**SUPPLEMENTARY TABLES**

**Supplementary Table 1.** The discriminative ability for all risk scores for liver transplantation or death between initiation of UDCA therapy up to 5 years thereafter

|  |  |  |  |
| --- | --- | --- | --- |
| **Model** | **N** | **C-statistic** | **95% CI** |
| ***First calculable score in first 5 years following initiation of UDCA*** | | | |
| MRS 1989 | 1171 | 0.789 | 0.752 - 0.825 |
| MRS 1994 | 1171 | 0.788 | 0.752 - 0.825 |
| MELD score | 1171 | 0.699 | 0.670 - 0.728 |
| UK-PBC scorea | 1171 | 0.756 | 0.713 - 0.800 |
| GLOBE scorea | 1171 | 0.794 | 0.759 - 0.830 |
| ***At start of UDCA therapy*** | | |  |
| MRS 1989 | 1100 | 0.768 | 0.728 - 0.809 |
| MRS 1994 | 1100 | 0.768 | 0.728 - 0.808 |
| MELD score | 1100 | 0.681 | 0.652 - 0.710 |
| GLOBE score | 1100 | 0.779 | 0.739 - 0.818 |
| ***At 12 months UDCA*** | |  |  |
| MRS 1989 | 905 | 0.764 | 0.716 - 0.813 |
| MRS 1994 | 905 | 0.765 | 0.717 - 0.812 |
| MELD score | 905 | 0.679 | 0.638 - 0.720 |
| UK-PBC scoreb | 905 | 0.738 | 0.671 - 0.805 |
| GLOBE scoreb | 905 | 0.799 | 0.756 - 0.842 |
| ***At 24 months UDCA*** | |  |  |
| MRS 1989 | 548 | 0.784 | 0.725 - 0.843 |
| MRS 1994 | 548 | 0.786 | 0.727 - 0.844 |
| MELD score | 548 | 0.691 | 0.641 - 0.741 |
| UK-PBC scorec | 548 | 0.741 | 0.691 - 0.791 |
| GLOBE scorec | 548 | 0.817 | 0.765 - 0.868 |
| ***At 36 months UDCA*** |  |  |  |
| MRS 1989 | 648 | 0.819 | 0.769 - 0.868 |
| MRS 1994 | 648 | 0.824 | 0.776 - 0.872 |
| MELD score | 648 | 0.757 | 0.711 - 0.802 |
| UK-PBC scored | 648 | 0.780 | 0.718 - 0.842 |
| GLOBE scored | 648 | 0.831 | 0.779 - 0.882 |
| ***At 48 months UDCA*** |  |  |  |
| MRS 1989 | 610 | 0.816 | 0.763 - 0.869 |
| MRS 1994 | 610 | 0.821 | 0.769 - 0.873 |
| MELD score | 610 | 0.746 | 0.698 - 0.793 |
| UK-PBC scoree | 610 | 0.804 | 0.749 - 0.859 |
| GLOBE scoree | 610 | 0.844 | 0.797 - 0.891 |
| ***At 60 months UDCA*** |  |  |  |
| MRS 1989 | 549 | 0.801 | 0.744 - 0.8590 |
| MRS 1994 | 549 | 0.806 | 0.749 - 0.8633 |
| MELD score | 549 | 0.704 | 0.657 - 0.7522 |
| UK-PBC scoref | 549 | 0.778 | 0.716 - 0.8400 |
| GLOBE scoref | 549 | 0.842 | 0.791 - 0.8931 |

Abbreviation: UDCA, ursodeoxycholic acid; CI, confidence interval.

ac-statistic for outcomes liver-related death or LT: 0.822 (0.785 - 0.859) for UK-PBC and 0.820 (0.784 - 0.857) for GLOBE score.

bc-statistic for outcomes liver-related death or LT: 0.809 (0.764 - 0.854) for UK-PBC and 0.814 (0.765 - 0.862) for GLOBE score.

c c-statistic for outcomes liver-related death or LT: 0.804 (0.737 - 0.870) for UK-PBC and 0.834 (0.775 - 0.893) for GLOBE score.

d c-statistic for outcomes liver-related death or LT: 0. 850 (0.798 - 0.903) for UK-PBC and 0.845 (0.790 - 0.901) for GLOBE score.

e c-statistic for outcomes liver-related death or LT: 0.870 (0.824 - 0.917) for UK-PBC and 0.860 (0.806 - 0.914) for GLOBE score.

f c-statistic for outcomes liver-related death or LT: 0.823 (0.764 - 0.882) for UK-PBC and 0.858 (0.805 - 0.912) for GLOBE score.

**Supplementary Table 2**. Difference between observed and median predicted transplant-free survival after 1 year of UDCA across different risk scores and time points

|  |  |  |  |
| --- | --- | --- | --- |
| **Predicted - observed survival (%)** | | | |
| **Time after 1 year of UDCA** | **Delta GLOBE** | **Delta MRS 1989** | **Delta MRS 1994** |
| 1 |  | +0.5 | +1.3 |
| 2 |  | +1.3 | +2.9 |
| 3 | +1.0 | +0.3 |  |
| 4 |  | +0.7 |  |
| 5 | +2.5 | +0.4 |  |
| 6 |  | +0.1 |  |
| 7 |  | -0.7 |  |
| 8 |  |  |  |
| 9 |  |  |  |
| 10 | +3.5 |  |  |
| 11 |  |  |  |
| 12 |  |  |  |
| 13 |  |  |  |
| 14 |  |  |  |
| 15 | +2.1 |  |  |

Abbreviation: UDCA, ursodeoxycholic acid; MRS, Mayo risk score.

**Supplementary Table 3.** Proportion of patients with any missing data in the first 5 years after start of UDCA therapy

|  |  |
| --- | --- |
|  | **Missing data**  **N, %** |
| Serum total bilirubin | 36 (3.3) |
| Serum ALP | 20 (1.8) |
| Serum AST | 18 (1.6) |
| Serum ALT | 20 (1.8) |
| Serum albumin | 103 (9.4) |
| Serum platelets | 34 (3.1) |
| Serum creatinine | 349 (31.7) |
| PTa | 203 (18.5) |
| INRa | 101 (9.2) |

Abbreviations: ALP, alkaline phosphatase; ALT, alanine aminotransferase; AST, aspartate aminotransferase; PT, prothrombin time; INR, international normalized ratio.

a In the cases were PT was missing, we assumed normal PT and INR values when albumin and bilirubin were within the normal range. Subsequently, the missing PT and INR values were imputed by multiple imputation.

**Supplementary Table 4.** Sensitivity analysis of the discriminative performance of the various risk prediction scores calculated after 1 year of UDCA therapy in the 25% of cases not used in the derivation of the GLOBE score.

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
|  |  | | | **C statistic (95% CI)** |
| ***At 1 year of UDCA therapy - 17 events*** | | | | |
| MRS 1989 | | 224 | 0.92 (0.88-0.96) | |
| MRS 1994 | | 224 | 0.92 (0.87-0.97) | |
| MELD score | | 224 | 0.81 (0.73-0.88) | |
| UK-PBC score | | 224 | 0.87 (0.80-0.93) | |
| GLOBE score | | 224 | 0.92 (0.88-0.97) | |