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ABSTRACT

An Evaluation of the Danish National Clinical Guidelines for von Hippel-Lindau (VHL)

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Purpose: von Hippel-Lindau (VHL) is a rare hereditary and potentially fatal cancer syndrome. Because of its unpredictable manifestations from various organ systems, surveillance is not linked to a single department and may therefore be incomplete. As one of the first countries in the world, Denmark published national guidelines for the surveillance of patients with manifest and possible VHL in 2005. The present study is the first of its kind where patients with suspected and manifest VHL are followed according to national guidelines at a single institution, and the purpose was to evaluate 1) to what extend the guidelines were being followed and 2) what findings were disclosed.

Methods: The study included 27 individuals with diagnosed (14 patients) or suspected (13 patients) VHL, observing the Danish VHL-guidelines at the Department of Neurosurgery, Rigshospitalet, Denmark from October 2002 to April 2008. The data were collected by reviewing the patients´ records. Results: 48% of the patients had manifestations revealed that influenced the treatment, and 26% of the patients had asymptomatic manifestations demonstrated. All investigations were conducted with a lower frequency than recommended.

Conclusions: The study shows that the national clinical guidelines were not being fully complied with. The investigations revealing the most serious VHL manifestations were carried out with a frequency closest to the recommendations. Many investigations lead to clinical consequences. Therefore we recommend that all patients suspected with or suffering from VHL are monitored according to structured clinical guidelines.