**Supplemental Table S3. Comparison of PDD and AGD subgroups of maltase deficiencies (Maltase < 100 from Total 30 K Series) by ANOVA (p) and Tukey test (values with same letter are not different).**

|  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Activity | PSID | N | Mean | StDev | Minimum | Maximum | ANOVA | p |
| Lactase | AGD | 834 | 16.514 | 6.901 | 10.000 | 68.600 | C | 0.000 |
|  | AGSID | 38 | 16.45 | 7.49 | 10.40 | 42.00 | C |  |
|  | CSID | 50 | 42.01 | 19.67 | 13.10 | 94.70 | A |  |
|  | PDD | 2477 | 3.9862 | 2.8644 | 0.1000 | 10.0000 | D |  |
|  | PMD | 52 | 22.25 | 18.43 | 0.10 | 62.70 | B |  |
|  | PSID | 375 | 2.593 | 2.468 | 0.100 | 9.800 | E |  |
| Sucrase | AGD | 834 | 24.039 | 6.038 | 0.000 | 46.900 | B | 0.000 |
|  | AGSID | 38 | 11.808 | 4.451 | 2.300 | 18.900 | D |  |
|  | CSID | 50 | 7.35 | 8.24 | 0.00 | 22.50 | DE |  |
|  | PDD | 2477 | 18.764 | 7.734 | 0.000 | 47.100 | C |  |
|  | PMD | 52 | 53.96 | 22.89 | 10.30 | 107.80 | A |  |
|  | PSID | 375 | 7.583 | 4.348 | 1.000 | 18.400 | E |  |
| Maltase | AGD | 834 | 80.381 | 14.633 | 2.600 | 99.700 | A | 0.000 |
|  | AGSID | 38 | 76.28 | 17.80 | 31.10 | 99.10 | A |  |
|  | CSID | 50 | 51.60 | 24.41 | 17.10 | 93.30 | C |  |
|  | PDD | 2477 | 65.048 | 23.682 | 0.000 | 99.800 | B |  |
|  | PMD | 52 | 37.63 | 32.55 | 3.00 | 92.90 | D |  |
|  | PSID | 375 | 46.10 | 22.54 | 5.50 | 98.60 | CD |  |
| Palatinase | AGD | 789 | 5.634 | 3.050 | 0.000 | 44.700 | B | 0.000 |
|  | AGSID | 36 | 3.347 | 1.309 | 0.000 | 5.600 | CD |  |
|  | CSID | 47 | 2.121 | 2.368 | 0.000 | 10.000 | D |  |
|  | PDD | 2318 | 4.0607 | 2.9046 | 0.0000 | 34.3000 | C |  |
|  | PMD | 49 | 9.402 | 5.441 | 2.800 | 25.200 | A |  |
|  | PSID | 352 | 1.9321 | 1.7046 | 0.0000 | 17.0000 | D |  |
| M/S | AGD | 831 | 3.4317 | 0.5638 | 0.7313 | 5.0000 | C | 0.000 |
|  | AGSID | 38 | 7.98 | 6.38 | 5.11 | 39.70 | A |  |
|  | CSID | 29 | 7.95 | 7.54 | 2.97 | 37.77 | A |  |
|  | PDD | 2449 | 3.5637 | 0.6302 | 1.2449 | 4.9915 | C |  |
|  | PMD | 52 | 0.7655 | 0.6852 | 0.0734 | 3.3301 | D |  |
|  | PSID | 375 | 6.714 | 2.651 | 5.000 | 36.826 | B |  |
| M-2.24S | AGD | 834 | 26.532 | 10.568 | -70.756 | 53.300 | D | 0.000 |
|  | ASSID | 38 | 49.83 | 14.51 | 20.12 | 86.15 | A |  |
|  | CSID | 50 | 35.14 | 13.88 | 13.26 | 78.54 | B |  |
|  | PDD | 2477 | 23.018 | 11.178 | 13.648 | 52.972 | E |  |
|  | PMD | 52 | -83.24 | 55.50 | -215.27 | 11.23 | E |  |
|  | PSID | 375 | 29.111 | 13.815 | 3.260 | 79.548 | C |  |
| L/S | AGD | 831 | 0.7392 | 1.1339 | 0.2701 | 32.3000 | C | 0.000 |
|  | AGSID | 38 | 2.032 | 2.823 | 0.603 | 17.174 | B |  |
|  | CSID | 29 | 6.84 | 9.87 | 1.27 | 39.46 | A |  |
|  | PDD | 2449 | 0.23978 | 0.20840 | 0.00228 | 3.03846 | E |  |
|  | PMD | 52 | 0.4006 | 0.2617 | 0.0009 | 1.1552 | CDE |  |
|  | PSID | 375 | 0.4368 | 0.6010 | 0.0056 | 7.9167 | D |  |
| Age | AGD | 828 | 10.065 | 7.324 | 0.003 | 79.280 | B | 0.000 |
|  | AGSID | 38 | 8.763 | 5.707 | 0.400 | 19.201 | ABC |  |
|  | CSID | 50 | 6.512 | 5.185 | 0.334 | 18.727 | C |  |
|  | PDD | 2470 | 11.440 | 6.543 | 0.005 | 71.439 | A |  |
|  | PMD | 52 | 9.825 | 5.044 | 0.235 | 21.481 | ABC |  |
|  | PSID | 374 | 10.503 | 7.001 | 0.003 | 90.136 | AB |  |

Supplementary Digital Content

**Brief Clinical Examples:**

*PMD1*

*Seven month old with chronic starch induced diarrhea*

*Normal duodenal histology*

*Lactase: 44. Sucrase, 41, Maltase: 87, Palatinase: 6*

*Reduced* 13*C-starch BT*

*Note: Onset of intolerance when complimentary starches fed.*

*PMD 2*

*Seven yr. with multiple allergies and chronic abdominal pain*

*Eosinophilia on duodenal biopsy*

*Lactase: 20, Sucrase: 75, Maltase: 59, Palatinase: 17*

*Note: Chronic abdominal pain with PMD*

*PDD 1:*

*4 mo with severe failure to thrive (20)*

*Chronic diarrhea from birth on lactose*

*Gaining on Total Parenteral Nutrition*

*Not gaining on adequate oral maltodextrin*

*Normal duodenal histology*

*Lactase: 3, Sucrase: 9. Maltase 38*

*Reduced* 13*C-starch BT*

*Note: Gained with 2% supplementary oral glucose*

*PDD 2*

*Adult-type lactase deficiency in teenage CSID*

*Normal duodenal histology*

*Elevated lactase at 11 mo. (Lactase: 46. Sucrase 0, Maltase: 19, Palatinase: 0)*

*Deficient lactase at 15 yr. (Lactase: 0. Sucrase: 0. Maltase: 21, Palatinase: 0)*

*Reduced* 13*C-starch BT at 15 yr*

*C/T – 13910 polymorphism present*

*Note: Non-persistent lactase in teenage with CSID*