



Published in final edited form as:

Pediatr Blood Cancer. 2023 February ; 70(2): e29983. doi:10.1002/pbc.29983.

Psychosocial Outcomes and Quality of Life Among School Age Survivors of Retinoblastoma

Melanie Morse, PhD¹, Kendra Parris, PhD¹, Ibrahim Qaddoumi, MD, MS¹, Sean Phipps, PhD¹, Rachel C. Brennan, MD¹, Matthew W. Wilson, MD², Carlos Rodriguez-Galindo, MD¹, Kristin Goode, MS¹, Victoria W. Willard, PhD¹

¹St. Jude Children's Research Hospital, Memphis, Tennessee

²University of Tennessee Health Sciences Center, Memphis, Tennessee

Abstract

Background: Retinoblastoma is the most common intraocular childhood cancer and is typically diagnosed in young children. With increasing number of survivors and improved medical outcomes, long-term psychosocial impacts need to be explored. Thus, the current study sought to assess functioning in school-aged survivors of retinoblastoma.

Procedure: Sixty-nine survivors of retinoblastoma underwent a one-time evaluation of psychosocial functioning. Survivors ($M_{age}=10.89$ years, $SD=1.07$ years; 49.3% male; 56.5% unilateral disease) and parents completed measures of quality of life (QoL; PedsQL) and emotional, behavioral, and social functioning (PROMIS Pediatric Profile, BASC-2 parent report). Demographic and medical variables were also obtained.

Results: On the whole, both survivors and caregivers indicated QoL and behavioral and emotional health within the typical range of functioning. Survivors reported better physical QoL compared to both parent report and a national healthy comparison sample, whereas caregivers reported that survivors experienced lower social, school, and physical QoL than a healthy comparison. Regarding behavioral and emotional health, survivors indicated more anxiety than a nationally representative sample. Parents of female survivors endorsed lower adaptive scores than parents of male survivors.

Conclusions: Results indicated that survivors of retinoblastoma reported QoL and behavioral and emotional health within normal limits, although parents appear to perceive greater impairment across several assessed domains. Understanding both survivor- and parent-report remains important for this population. Future research should explore psychosocial functioning of these survivors as they transition to adolescence and early adulthood given the increased independence and behavioral and emotional concerns during these developmental periods.

Correspondence Victoria W. Willard, Department of Psychology, 262 Danny Thomas Place, Memphis, TN 38105, USA; Phone: (901) 595-5336; victora.willard@stjude.org.

This study was accepted for poster presentation at the 2021 Virtual Society of Pediatric Psychology Annual Conference (April 8-10, 2021).

Keywords

retinoblastoma; psychosocial outcomes; quality of life; pediatric childhood survivors

Retinoblastoma is the most common intraocular tumor of childhood, affecting approximately 8,000 children worldwide each year.¹ In the United States, 200-300 new cases are diagnosed annually.² The disease primarily impacts young children, with most cases occurring before five years of age; average age of onset in the United States is two years.^{2,3} Survival rates currently exceed 95% in the United States and other developed countries.¹ As advances in medicine have improved survival rates, more long-term survivors are at risk of treatment-related medical and psychological morbidities, including visual deficits, cosmetic deformities, other health conditions, and impairments in mental health functioning.⁴⁻⁶ Medical efforts to maximize ocular salvage and preserve vision could allay some of these long-term consequences and positively impact long-term quality of life (QoL).⁵

Extant literature has focused largely on the visual, physical, and neurocognitive outcomes of retinoblastoma diagnosis and treatment.⁷⁻¹⁰ While medical outcomes and long-term medical risks are well documented,^{7-9,11} less is known about long-term psychosocial outcomes. As more children with retinoblastoma are surviving, understanding the impact of their disease on their QoL and psychosocial functioning is crucial. In one study of adult survivors, over one-third of the sample reported that their illness experience continued to impact their life “a lot” in adulthood.¹² In another study of adult survivors, over half of the sample perceived restrictions in their daily life as a result of their prior illness experience.¹³ Not surprisingly, adult survivors who experienced bilateral disease or complete blindness reported worse vision-targeted QoL.⁵ In contrast, more general QoL may be less compromised. For example, Ford and colleagues⁴ found that adult survivors were less likely to report depression, somatization, and global problems than a control sample. Certain childhood experiences, such as bullying, may serve as specific risk factors for later perceived impairments in QoL.⁶

