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Title: Hepatopulmonary syndrome: Long-term survival in the Mayo Clinic experience

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Body: Introduction:Hepatopulmonary syndrome (HPS) is an uncommon pulmonary vascular disorder occurring in advanced liver disease, characterized by hypoxemia due to intrapulmonary vascular dilatations. Liver transplantation (LT) improves survival in HPS. We present the largest consecutive series of HPS patients specifically addressing long-term survival relative to the degree of pre-LT hypoxemia. Methods: Survival was assessed using Kaplan-Meier methodology for 106 HPS patients from 1986 through 2010. Results: 49 HPS patients underwent LT. Post-LT survival (1, 3, 5 and 10 year) did not differ between groups based on Pa0₂ at the time of HPS diagnosis. Improvements in overall survivals at 1, 3 and 5 years post-LT in those HPS patients transplanted after 2002 (MELD exception era, n=28) (92, 87 and 87%, respectively) as compared to those transplanted prior to that time (pre MELD era, n=21) (71, 67 and 67%, respectively) did not reach statistical significance (P= 0.09) (figure 1). Model for Endstage Liver Disease (MELD) exception to facilitate LT was granted to 18 patients since 2002 with post-LT survival of 15/18 patients (83%) and no wait-list mortality. Conclusion: Long-term outcome after LT in HPS is favorable. The survival patterns from the time of LT were not influenced by pre-LT PaO₂. Limited experience with HPS-MELD exception suggests a positive impact on survival with no wait list mortality.