

Long Term Follow-Up Among Survivors of Childhood Ewing's Sarcoma: A Report from the Childhood Cancer Survivor Study (CCSS)

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ABSTRACT

Purpose: Long-term survival for Ewing's sarcoma (ES) has improved, although often at considerable cost to affected individuals. We report mortality, second neoplasms (SN), chronic medical condition (CMC) rates, and health status of ES survivors.

Patients and Methods: A retrospective cohort of 5-year survivors of ES (n=403) diagnosed between 1970-1986, under age 21 at diagnosis was compared to general population rates or a sibling control group (n=3,899) using data from the CCSS.

Results: Mean age at time of interview was 26 years. Interval from cancer diagnosis to interview for survivors was 14 years. Cumulative mortality for survivors was 24.5% at 25 years from diagnosis. Standardized mortality ratio (SMR) was 13.3 (95% confidence interval [CI], 10.8 – 16.1). SMR for irradiated survivors was 14.0 (95% CI, 11.1-17.4) compared to 4.1 (95% CI, 1.5-8.7) for non-irradiated survivors (P = 0.007). A total of 33 SN occurred among 403 survivors, 26 SN were in radiation-treated patients. Standardized incidence ratio (SIR) for all SN was 5.5 (95% CI, 3.8 – 7.9). The SIR for irradiated survivors was 6.1 (95% CI, 4.1 – 9.1) whereas the SIR was 3.3 (95% CI, 1.1 – 10.2) for non-irradiated survivors. Survivors were more likely to report a CMC (hazard ratio [HR], 5.9; 95% CI, 4.8-7.2) and severe or life threatening CMC (HR, 11.8; 95% CI, 9.0 -15.4) compared to siblings. After adjusting for gender and ethnicity, survivors were 21.2 times as likely to have a grade 3-4 cardiac condition (95% CI, 9.4-47.6). Survivors also reported significantly higher adverse general health, mental health, functional impairment and activity limitations compared to siblings (P <0.001).

Conclusions: Long-term survivors of ES exhibit excess mortality and morbidity from therapy, particularly those who received radiation therapy as part of their treatment. Recognition of these outcomes and interventions designed to reduce or eliminate them will improve overall health of ES survivors.