

Adult survivors of childhood eye cancer experience few cognitive or social setbacks

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Most long-term survivors of retinoblastoma, particularly those who had been diagnosed with tumors by their first birthdays, have normal cognitive function as adults, according to a St. Jude Children's Research Hospital study. The research, which appears in the current issue of the journal *Cancer*, found that the vast majority of survivors work full time, live independently and fulfill other milestones of adult life.

The study is the first to examine how adult survivors of retinoblastoma fare cognitively and socially decades after their diagnosis. The findings contrast with research involving survivors of other childhood cancers that suggest a younger age at cancer diagnosis may put survivors at risk for reduced cognitive functioning later in life.

"As a group, adult survivors of retinoblastoma are doing quite well based on their cognitive functioning and attainment of adult social milestones," said the study's first and corresponding author Tara Brinkman, Ph.D., an assistant member of the St. Jude Department of Epidemiology and Cancer Control and the Department of Psychology.

"This suggests that for children whose visual system is damaged very early in life, the brain may compensate by reorganizing areas responsible for processing visual information to enhance processing of verbal and auditory information," Brinkman said. "This highlights the importance of early intervention and rehabilitation for these patients."

Retinoblastoma is diagnosed in about 350 children annually in the U.S.,



and 95 percent of patients are younger than 5 years old when their tumor is identified. Today, more than 95 percent of retinoblastoma patients become long-term survivors. Previous research with retinoblastoma survivors focused on cognitive functioning in childhood. The results were mixed.

The 69 adult survivors in this study scored in the normal range on measures of verbal intelligence, attention, memory, executive functioning and their ability to process information. Their performance was above average for certain tasks, including non-verbal reasoning, but well below average for the fine motor dexterity needed for tasks like handwriting.

Seventy percent of survivors lived independently, and 62 percent were married or living as married couples. Fifty-eight percent had completed college. Three-quarters of survivors worked full-time, but more than half reported earning less than \$20,000 annually.

Age at diagnosis was a more powerful predictor of adult cognitive functioning than whether survivors developed tumors in one or both eyes. Survivors whose retinoblastoma was identified by their first birthdays scored better on almost all measures of verbal functioning than did survivors diagnosed later. Diagnosis during the first year of life was associated with significantly better performance on tests of short- and long-term verbal memory, verbal learning and verbal intelligence.

While additional research is needed to understand the results, Brinkman and her colleagues noted that the findings are consistent with other reports that the developing central nervous system can compensate for early vision loss.

Patients diagnosed as infants are also more likely to have tumors in both eyes than children whose cancer is identified later, said co-author Rachel



Brennan, M.D., an assistant member of the St. Jude Department of Oncology. "These children start rehabilitation and other interventions very early in life that stimulate sensory development and learning to compensate for possible visual impairment," she said.

Study participants were on average 33 years old and 31 years from their retinoblastoma diagnoses. All were enrolled in the St. Jude Lifetime Cohort Study (St. Jude LIFE), which brings survivors treated at St. Jude as children back to the hospital for several days of extensive testing. St. Jude LIFE is an ongoing study designed to better understand the causes and severity of the challenges facing childhood cancer survivors as they age.

Whole brain irradiation also surfaced as a risk factor for poor performance on measures of short- and long-term verbal memory, even among survivors whose overall cognitive functioning fell within the normal range. Because few survivors who developed retinoblastoma in a single eye were treated with radiation, researchers analyzed the treatment's impact on the 24 survivors who had tumors in both eyes and factored in patient age at diagnosis. Thanks to novel chemotherapy regimens and aggressive focal therapy such as laser therapy or cryotherapy, Brennan said less than 3 percent of retinoblastoma patients at St. Jude receive radiation now to save an eye.

When asked to rate their own cognitive and behavior functioning, survivors were within the normal range for most measures. But survivors were significantly more likely than U.S. adults of a similar age to report problems with working memory or in completing tasks.

More information: "Cognitive function and social attainment in adult survivors of retinoblastoma: A report from the St. Jude Lifetime Cohort Study." Tara M. Brinkman, Thomas E. Merchant, Zhenghong Li, Rachel Brennan, Matthew Wilson, Mary Ellen Hoehn, Ibrahim Qaddoumi, Sean



Phipps, Deokumar Srivastava, Leslie L. Robison, Melissa M. Hudson, and Kevin R. Krull. *Cancer*; Published Online: November 24, 2014 DOI: 10.1002/cncr.28924

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