Results of studies examining QoL in childhood survivors of retinoblastoma are variable. In some studies, survivors¹⁴⁻¹⁵ and parents¹⁶⁻¹⁷ reported lower QoL than controls. However, another study found that parent and child reports differed significantly, with parents reporting much poorer QoL for their child, and survivors themselves indicating good QoL comparable to controls.¹⁸ Variable findings from these past studies likely relate to multiple factors, such as severity of disease, types of treatments received, use of parent versus child report, and age of the sample. Specifically, some evidence suggests that younger survivors^{15,18} and those with normal visual acuity¹⁸ report better QoL, whereas survivors with more motor problems may experience worse QoL.¹⁷ Lastly, disease laterality has been examined as a predictor of QoL. Ford and colleagues⁴ found that survivors who experienced bilateral disease were more likely than those with unilateral disease to be unmarried, have a lower income, and have a history of special education; however, the two groups did not differ in mental health functioning or post-traumatic stress symptoms. In fact, survivors

of bilateral disease reported significantly more post-traumatic growth than survivors of unilateral disease.⁴

Taken together, the existing literature highlights variability in ratings of QoL among survivors of retinoblastoma. Additionally, long-term psychosocial functioning of survivors remains unclear. Previous research found that the first onset of psychiatric disorders typically occurs in childhood and adolescence,¹⁹ making childhood a particularly critical timepoint to consider. Additional factors that may impact survivors' functioning including enucleation status and respondent type (e.g., self versus proxy report) are also less studied in the literature. To address limitations in past studies, the current study aimed to examine the psychosocial functioning and QoL of a sample of survivors of retinoblastoma who were approximately 10 years of age. Measures were obtained from both survivors and parents, and demographic and medical variables were explored as predictors of outcomes.

Materials and Methods

Participants

Participants included children who were treated for retinoblastoma on an institutional treatment protocol. As part of their enrollment, patients participated in a protocol evaluation of psychosocial functioning. The protocol was approved by the Institutional Review Board and consent/assent was provided at the time of diagnosis and again at the age 10 assessment. Sixty-nine survivors of retinoblastoma, and a caregiver, completed measures as part of this evaluation ($M_{age}=10.89$ years, $SD=1.07$ years; 49.3% male; 56.5% unilateral disease). On average, participants were diagnosed with retinoblastoma 9.37 years prior to the evaluation (range 6.5-12 years since diagnosis). See Table 1 for demographic and medical information.

Measures

QoL—The Pediatric Quality of Life Inventory (PedsQL)²⁰⁻²¹ was administered to both participants and their caregivers to assess QoL. The PedsQL has 23 questions distributed in the physical, emotional, social, and school dimensions, with the latter three scales comprising the psychosocial health domain. A 5-point Likert scale is utilized across both forms, and items are reverse scored and linearly transformed to a 0-100 scale with higher scores indicating better QoL. Although it is at present unclear what scores may be normative for young survivors of retinoblastoma, research has identified means and standard deviations for healthy, chronically ill, and acutely ill children across the total and scale scores, respectively by both self- and parent-proxy report.²¹ Both the PedsQL child and caregiver reports demonstrated excellent internal consistency reliability for the four identified scales (i.e., physical, emotional, social, school).²¹ The present study utilized scores from physical, emotional, social, and school dimensions of the PedsQL.

Behavioral and Emotional Health—Retinoblastoma survivors completed the PROMIS Pediatric Profile,²²⁻²³ a self-report measure of internalizing and externalizing symptoms. The PROMIS includes 49 items assessing anxiety, depression, fatigue, peer relations, and pain; corresponding T-Scores are provided for each of these five domains. Although T-scores may be theoretically interpreted relative to a normal distribution with a mean of 50 and

a standard deviation of 10, recent research has utilized nationally representative data to establish cut-points for child and proxy reports on the PROMIS.²⁴ Higher scores on the PROMIS are indicative of more problems in the respective domain. The current study utilized scores from three key domains of the PROMIS: anxiety, depressive symptoms, and peer relationships.

