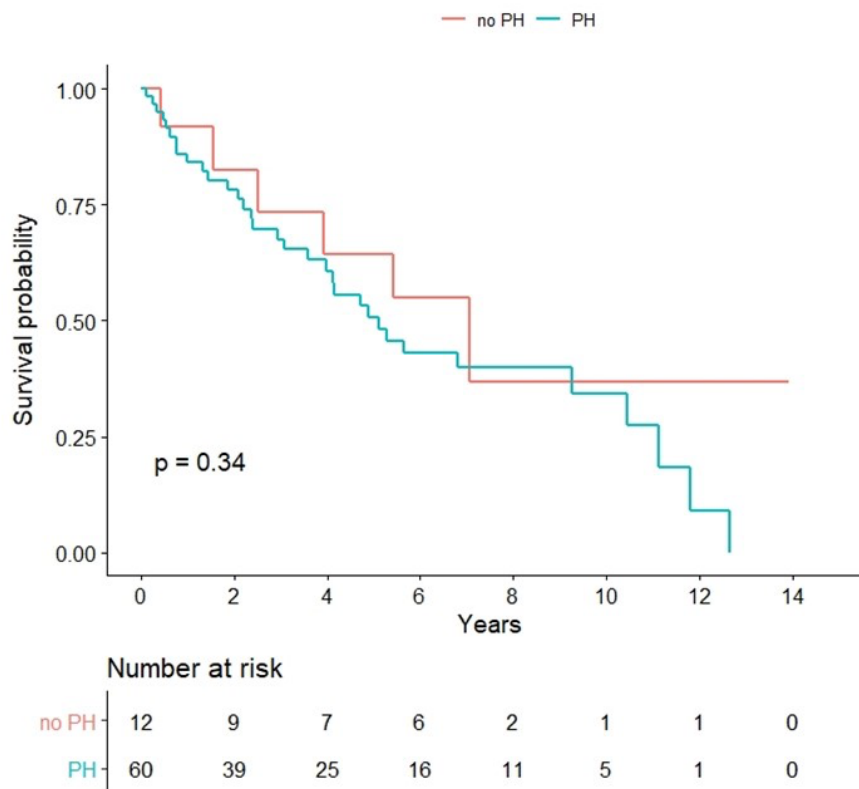
The comparisons were performed using log-rank test. There was a statistically significant difference between the two groups: median conditional survival 9.3 years in female transplant recipients vs. 4.5 years in male transplant patients ($p=0.003$).

Figure S3: Kaplan-Meier curve demonstrating the probability of survival following primary lung transplantation in patents with PLCH after segregating the cohort on the basis of type of lung transplant: single LT versus bilateral sequential LT.



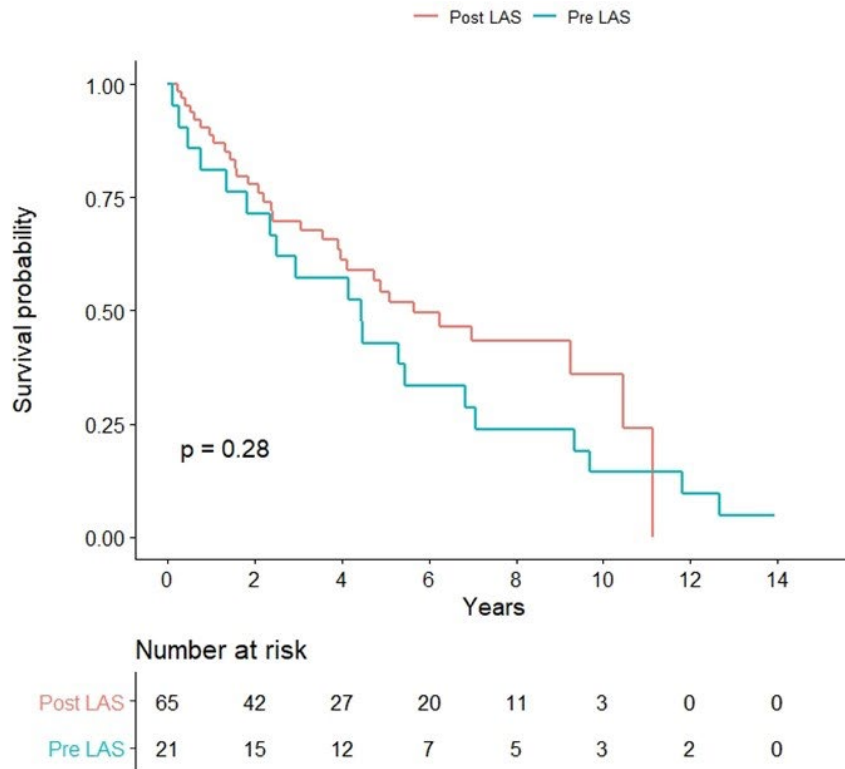
The comparisons were performed using the log-rank test. There was no statistically significant difference found between the two groups: median survival 4.9 years in patients with single lung transplant as compared to 5.4 years in patients with bilateral sequential lung transplant ($p=0.69$).

Figure S4: Kaplan-Meier curve demonstrating the probability of survival following primary lung transplantation in patents with PLCH after segregating the cohort on the basis of presence or absence of pulmonary hypertension, defined by mean pulmonary artery pressure ≥ 25 mmHg determined by right heart catheterization.



The comparisons were performed using the log-rank test. There was no statistically significant difference found between the two groups: median survival of 5.1 years in patients with pulmonary hypertension compared with median survival of 7.1 years in patients without pulmonary hypertension (p=0.34).

