Abstract

Wilson's disease is an autosomal recessive inherited disorder of copper metabolism resulting in accumulation of copper in various tissues. The hallmarks of the disease are the presence of liver disease, neurologic symptoms, and Kayser-Fleischer corneal rings. Until very recently, Wilson's disease was believed to be very rare. By a population-based approach, the incidence of Wilson's disease was estimated to be approximately 1:30,000 to 50,000 (Ireland: 17/10^6 live births [34]; former East-Germany: 29 [1]) with a gene frequency of 1:90 to 1:150; however, these estimations were mostly based on adolescents or adults presenting with neurologic symptoms. More recent data, however, indicate that neurologic symptoms occur only in about half of patients with Wilson's disease. Thus, the incidence of Wilson's disease was underestimated by these studies.