Parents of retinoblastoma survivors completed the second edition of the Behavior Assessment System for Children (BASC-2),²⁵ a broad-band assessment of children's behavioral and emotional functioning. On the BASC-2, T-scores are provided for both clinical scales (Hyperactivity, Aggression, Conduct Problems, Anxiety, Depression, Somatization, Atypicality, Withdrawal, Attention Problems) and adaptive scales (Adaptability, Social Skills, Leadership, Activities of Daily Living, Functional Communication). Additionally, scoring yields T-Scores for composite scales (Externalizing Problems, Internalizing Problems, Behavioral Symptoms Index, Adaptive Skills). Higher scores are indicative of more problems for scores on the clinical scales of the BASC-2, whereas lower scores are indicative of greater difficulties for the adaptive scales. For the clinical scales, T-scores less than or equal to 60 indicate functioning in the average range. T-scores between 61 and 70 indicate domains that are at-risk for difficulties and scores 71 and above indicate clinically significant concerns for clinical scales. In contrast, T-Scores between 31 and 40 indicate at-risk concerns for adaptive scales on the BASC-2, and those scores 30 or below suggest clinically significant concerns within a given adaptive scale. For the current study, only the four composite scales were used in analyses.

Data Analytic Plan

Independent samples t-tests were utilized to analyze mean differences in QoL scores (social, emotional, school, physical) by respondent (survivor, caregiver), laterality (unilateral, bilateral), enucleation status (yes, no), and sex (male, female). One-sample t-tests were used to assess mean differences between QoL as reported by survivors and caregivers in the current sample and previously established normative scores among healthy children.

Survivors' scores on key behavioral and emotional health outcomes (anxiety, depression, peer relations) were compared to normative cut-points from a nationally representative sample. Independent samples t-tests were also used to test mean differences in key areas of survivor-reported behavioral and emotional health (anxiety, depression, peer relations) and caregiver-reported behavioral and emotional health (internalizing symptoms, externalizing symptoms, behavioral symptoms, adaptive behavior) by laterality, enucleation status, and sex.

Results

QoL

Generally, both survivors and caregivers reported QoL within normal limits across domains of day-to-day functioning. There were no significant differences between parent-proxy and child self-report for social, emotional, and school functioning domains. However, patient-reported physical QoL ($M=89.23$, $SD=13.39$) was significantly higher than caregiver-

reported physical QoL ($M=81.49$, $SD=22.42$), $t(133)=-2.43$, $p<0.05$; see Figure 1. No other differences in patient and caregiver reported QoL were observed across domains.

With respect to disease laterality, there were no differences in ratings of QoL from patients with unilateral versus bilateral diagnoses across the social ($t(65)=0.48$), emotional ($t(64)=-0.71$), school ($t(64)=-1.18$), or physical functioning ($t(65)=-1.49$) domains. Similarly, there were no differences by laterality in caregiver ratings of QoL across social ($t(66)=0.73$), emotional ($t(66)=0.37$), school ($t(66)=0.04$), or physical functioning ($t(66)=-1.80$) domains.

Given the influence of enucleation on vision-related QoL in the extant literature, the impact of enucleation status on broad domains of QoL as assessed by the PedsQL was examined. There was no difference in scores of survivors who underwent enucleation versus those who did not on social ($t(64)=-1.84$), emotional ($t(63)=-1.96$), school ($t(63)=-1.02$), or physical functioning ($t(64)=-1.06$). Similarly, there was no difference in QoL scores reported by parents of survivors of retinoblastoma with or without enucleation across assessed domains including social ($t(65)=-1.13$), emotional ($t(65)=-1.17$), school ($t(65)=-0.32$), or physical functioning ($t(65)=-0.56$).

In order to assess any differences by sex, QoL scores of male and female survivors were examined. Male and female survivors of retinoblastoma did not differ in their ratings of social ($t(65)=.26$), emotional ($t(64)=1.50$), school ($t(64)=.74$), or physical functioning ($t(65)=-1.13$). Parents' ratings of QoL for male and female survivors did not differ across assessed domains of social ($t(66)=0.98$), emotional ($t(66)=1.38$), school ($t(66)=.18$), or physical functioning ($t(66)=-0.79$). See Table 2 for a summary of QoL ratings by caregiver, laterality, enucleation status, and sex.

Both survivors' and caregivers' reported QoL scores domains were also compared to normative scores among healthy children (Table 3).²¹ Survivor report was significantly higher than normative scores for healthy children for the physical domain of QoL ($t(66)=2.95$, $p<.01$), but no differences between scores were observed for the social ($t(66)=-1.99$), emotional ($t(65)=-0.98$), or school ($t(65)=-0.95$) domains. In contrast, caregiver proxy-report was significantly lower than normative proxy scores for healthy children on the social ($t(67)=-2.97$, $p<0.01$), school ($t(67)=-2.65$, $p<0.05$, and physical ($t(67)=-2.88$, $p<0.01$) domains of QoL. Caregiver report was not significantly different than proxy comparison scores for emotional QoL, $t(67)=-1.18$.