Figure S5: Kaplan-Meier curve demonstrating the probability of survival following primary lung transplantation in patents with PLCH after segregating the cohort on the basis of time of lung transplant: before institution of the lung allocation score (LAS) system versus post-LAS.



The comparisons were performed using the log-rank test. There was no statistically significant difference found between the two groups: median survival before institution of the LAS system was 4.5 years as compared with post-LAS median survival of 5.6 years ($p=0.28$).

Table S1: Details regarding the development of bronchiolitis obliterans post lung transplantation in our cohort.

Variable	N (%)
No BOS	33 (42%)
BOS at any time	45 (58%)
BOS Grade at diagnosis (n=45)	
Grade 0p	11 (24%)
Grade 1	4 (9%)
Grade 2	3 (7%)
Grade 3	8 (18%)
Grade unknown	19 (42%)

Data regarding BOS was available in 78 patients.

Abbreviations: BOS = Bronchiolitis obliterans syndrome

Table S2: Details regarding the timing and cause of death post lung transplantation in our cohort.

Pt. No.	Date of Transplant	Gender	Type of transplant	Survival	Cause of Death
1	2/9/1996	Male	BSLT	39 days	Pulmonary dehiscence
2	4/7/2006	Female	BSLT	88 days	Infection: Pseudomonal Sepsis
3	11/21/2001	Male	SLT	92 days	Respiratory failure
4	10/30/2005	Male	BSLT	120 days	Infection: Pseudomonas Bacteremia
5	09/21/07	Male	BSLT	151 days	Bacterial septicemia
6	11/12/1998	Male	SLT	5.6 months	Respiratory failure
7	1/19/2014	Female	BSLT	6.4 months	Respiratory failure
8	1/26/2014	Male	BSLT	7.4 months	Unknown
9	2/25/1997	Female	SLT	9.3 months	Graft failure: non-specific
10	4/18/2012	Male	BSLT	9.4 months	CMV Infection
11	5/9/2008	Female	BSLT	11.9 months	Graft failure: graft infection
12	10/18/2007	Male	BSLT	1.1 years	Respiratory failure
14	8/21/1993	Male	BSLT	1.2 years	BOS
15	9/20/2008	Male	BSLT	1.3 years	Unknown
16	6/26/2009	Female	BSLT	1.4 years	BOS
16	12/13/2005	Female	SLT	1.6 years	Metastatic Liver Cancer
17	06/24/07	Male	BSLT	1.6 years	Cardiovascular: Other
18	1/24/2005	Female	BSLT	1.8 years	Cardiovascular: cardiac arrest
19	7/16/2010	Male	BSLT	1.9 years	BOS
20	12/9/2007	Male	BSLT	2.1 years	Cerebrovascular: Stroke
21	6/5/2005	Male	BSLT	2.2 years	Infection: bacterial septicemia
22	7/13/2011	Male	BSLT	2.4 years	BOS
23	06/18/10	Male	BSLT	2.4 years	Multiple organ failure
24	1/14/2003	Female	SLT	2.4 years	BOS
25	3/7/2005	Female	BSLT	2.5 years	Graft failure: graft infection
26	12/19/2002	Male	BSLT	2.9 years	BOS
27	5/19/2013	Female	BSLT	3.1 years	BOS
28	10/2/2008	Female	BSLT	3.6 years	Acute respiratory distress disease
29	4/10/2007	Male	BSLT	3.9 years	Respiratory failure
30	1/26/2013	Male	BSLT	4 years	Unknown
31	1/22/2013	Male	BSLT	4.1 years	BOS
32	3/14/1999	Female	BSLT	4.2 years	BOS
33	9/13/2000	Female	SLT	4.4 years	Respiratory failure
34	5/31/1998	Male	SLT	4.5 years	BOS
35	1/10/2006	Female	BSLT	4.7 years	Suicide
36	09/10/12	Male	BSLT	4.9 years	BOS
37	10/26/2011	Male	BSLT	5.1 years	Acute rejection
38	4/6/2002	Female	SLT	5.3 years	Unknown
39	10/6/2002	Female	SLT	5.4 years	BOS
40	7/1/2008	Male	BSLT	5.6 years	BOS
41	3/12/2008	Female	BSLT	6.2 years	Acute respiratory distress disease

42	6/15/1998	Female	SLT	6.8 years	Infection: Disseminated Aspergillus
43	8/9/2006	Male	BSLT	7 years	Unknown
44	4/18/2001	Male	BSLT	7.1 years	PTLD
45	11/14/07	Male	BSLT	9.3 years	Missing
46	10/13/1998	Female	SLT	9.3 years	Respiratory failure
47	11/10/1997	Male	BSLT	9.7 years	Respiratory failure
48	1/17/2006	Male	BSLT	10.5 years	Unknown
49	8/18/2005	Female	SLT	11.1 years	BOS
50	11/21/2002	Female	BSLT	11.8 years	Acute rejection
51	8/2/2001	Female	SLT	12.7 years	Respiratory hemorrhage

There were a total of 51 deaths in our cohort. Primary cause of death was reported to the OPTN/UNOS database by the individual institutions. For patients 16 and 39 there was no primary cause of death listed, so secondary cause of death was used instead.

Abbreviations: CMV = cytomegalovirus, BOS= bronchiolitis obliterans syndrome, BSLT = bilateral sequential lung transplantation, MI = myocardial infarction, PTLD = post-transplant lymphoproliferative disorder, SLT = single lung transplantation.