**SDC Table 5.** Study cohort characteristics pre- and post-LT. Data are shown for all participants (“all PFIC”), and for FIC1, BSEP, and MDR3 disease subgroups. Median age at LT for only 18 of the 19 BSEP post-LT participants is noted because transplant age was unavailable for one BSEP participant.

| ***n (%) or******Median (Q1, Q3)*** | ***FIC1******(n=26)*** | ***BSEP******(n=53)*** | ***MDR3******(n=19)*** | ***All PFIC******(n=98)*** | ***p-value\**** |
| --- | --- | --- | --- | --- | --- |
| **Sex** |  |  |  |  | 0.48 |
|  Female | 17 (65%) | 31 (58%) | 9 (47%) | 57 (58%) |  |
|  Male | 9 (35%) | 22 (42%) | 10 (53%) | 41 (42%) |  |
| **Race** |  |  |  |  | <0.001 |
|  White | 19 (73%) | 40 (75.5%) | 11 (58%) | 70 (71%) |  |
|  Black or African-American |  |  | 5 (26%) | 5 (5%) |  |
|  Asian | 1 (4%) | 2 (4%) | 2 (11%) | 5 (5%) |  |
|  Other\*\* | 6 (23%) | 11 (21%) | 1 (5%) | 18 (18%) |  |
| **Ethnicity** |  |  |  |  | 0.07 |
|  Hispanic, Latino, or Spanish origin | 2 (8%) | 10 (19%) | 7 (37%) | 19 (19%) |  |
|  Other origin | 23 (88%) | 43 (81%) | 11 (58%) | 77 (79%) |  |
|  Unknown or not reported | 1 (4%) |  | 1 (5%) | 2 (2%) |  |
| **Age (months) at symptom onset** | n=26 | n=49 | n=15 | n=90 |  |
|  Median (Q1, Q3) | 2 (0, 4) | 4 (1, 7) | 15 (0, 63) | 4 (1, 8) | 0.01 |
| **Age (months) at diagnosis** | n=25 | n=49 | n=18 | n=92 |  |
|  Median (Q1, Q3) | 4 (3, 10) | 9 (6, 17) | 57 (32, 131) | 10 (4, 30) | <0.001 |
| **Age (years) at enrollment** |  |  |  |  |  |
|  Enrolled pre-LT | n=24 | n=34 | n=18 | n=76 |  |
|  Median (Q1, Q3) | 4 (1, 11) | 2 (1, 6) | 10 (7, 12) | 4 (1, 11) | 0.002 |
|  Enrolled post-LT | n=2 | n=19 | n=1 | n=22 |  |
|  List of ages or median (Q1, Q3) | 18, 20 | 10 (7, 11) | 13 | 10 (7, 13) | 0.11 |
|  All participants | n=26 | n=53 | n=19 | n=98 |  |
|  Median (Q1, Q3) | 4 (1, 13) | 5 (1, 10) | 11 (7, 12) | 7 (2, 11) | 0.03 |
| **Age (years) at LT (enrolled post-LT)** | n=2 | n=18 | n=1 | n=21 |  |
|  Age list or median (Q1, Q3) | 6, 13 | 3 (2, 5) | 4 | 3 (2, 6) | 0.004 |
| **Time (years) from symptom onset to enrollment (enrolled pre-LT)** | n=24 | n=33 | n=14 | n=71 |  |
|  Median (Q1, Q3) | 3 (1, 11) | 1 (1, 4) | 7 (1, 10) | 3 (1, 9) | 0.82 |

\*p-values for categorical variables were from chi-square tests; p-values for continuous variables were from Kruskal-Wallis tests.

\*\*Other race includes American Indian, Native Hawaiian, Multi-Racial, Other, Don’t Know, and Refused.

Abbreviations: BSEP, bile salt export pump; FIC1; familial intrahepatic cholestasis; LT, liver transplant; MDR3, multidrug resistance; n, number; PFIC, progressive familial intrahepatic cholestasis; Q1, quartile 1; Q3, quartile 3.