Behavioral and Emotional Health

Survivor Report—Survivors of retinoblastoma rated their behavioral and emotional health symptoms in the average range of functioning across assessed domains. Results of the PROMIS indicate that patient-reported behavioral and emotional health symptoms were above the nationally established median value for the PROMIS²⁴ with respect to anxiety ($M=46.46$, $SD=11.82$; $t(65)=3.07$, $p<0.01$), but were no different than the median value for depression ($t(63)=-1.61$) or peer relations ($t(64)=-.20$). Results are summarized in Table 4.

Analyses of disease laterality indicated that unilateral versus bilateral disease did not have an effect on survivors' ratings of anxiety ($t(64)=0.10$), depression ($t(61)=-0.55$), or peer relations ($t(62)=1.88$). Comparison of survivors who underwent enucleation versus those who did not revealed that anxiety ($t(64)=0.45$), depression ($t(61)=0.66$), and peer relations ($t(62)=-1.99$) did not differ by enucleation status. Similarly, male and female survivors of retinoblastoma did not report significantly different anxiety ($t(64)=-1.7$), depression ($t(61)=-1.52$), or peer relations ($t(62)=-0.22$) scores. Mean scores for each group are summarized in Table 5.

Parent Report—Parents generally reported behavioral and emotional health symptoms in the average range of functioning for survivors as measured by the BASC-2.

Specifically, internalizing symptoms ($M=50.75$, $SD=11.71$), externalizing symptoms ($M=46.82$, $SD=9.28$), behavioral problems ($M=48.84$, $SD=9.66$), and adaptive skills ($M=50.40$, $SD=9.64$) were consistent with normative expectation. However, 19 (28.36%) parents rated their children as having elevated internalizing problems, although only 3 (4.48%) parents reported internalizing scores in the clinically significant range. Even fewer parents ($n=6$; 8.96%) rated their children as having elevated externalizing behaviors, with half of those parents endorsing clinically significant externalizing concerns in their children ($n=3$; 4.48%). Nine (13.43%) parents endorsed symptoms that produced elevations on the behavioral symptoms scale. Of these nine parents, 4 (5.97%) reported that behavior of their children fell within the clinically significant range. Similarly, 9 (13.43%) parents rated the adaptive skills of their children in either the at-risk or clinical range on the BASC-2, with 3 (4.48%) parents endorsing concerns in the clinical range.

Disease laterality was examined to determine whether parents of survivors with unilateral versus bilateral retinoblastoma endorsed different rates of behavioral and emotional health symptoms. Results indicated no differences in reports from parents of children with unilateral versus bilateral disease for assessed domains of internalizing ($t(65)=-0.40$), externalizing ($t(65)=0.02$), behavioral ($t(65)=-0.71$), or adaptive skills ($t(65)=0.68$). Similarly, parents of children with and without enucleation did not differ in reports of internalizing problems ($t(64)=1.68$), externalizing problems ($t(64)=-0.63$), behavioral symptoms ($t(64)=0.41$), or adaptive skills ($t(64)=-0.01$).

Sex was also examined to assess any differences in reports of behavioral and emotional health between parents of male and female survivors of retinoblastoma. Parents of females reported lower adaptive scores ($M=47.85$, $SD=8.30$) than parents of males ($M=52.88$, $SD=10.31$), $t(65)=2.20$, $p=0.03$. Specifically, parents of female survivors of retinoblastoma reported more concerns related to adaptive behavior (e.g., adjusts well to changes in family plans, attends to issues of personal safety, communicates clearly, will speak up if the situation calls for it),²⁵ although the mean still falls within the average range of functioning. Parents of male and female survivors of retinoblastoma did not differ in their reports of internalizing problems ($t(65)=-2.08$), externalizing problems ($t(65)=-0.99$), or behavioral symptoms ($t(65)=-1.49$). A summary of parent-reported behavioral and emotional health scores by laterality, enucleation status, and sex is provided in Table 5.

