PAPER 55

Do High- and Low-Risk Pediatric Rhabdomyosarcoma Survival Estimates Equalize 5 Years After Diagnosis? *A Cause-specific Conditional Survival Analysis*

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Background: Conditional survival (CS) describes a patient's prognosis based on the time a patient has survived since initial diagnosis. CS provides valuable information on prognosis and how it changes over time. This is especially important during patient counseling. Rhabdomyosarcoma is the most common soft tissue sarcoma in childhood, with multiple factors impacting overall survival including age, tumor size, stage, and histology. Although overall survival of pediatric rhabdomyosarcoma (RMS) has been extensively studied, CS related to RMS has not been reported.

Questions/Purposes: The purpose of this study was to determine how cause-specific 5-year survival is impacted based on each additional year of survival. Furthermore, patient specific demographics, tumor characteristics, and surgical management were collected to determine their impact on CS.

Patients and Methods: A total of 1,997 patients ages 18 years and younger with rhabdomyosarcoma diagnosed between 2000-2018 were identified through the Surveillance, Epidemiology, and End Results (SEER) database. All subtypes of rhabdomyosarcoma were included based on ICD-O-3 morphology. Demographics, stage, histology, grade, size, site, and surgical procedure were collected. Tumor sites were classified as "favorable" in the head/neck, biliary tract, or genitourinary system, and "unfavorable" at all other sites. Survival for an additional 5 years conditional on a patient having already survived for 0 to 5 years from diagnosis was evaluated with an actuarial life table analysis. An unpaired, two-tailed t test was used to compare continuous variables.

Results: The overall cause-specific 5-year survival at time of diagnosis was 67.9%. At 1 and 5 years from diagnosis the estimated 5-year survival increased to 72% and 95.3%, respectively. The baseline 5-year survival was 81.1% for local/regional disease and 38.4% for metastatic disease. At 1 year from diagnosis, the 5-year survival for metastatic RMS had no significant improvement from baseline. However, those at the 5-year survival interval had a marked improvement in survival for an additional 5 years compared to baseline (38.4% vs 90.7%, $p<0.001$). There were no significant differences in 5-year survival between grades at diagnosis, 1 year, or 5 years. Survival was lowest in patients who were ≥10 years old, tumor size >5 cm, alveolar histology, distant metastases, and unfavorable locations **(Table 1)**. Although these factors portend the worst prognosis at initial diagnosis, they demonstrated the greatest increase in conditional survival from baseline to 5 years. At 5 years from diagnosis, there was no longer a difference in 5-year survival between patients ages 10-18 vs. ages 1-9 ($p=0.95$), and tumors <5 cm vs. >5cm ($p=0.13$). A small difference in survival remained between alveolar vs. embryonal histology ($p=0.01$), distant disease vs. local/regional ($p=0.01$), and unfavorable vs. favorable tumor sites ($p=0.02$) at 5 years from diagnosis **(Figure 1)**.

Conclusion: 5-year conditional survival for patients with RMS improves with each additional year of survival, especially for patients with initial high-risk characteristics. This indicates that patients with high-risk characteristics that survive for 5 years may have similar long-term prognosis as some patients with less severe features. This conditional survival data provides a valuable resource to counsel patients and direct current and future treatment.

Figure 1. Cause-specific conditional survival of pediatric patients with rhabdomyosarcoma. Each point represents the percent survival for an additional 5 years, given the patient has already survived 0 to 5 years after diagnosis. Error sheaths represent 95% confidence interval.

