**Supplementary Table 1. Comparative data for patients with WD-ALF and ALF from others**

|  |  |  |  |
| --- | --- | --- | --- |
| **Parameters** | **WD-ALF** | **Indeterminate ALF** | **ALF from others** |
| **(N=15)** | **(N=9)** | **(N=61)** |
| Age (year)  Female (%)  Zn (μmol/L)  Corrected Zn (μmol/L)  Copper (μmol/L)  Free copper (μmol/L)  ALP (IU/L)  Low ALP for age (N)  Albumin (g/L)  TB (μmol/L)  AST (IU/L)  ALT (IU/L)  AST to ALT  AST to ALT > 2 (N)  AST to ALT > 4 (N) | 13.2 (7.7-16.0)  5 (33.3%)  5.8 (4.1-8.3) (N=8)  7.3 (5.2-11.6)  14.5 (4.5-36.3)  7.92 (3.08-31.58) (N=13)  92 (19-1120)  10  22 (19-35)  57 (19-625)  132 (39-403)  54 (11-190)  2.83 (0.68-12.36)  10  4 | 7.6 (1.0-12.2)\*  1 (11%)  9.8 (7.0-12.1)†  11.4 (7.6-17.8)\*  18.1 (11.4-36.7)  6.41 (3.38-23.48)  345 (159-758)\*  0\*  36 (25-41)\*  282 (48-448)\*  2778 (2039-6637)†  2744 (1816-2857)\*  1.05 (0.74-1.53)†  0\*  0\* | 6 (1.1-16.6)\*  24 (39%)  -  -  -  -  255 (102-1286)\*  2†  34 (19-41)\*  150 (6-844)  1421 (34-11303)†  974 (8-7833)†  1.37 (0.18-7.30)\*  19\*  7 |

Value = Median (range)

\* P < 0.05 vs WD-ALF, †P < 0.001 vs WD-ALF

**Supplementary Table 2. The Diagnostic Value of 3 Parameters in the diagnostic score published by**

**Ferenci *et al.***

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| **Parameter** | **Cut-off** | **sensitivity** | **specificity** | **PPV** | **NPV** | **Accuracy** | **AUC (95%CI)** |
| Liver copper (μg/g dry weight)  Basal urine copper (μg/day)  Urine copper post PCT (μg/day)  Ceruloplasmin (g/L) | >50  >120  >250  >0.5  >0.6  >1.6  >2.0  >3.0  >5  >8  >15  >25  <0.1  <0.15  <0.2  <0.25 | μ  93.9%  86.8%  76.3%  100%  95.5%  81.8%  77.3%  59.1%  83.8%  81.1%  56.8%  37.8%  53.5%  74.4%  90.7%  97.7% | 71.5%  87.4%  97.7%  49.3%  55.6%  91.8%  95.2%  96.6%  19.0%  40.5%  83.6%  99.5%  100%  96.4%  79.5%  37.4% | 47.0%  75.0%  93.5%  17.3%  18.6%  51.4%  63.0%  65.0%  16.4%  20.5%  39.6%  93.3%  100%  76.2%  40.6%  19.4% | 97.8%  93.8%  90.4%  100%  99.1%  97.9%  97.5%  95.7%  86.0%  91.9%  91.1%  89.4%  93.3%  93.5%  81.5%  45.5% | 76.3%  87.2%  91.2%  54.1%  59.4%  90.8%  93.4%  93.0%  29.3%  47.0%  79.3%  89.7%  93.8%  93.5%  81.5%  45.5% | 0.931 (0.869-0.992)  0.939 (0.888-0.990)  0.726 (0.613-0.838)  0.935 (0.884-0.986) |

**Supplemantary table 3. Data of false-positive and false-negative results among the study population**

|  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Case** | **Sex** | **Age at**  **diagnosis**  **(years)** | **Diagnosis** | **Liver Cu**  **(μg/g dry**  **liver)** | **Basal urine**  **Cu (μmol/**  **day)** | **Urine Cu**  **post PCT**  **(μmol/day)** | **CP**  **(g/L)** | **KF**  **rings** | **Neuro**  **Symp** | **Zn**  **(μmol/L)** | **ALP**  **(IU/L)** | **AST to**  **ALT**  **≥ 2** | **Diagnostic**  **score by**  **Ferenci *et al***  **(marks)** | **Proposed WD**  **diagnostic**  **Score (marks)** |
| **Patients with false negative results** | | | | | | | | | | | | | | |
| 1  2  3  4  5  6  7  8  9 | F  F  F  M  F  M  F  M  F | 1.64  4.50  9.50  11.23  11.96  13.04  13.43  14.57  15.89 | WD-non-ALF  WD-non-ALF  WD-ALF  WD-ALF  WD-non-ALF  WD-ALF  WD-non-ALF  WD-ALF  WD-ALF | 513  38  -  -  7  -  875  -  - | -  0.7  16.7  -  -  5.8  0.5  30.9  - | -  4.6  98.7  -  0.9  19.2  2.4  67.7  68.6 | 0.11  0.17  0.13  0.20  0.05  0.10  0.11  0.17  0.16 | N  N  N  N  N  N  N  N  N | N  N  N  N  N  N  N  N  N | -  -  4.9  -  11.9  4.1  -  -  6.5 | 357  28  22  31  74  189  522  168  19 | N  N  Y  Y  N  N  N  Y  Y | 3  2  3  1  1  3  3  3  3 | 3  3  5  2  3  3  3  4  5 |
| **Patients with false positive results** | | | | | | | | | | | | | | |
| 1  2  3  4  5  6  7  8  9  10  11 | F  F  F  F  M  M  M  F  F  M  M | 8.69  8.87  13.21  13.73  13.77  13.80  14.79  15.41  15.45  15.73  16.76 | AIH  AIH  AIH  AIH  AIH type I  AIH  AIH  AIH type I  AIH  AIH type I  AIH | 247  401  97  77  -  53  107  66  52  196  108 | 0.6  0.8  2.3  0.7  0.5  1.4  1.6  1.5  0.1  0.2  1.6 | 11.25  12.4  21.6  9  15.3  17.6  22.1  16.6  11.75  8.6  9 | 0.19  0.29  0.16  0.12  0.17  0.15  0.17  0.15  0.15  0.16  0.20 | N  N  N  N  N  N  N  N  N  N  N | N  N  N  N  N  N  N  N  N  N  N | -  -  -  -  -  12.9  -  8.4  -  9.5  - | 432  277  1164  381  912  341  355  220  235  970  242 | N  Y  N  N  N  N  N  N  N  N  N | 4  4  4  4  4\*  4  4  4  4  4  4 | 3  3  4  3  3\*  3  3  3  3  3  3 |

\* One mark from positive Rhodanine staining

\*\* All has negative results from the Coomb’s negative hemolysis