

# Analysis of Unusual Accumulation of Creutzfeldt-Jakob Disease Cases in Orava and Liptov Regions (Northern Slovak Focus) 1983-2000

Maďar R.<sup>1</sup>, Maslenová D.<sup>2</sup>, Ranostajová K.<sup>3</sup>, Straka Š.<sup>1</sup>, Baška T.<sup>1</sup>

<sup>1</sup> Institute of Epidemiology, Jessenius Faculty of Medicine, Comenius University, Martin

<sup>2</sup> Department of Epidemiology, State Health Institute, Liptovský Mikuláš

<sup>3</sup> Department of Epidemiology, State Health Institute, Dolný Kubín, Slovak Republic

## Summary

*While familial cases of Creutzfeldt-Jakob disease are extremely rare all over the world, 3 familial clusters were observed between 1983-2000 in a relatively small area situated in the North of Slovakia. Prevalence of CJD in this area exceeded the overall prevalence in Slovakia more than 8 times. The majority of CJD patients admitted consuming sheep brain. Most patients lived in small secluded villages with rather common familial intermarriage. CJD affected both sexes equally. All patients were prior to the disease mentally normal individuals. Shortly after the onset of CJD their mental status deteriorated remarkably with an average survival rate of 3.6 months.*

**Key words:** Creutzfeldt-Jakob disease, cluster, prevalence, familial intermarriage, sheep breeding

**Address for correspondence:** R. Maďar, Institute of Epidemiology, Jessenius Faculty of Medicine, Comenius University, Sklabinská 26, Martin, 037 53 Slovakia. E-mail: [MADAR@jfmed.uniba.sk](mailto:MADAR@jfmed.uniba.sk),