

PubMed

U.S. National Library of Medicine
National Institutes of Health



Display Settings: Abstract

[Cancer.](#) 2010 Aug 24. [Epub ahead of print]

Incidence of atypical teratoid/rhabdoid tumors in children: a population-based study by the Austrian Brain Tumor Registry, 1996-2006.

Woehrer A, Slavic I, Waldhoer T, Heinzl H, Zielonke N, Czech T, Benesch M, Hainfellner JA, Haberler C; on behalf of the Austrian Brain Tumor Registry.

Institute of Neurology, Medical University of Vienna, Vienna, Austria.

Abstract

BACKGROUND: Atypical teratoid/rhabdoid tumors are highly malignant embryonal central nervous system (CNS) tumors that were defined as an entity in 1996. As compared with other malignant CNS tumors, their biological behavior is particularly aggressive, but patients may benefit from an intensified treatment. Atypical teratoid/rhabdoid tumors display a complex histomorphology, which renders them prone to misdiagnosis. They occur predominantly in young children, with an estimated prevalence of 1% to 2% among all pediatric CNS tumors. However, population-based data on the incidence of these tumors are not yet available.

METHODS: A nation-wide survey of malignant high-grade CNS tumors (World Health Organization grade III/IV), diagnosed in children (aged birth to 14 years) from 1996 to 2006 was conducted by the Austrian Brain Tumor Registry. A central histopathology review was performed including the assessment of SMARCB1 (INI1) protein status.

RESULTS: A total of 311 newly diagnosed, malignant CNS tumors were included. Atypical teratoid/rhabdoid tumors constituted the sixth most common entity (6.1%), referring to an age-standardized incidence rate of 1.38 per 1,000,000 person-years in children. Peak incidence was found in the birth to 2 years age group, where they were as common as CNS primitive neuroectodermal tumors and medulloblastomas. A total of 47.4% of atypical teratoid/rhabdoid tumors were initially diagnosed, whereas 52.6% were retrospectively detected by the central review. The 5-year survival of atypical teratoid/rhabdoid tumor patients was 39.5%, with 66.7% in the correctly diagnosed group versus 15.0% in the not recognized group ($P = .0469$).

CONCLUSIONS: Clinicians and pathologists should be aware of the high incidence of atypical teratoid/rhabdoid tumors in young children to optimize diagnostic and therapeutic management of patients with these tumors. *Cancer* 2010. (c) 2010 American Cancer Society.

PMID: 20737418 [PubMed - as supplied by publisher]

[LinkOut - more resources](#)