**Table, SDC 2: Biochemical parameters and copper metabolic parameters of group 1 (< 20mg/kg TD) and group 2 (> 20mg/kg TD) at initation, after 24 and 60 months of trientine therapy**

|  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Parameter** | **baseline** | | | | |  | **24 months** | | | | |  | **60 months** | | | | |
|  | **Group 1** | | **Group 2** | |  |  | **Group 1** | | **Group 2** | |  |  | **Group 1** | | **Group 2** | |  |
|  | n | median (range) | n | median (range) | **p value** |  | n | median (range) | n | median (range) | **p value** |  | n | median (range) | n | median (range) | **p value** |
| **AST [U/I]** | 13 | 27 (8-123) | 10 | 40 (21-168) | 0.147 |  | 10 | 23 (17-55) | 10 | 36 (23-68) | **0.027** |  | 6 | 29 (19-45) | 8 | 32 (16-69) | 0.419 |
| **ALT [U/l]** | 13 | 24 (8-310) | 10 | 56 (12-371) | 0.181 |  | 10 | 28 (10-92) | 10 | 48 (27-151) | 0.052 |  | 7 | 34 (18-137) | 8 | 62 (26-203) | 0.356 |
| **GGT [U/l]** | 13 | 20 (6-122) | 10 | 35 (9-93) | 0.744 |  | 10 | 17 (8-36) | 9 | 24 (12-48) | 0.229 |  | 7 | 34 (11-63) | 8 | 23 (13-87) | 0.811 |
| **Bilirubin [U/l]** | 12 | 0.60  (0.13-10.60) | 9 | 0.40  (0.2-1.4) | 0.375 |  | 10 | 0.45  (0.20-0.90) | 9 | 0.6  (0.2-1.0) | 0.550 |  | 6 | 0.35  (0.20-0.50) | 8 | 0.50  (0.4-1.3) | 0.079 |
| **Creatinine [mg/dl]** | 12 | 0.62  (0.33-0.98) | 9 | 0.48  (0.35-0.99) | 0.323 |  | 10 | 0.63  (0.13-1.00) | 9 | 0.54  (0.29-0.79) | 0.166 |  | 7 | 0.56  (0.37-1.04) | 8 | 0.63  (0.35-0.84) | 0.805 |
| **Albumin [g/l]** | 12 | 43  (27-49) | 9 | 43.0  (38.8-46.0) | 0.477 |  | 9 | 43  (40.1-50.6) | 9 | 45.6  (35-51.9) | 0.498 |  | 5 | 44.9  (41.0-46.2) | 8 | 46.5  (42.2-52.9) | 0.072 |
| **INR** | 10 | 1.01  (0.9-2.0) | 9 | 1.01  (1.00-1.17) | 0.628 |  | 9 | 1.00  (0.95-1.09) | 9 | 1.06  (0.99-1.10) | 0.200 |  | 5 | 1.05  (0.95-1.07) | 7 | 1.00  (0.98-1.11) | 0.878 |
| **WBC [109/L]** | 13 | 6.60  (3.25-16.4) | 9 | 6.01  (4.13-9.86) | 0.370 |  | 10 | 5.87  (3.92-10.90) | 9 | 5.88  (4.25-9.60) | 0.982 |  | 8 | 5.63  (4.1-10.4) | 8 | 6.29  (4.00-9.91) | 0.818 |
| **Hb [g/dl]** | 13 | 12.9  (6.2-15.7) | 9 | 13.0  (12.2-14.3) | 0.635 |  | 10 | 12.1  (11.4-15.7) | 9 | 12.9  (11.8-14.1) | 0.943 |  | 8 | 12.7  (11.4-15.4) | 8 | 13.2  (12.0-15.4) | 0.295 |
| **Platelets [109/L]** | 13 | 257  (104-514) | 9 | 285  (173-465) | 0.499 |  | 10 | 255  (146-385) | 10 | 264  (156-382) | 0.855 |  | 8 | 274  (163-417) | 8 | 250  (204-425) | 0.998 |
| **Cerulplasmin [g/l]** | 9 | 0.09  (0.03-0.14) | 9 | 0.11  (0.01-0.20) | 0.680 |  | 8 | 0.09  (0.02-0.23) | 9 | 0.10  (0.03-0.15) | 0.919 |  | 6 | 0.08  (0.04-0.16) | 7 | 0.08  (0.02-0.17) | 0.545 |
| **Serum copper [µmol/l]** | 8 | 5.3  (1.9-10.0) | 9 | 7.8  (4.0-9.5) | 0.394 |  | 7 | 5.0  (0.8-11.9) | 9 | 5.0  (2.7-9.4) | 0.659 |  | 6 | 4.2  (1.8-9.8) | 7 | 4.0  (1.5-5.3) | 0.359 |
| **NCC [µmol/l]** | 8 | 1.26  (0.01-6.22) | 9 | 1.95  (0.01-6.95) | 0.603 |  | 7 | 0.66  (0.01-2.17) | 9 | 0.74  (0.01-3.08) | 0.496 |  | 5 | 1.21  (0.01-4.57) | 7 | 0.32  (0.01-3.63) | 0.350 |
| **Urinary copper excretion [µmol/day]** | 11 | 4.21  (0.36-187.2) | 10 | 2.36  (0.71-12.58) | 0.335 |  | 9 | 1.94  (0.86-4.62) | 10 | 1.30  (0.9-6.030) | 0.344 |  | 7 | 1.75  (0.80-3.99) | 6 | 2.70  (1.42-9.60) | 0.166 |

ALT = alanine aminotransferase; AST = aspartate aminotransferase; GGT = gamma-glutamyl transferase; Hb = hemoglobin; INR = international normalized ratio; n = number; NCC = non-ceruloplasmin bound copper; TD = trientine-dihydrochloride; WBC = white blood cells;