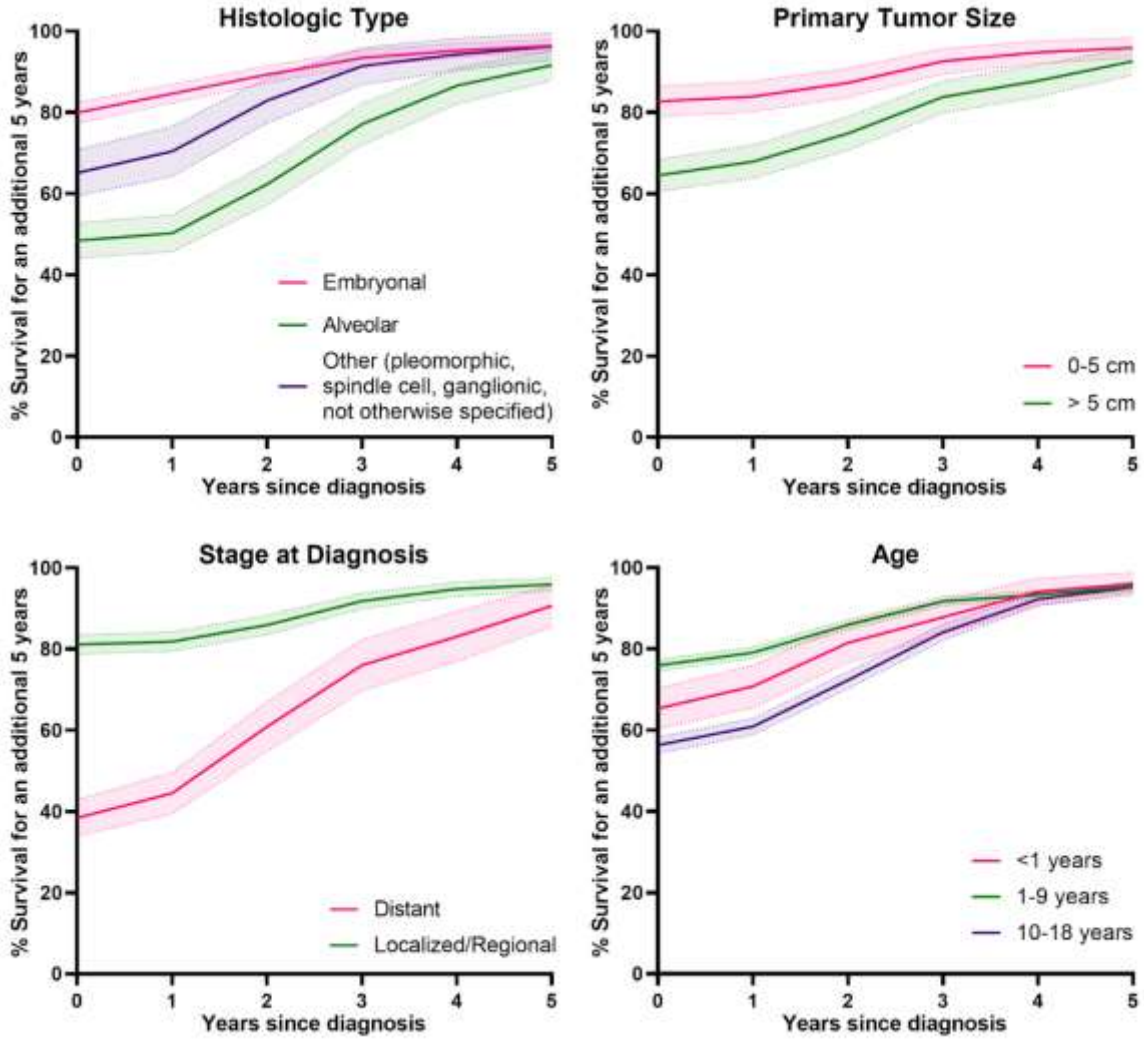

Table 1. Cause-specific 5-year survival of pediatric rhabdomyosarcoma at baseline, conditional on 1 year of survival, and conditional on 5-years of survival after diagnosis

