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Comparison of Long-Term Outcome of Patients with Wilson's Disease Presenting with Acute Liver Failure versus Acute-on-Chronic Liver Failure

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Abstract

Objective: Wilson's disease (WD) is an inherited disorder of copper metabolism, leading to hepatic and neuropsychiatric manifestations. The long-term outcome of patients with Wilson' disease presenting with acute liver failure (ALF) versus those with acute-on-chronic liver failure (ACLF) has not been well documented. The authors' aimed to compare the clinical presentations and long-term outcome after standard treatments between patients with ALF versus those with ACLF.

Material and Method: The authors performed a retrospective review of 39 patients with Wilson's disease, at the King Chulalongkorn Memorial Hospital between January 2003 and December 2011. Primary outcome of the present study was liver complications or deaths from any cause. ACLF was defined as an acute hepatic insult in patients with previously diagnosed or undiagnosed chronic liver disease, whereas ALF was defined as an occurrence in the absence of any preexisting liver disease.

Results: Twenty-two of 39 patients (56.4%) presented predominantly with hepatic symptoms with the mean duration of follow-up of 7.7±8.5 years. Ten of them (45%) presented with ALF, whereas 12 patients (55%) presented with ACLF. Patients with ALF showed a significantly earlier age of onset of presenting symptoms than those with ACLF (15.4±4.5 vs. 28.1±13.0 years; $p<0.05$). The mean baseline of 24-hour urinary copper in patients with ALF was higher than those found in ACLF (1,645±1,406 vs. 441±434 mg/day; $p<0.05$, respectively). Fourteen patients (63.6%) improved with supportive care and chelating agents. No significant difference of clinical improvement was found between patients presented with ALF and ACLF (80% vs. 50%; $p = 0.19$). By using the survival analysis, the mean duration time to liver complications or all cause of death in patients with ALF was significantly longer than those with ACLF (16.2±2.3 years vs. 8.5±3.2 years; $p = 0.012$) as well as higher cumulative percent of free a period from liver complication or death during a 9-year period (80% vs. 21%, $p = 0.012$).

Conclusion: Patients with Wilson's disease presenting with acute-on-chronic liver failure manifested symptoms later and had more liver complications than patients with acute liver failure, as well as a lower cumulative free period from liver complication or death.

Keywords: Long-term outcome, Wilson's disease, Acute liver failure, Acute-on-chronic liver failure

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