Discussion

The present study examined both QoL and broad behavioral and emotional symptoms among school-aged survivors of retinoblastoma. The findings add to the existing literature base in characterizing and describing key elements of psychosocial functioning among this population. Generally, survivors reported positive QoL within assessed domains of social, emotional, school, and physical functioning, with survivor-reported physical QoL significantly higher than established norms among healthy children. In contrast to survivor report, caregivers reported that their children experienced significantly lower social, school, and physical QoL than a normative sample of parents of healthy children. Interestingly, patients reported greater physical QoL than caregivers, although both patient and caregiver ratings indicated QoL within normal limits. This finding may be reflective of parents' potential lingering overprotection aimed at shielding any further ocular damage and is consistent with previous research that survivors report better perceptions of QoL than caregivers.¹⁸ It follows that parents who experience lingering concerns or worry about potential further damage may feel their children are less capable physically, thus reporting lower QoL related to physical functioning.

Patient- and caregiver-reported behavioral and emotional health symptoms fell within expectations across domains as well. While survivors in the current study indicated greater concerns related to anxiety compared to the median value from a nationally representative sample, it is important to qualify that the mean value of patients' anxiety scores fell within the average range and was not indicative of elevated anxiety symptoms. The current findings are consistent with previous research that adolescent cancer survivors report psychosocial functioning within normative outcomes,²⁶ and suggest that children with retinoblastoma are also resilient in post-treatment functioning.

Notably, parents of female survivors of retinoblastoma reported lower adaptive scores than parents of males. This finding contrasts previous research on parent-rated adaptive functioning of children who are survivors of brain tumors, which indicate no significant gender differences in perceived adaptive functioning.²⁷ Although the current finding is a statistically significant difference, it is likely not a clinically significant difference, especially considering average scores were well within normal limits for both sexes. Previous research has found that girls are more likely than boys to assist with household chores, childcare, and related tasks.²⁸ It may be the case that parents of girls have higher expectations within this domain, and that girls are tasked with assisting in key areas of adaptive behavior within a household (e.g., sibling care, additional chores), leading parents in turn to provide more stringent ratings of girls within this domain. Future research could continue to examine adaptive skills among survivors of retinoblastoma to further explore and clarify nuances within this finding.

For all assessed domains within both QoL and behavioral and emotional symptoms, there were no differences by disease laterality or enucleation status. This may indicate that school-age survivors of retinoblastoma are a resilient group and have been able to avoid long-term psychosocial impact. Given that retinoblastoma diagnosis typically occurs in the first years of life and survivors in this sample were, on average, nearly a decade post-diagnosis, it

may be the case that young age at diagnosis and treatment helps to mitigate the impact of disease-related insult. Survivors may feel that they have grown up knowing what life is like post-diagnosis and treatment, and consequently may have appropriate, longstanding coping skills in place.

Current findings have several implications for clinical practice. Given the observed difference between survivor- and parent-reported functioning across several domains, it is important to continue to consider both patients' and parents' perspectives of functioning. Indeed, previous research has established that parent proxy reports are often discrepant from survivor reports, so understanding both perspectives is crucial.²⁹ Additionally, although findings suggest overall resilience among survivors, a small percentage of parents did endorse clinically significant behavioral and emotional health concerns. Providers should continue to assess behavioral and emotional health during routine follow-up screenings and provide appropriate resources or referrals for those survivors who are experiencing elevated concerns.

Although the current study contributes to the existing literature of long-term functioning among survivors of retinoblastoma, it is not without limitations. First, it is possible that the PedsQL was not sensitive enough to detect any differences in vision-related QoL across assessed domains (e.g., laterality, enucleation status, sex). Previous research has utilized vision-specific measures of QoL among survivors of retinoblastoma,⁵ and a more targeted measure may prove more useful than broad domains such as physical QoL. Additionally, the current study was cross-sectional, which limits the ability to understand survivors' psychosocial functioning over time.

Research should continue to examine adjustment to help understand long-term psychosocial effects of retinoblastoma on survivors' functioning. The current sample included school-age survivors and previous research has examined outcomes of adult survivors of retinoblastoma.¹² In order to fully understand the impact, however significant, of this disease on survivors' functioning, it is critical to continue to study this population. It would be beneficial to better understand psychosocial functioning among adolescent survivors of retinoblastoma, as this is a crucial period in which perceived QoL may decline, and behavioral and emotional health symptoms may increase. Additionally, adolescence is a time period in which body image tends to become much more salient and dissatisfaction with body image often increases for both sexes.³⁰ Recent research also points to a global increase in adolescent mental health concerns, particularly within internalizing problems.³¹ As such, continuing to follow survivors of retinoblastoma through adolescence and early adulthood to monitor psychosocial outcomes and QoL is merited. Should such survivors continue to show resilience in these domains, perhaps research could seek to better understand factors contributing to such resilience.