Table 1: Cause-Specific 5-Year Survival of Rhabdomyosarcoma at Baseline, conditional on 1 year of survival, and conditional on 5 years of survival				
Variables	**N**	**Baseline (95% CI)**	**1 year (95% CI)**	**5 years (95% CI)**
Overall	1,997	67.9% (65.7%-70.1%)	72.0% (69.6%-74.1%)	95.3% (93.6%-96.5%)
Age				
<1 year	107	65.3% (54.6%-74.0%)	70.8% (59.3%-79.6%)	96.0% (84.8%-99.0%)
1-9 years	1,142	75.9% (73.1%-78.5%)	79.1% (76.3%-81.7%)	95.3% (93.1%-96.8%)
10-18 years	748	56.3% (52.3%-60.0%)	60.9% (56.8%-64.8%)	95.2% (91.8%-97.2%)
Sex				
Male	1,145	68.2% (65.2%-71.0%)	71.3% (68.2%-74.2%)	94.8% (92.4%-96.4%)
Female	852	67.6% (64.1%-70.9%)	72.9% (69.3%-76.1%)	96.0% (93.4%-97.6%)
Race				
White	988	69.8% (66.6%-72.7%)	73.3% (70.0%-76.2%)	95.8% (93.5%-97.3%)
Black	312	66.2% (60.2%-71.5%)	70.7% (64.5%-76.1%)	94.5% (88.7%-97.3%)
American Indian/Alaska Native	19	44.5% (21.6%-65.2%)	50.0% (24.5%-71.0%)	100.00%
Asian/Pacific Islander	133	63.9% (53.9%-72.2%)	70/7% (60.2%-78.8%)	100.00%
Hispanic	535	67.2% (62.6%-71.3%)	71.1% (66.3%-75.3%)	93.4% (89.2%-96.1%)
Stage				
Localized/Regional	1,077	81.1% (78.6%-83.3%)	81.8% (79.3%-84.1%)	95.9% (94.0%-97.2%)
Distant	480	38.4% (33.9%-42.8%)	44.5% (39.3%-49.5%)	90.7% (84.1%-94.6%)
Grade				
Grade I/II	44	75.2% (58.6%-85.8%)	76.9% (60.2%-87.3%)	96.0% (74.8%-99.4%)
Grade III	136	70.9% (62.3%-77.9%)	73.3% (64.3%-80.3%)	92.5% (82.7%-96.8%)
Grade IV	250	74.6% (68.5%-79.8%)	77.8% (71.4%-82.9%)	94.6% (89.0%-97.4%)
Size				
< 5 cm	427	82.7% (78.6%-86.0%)	83.9% (79.7%-87.3%)	95.9% (92.4%-97.8%)
> 5 cm	596	64.5% (60.3%-68.3%)	67.9% (63.6%-71.9%)	92.6% (88.3%-95.3%)
Surgery				
Local excision or limb salvage	583	74.3% (70.3%-77.9%)	74.3% (70.1%-78.0%)	93.7% (90.2%-96.0%)
Partial or total amputation of limb	232	86.3% (80.7%-90.3%)	86.2% (80.5%-90.3%)	96.9% (91.9%-98.8%)
No surgery	866	54.3% (50.6%-57.8%)	60.9% (57.0%-64.7%)	95.2% (92.0%-97.1%)
Histology				
Embryonal	1,087	79.9% (77.2%-82.3%)	84.6% (82.0%-86.9%)	96.4% (94.4%-97.8%)
Alveolar	590	48.4% (44.0%-52.6%)	50.2% (45.5%-54.6%)	91.6% (86.9%-94.7%)
Other	320	65.1% (59.1%-70.5%)	70.4% (64.0%-76.0%)	96.2% (91.2%-98.4%)
Primary Tumor Site				
Favorable sites (head/neck, biliary and genitourinary)	816	80.4% (77.3%-83.1%)	83.5% (80.4%-86.2%)	97.1% (94.9%-98.3%)
Unfavorable sites	1,181	59.3% (56.2%-62.3%)	63.6% (60.3%-66.7%)	93.6% (90.9%-95.6%)