Abbreviations Key

QoL	quality of life
HRQoL	Health-related quality of life

References

1. Dimeras H, Corson TW, Cobrinik D, et al. Retinoblastoma. *Nat Rev Dis Primers*. 2015; 1: 15021. doi:10.1038/nrdp.2015.21 [PubMed: 27189421]
2. Key statistics for retinoblastoma. American Cancer Society website. <https://www.cancer.org/cancer/retinoblastoma/about/key-statistics.html>. Accessed February 16, 2021
3. Abramson DH, Frank CM, Susman M, Whalen MP, Dunkel IJ, Boyd NW. Presenting signs of retinoblastoma. *J Pediatr*. 1998; 132(3): 505–508. [PubMed: 9544909]
4. Ford JS, Chou JF, Sklar CA, et al. Psychosocial outcomes in adult survivors of retinoblastoma. *J Clin Oncol*. 2015; 33(31): 3608–3614. doi:10.1200/JCO.2014.60.5733 [PubMed: 26417002]
5. Friedman DF, Chou JF, Francis JH, et al. Vision-targeted health-related quality of life in adult survivors of retinoblastoma. *JAMA Ophthalmol*. 2018; 136(6): 637–641. doi:10.1001/jamaophthalmol.2018.1082
6. vanDijk J, Imhof SM, Moll AC, et al. Quality of life of adult survivors of retinoblastoma in the Netherlands. *Health Qual Life Outcomes*. 2007; 5: 30. doi:10.1186/1477-7525-5-30 [PubMed: 17547767]
7. Hall LS, Ceisler E and Abramson DH. Visual outcomes in children with bilateral retinoblastoma. *J AAPOS*. 1999; 3(3): 138–142. doi:10.1016/S1091-8531(99)70058-3 [PubMed: 10428586]
8. Kelly KR, McKetton L, Schneider KA, Gallie BL, Steeves JKE. Altered anterior visual system development following early monocular enucleation. *Neuroimage Clin*. 2014; 4: 72–81. Doi:10.1016/j.nicl.2013.10.014 [PubMed: 24319655]
9. Kaste SC, Chen G, Fontanesi J, Crom DB, Pratt CB. Orbital development in long-term survivors of retinoblastoma. *J Clin Oncol*. 1997; 15(3): 1183–1189. [PubMed: 9060562]
10. Willard VW, Qaddoumi I, Pan H, Hsu C, Brennan RC, Wilson MW, Rodriguez-Galindo C, Goode K, Parris K, Phipps S. Cognitive and adaptive functioning in youth with retinoblastoma: A longitudinal investigation through 10 years of age. *J. Clin. Oncol* 2021; 39(24). doi: 10.1200/JCO.20.03422
11. Friedman DV, Chou JF, Oeffinger KC, et al. Chronic medical conditions in adult survivors of retinoblastoma: Results of the Retinoblastoma Survivor Study. *Cancer*. 2016; 122(5): 773–781. doi:10.1002/cncr.29704 [PubMed: 26755259]
12. Banerjee SC, Pottenger E, Petriccione M, et al. Impact of enucleation on adult retinoblastoma survivors' quality of life: A qualitative study of survivors' perspectives. *Palliat Support Care*. 2019; 18(3): 1–10. doi:10.1017/S1478951519000920
13. van Dijk J, Oostrom KJ, Huisman J, et al. Restrictions in daily life after retinoblastoma from the perspective of the survivors. *Pediatr Blood Cancer*. 2010; 54(1): 110–115. doi:10.1002/pbc.22230 [PubMed: 19760766]
14. Batra A, Kumari M, Paul R, Patekar M, Dhawan D, Bakhshi S. Quality of life assessment in retinoblastoma: A cross-sectional study of 122 survivors from India. *Pediatr Blood Cancer*. 2013; 63(2): 313–317. doi:10.1002/pbc25781
15. Zhang L, Gao T, Shen Y. Quality of life in children with retinoblastoma after enucleation in China. *Pediatr Blood Cancer*. 2018; 65(7): e27024. doi:10.1002/pbc27024 [PubMed: 29528176]
16. Batra A, Kain R, Kumari M, Paul R, Dhawan D, Bakhshi S. Parents' perspective of quality of life of retinoblastoma survivors. *Pediatr Blood Cancer*. 2016; 63(7): 1287–1289. doi:10.1002/pbc.25982 [PubMed: 27038275]
17. Weintraub N, Reshef N, Pe'er J, et al. The impact of monocular vision on motor function and quality of life in survivors of retinoblastoma. *Pediatr Blood Cancer*. 2018; 66(5): e27623. Doi:10.1002/pbc.27623
18. van Dijk J, Huisman J, Moll AC, et al. Health-related quality of life of child and adolescent retinoblastoma survivors in the Netherlands. 2007: Health Related QOL of child and adol retino survivors in Netherlands. *Health Qual Life Outcomes*. 2007; 5: 65. doi:10.1186/1477-7525-5-65 [PubMed: 18053178]
19. Kessler RC, Amminger P, Aguilar-Gaxiola S, Alonso J, Lee S, Ustun TB. Age of onset of mental disorders: A review of recent literature. *Curr Opin Psychiatry* 2007; 20(4): 359–364. [PubMed: 17551351]

20. Varni JW, Seid M, Knight TS, Uzark K, Szer IS. The PedsQL Generic Core Scales: Sensitivity, responsiveness, and impact on clinical decision-making. *J Behav Med.* 2002; 25: 175–193. [PubMed: 11977437]
21. Varni JW, Seid M, Kurtin PS. The PedsQL 4.0: Reliability and validity of the Pediatric Quality of Life Inventory 4.0 Generic Core Scales in healthy and patient populations. *Med Care.* 2001; 39:800–812. [PubMed: 11468499]
22. Forrest CB, Bevans KB, Tucker C, et al. Commentary: The patient-reported outcome measurement information system (PROMIS[®]) for children and youth: application to pediatric psychology. *J Pediatr Psychol.* 2012; 37:614–621. [PubMed: 22362923]
23. Cella D, Yount S, Rothrock N, et al. The Patient-Reported Outcomes Measurement Information System (PROMIS): Progress on an NIH Roadmap cooperative group during its first two years. *Med Care.* 2007; 45:S3–S11.
24. Carle AC, Bevans DB, Tucker CA, Forrest CB. Using nationally representative percentiles to interpret PROMIS pediatric measures. *Qual Life Res.* 2021; 30:997–1004. [PubMed: 33201388]
25. Reynolds CR, Kamphaus RW. *Behavior Assessment Scale for Children, Second Edition.* Circle Pines, MN: AGS; 2004.
26. Castellano-Tejedor C, Pérez-Campedarós M, Capdevila L, Blasco-Blasco T. Surviving cancer: The psychosocial outcomes of childhood cancer survivors and its correlates. *J. Health Psychol.* 2016; 21(7): 1491–1502. [PubMed: 25411198]
27. Ashford JM, Netson KL, Clark KN, Merchant TE, Santana VM, Wu S, Conklin HM. Adaptive functioning of childhood brain tumor survivors following conformal radiation therapy. *J Neurooncol.* 2014; 118(1):193–199. [PubMed: 24658934]
28. Allais FB. Assessing the gender gap: Evidence from SIMPOC surveys. Geneva: International Labour Organization; 2009. Retrieved from http://www.ilo.org/ipecinfo/product/download.do?sessionId=F9hFsM_emBhRZtaAN5kraO-TeHGgq-8W8LDTM9T8ktXaKaWVnzhL!866796678?type=document&id=10952
29. Schulte F, Wurz A, Reynolds K, Strother D, Dewey D. Quality of life in survivors of pediatric cancer and their siblings: The consensus between parent-proxy and self-reports. *Pediatr Blood Cancer.* 2016; 63(4): 677–683. [PubMed: 26739262]
30. Hargreaves DA, Tiggemann M. Idealized media images and adolescent body image: “Comparing” boys and girls. *Body Image.* 2004; 1(4): 351–361. Doi: 10.1016/j.bodyim.2004.10.002 [PubMed: 18089166]
31. Blomqvist I, Blom EH, Hagglof B, Hammarstrom A. Increase of internalized mental health symptoms among adolescents during the last three decades. *Eur J Public Health.* 2019; 29(5): 925–931. doi: 10.1093/eurpub/ckz028 [PubMed: 30859217]

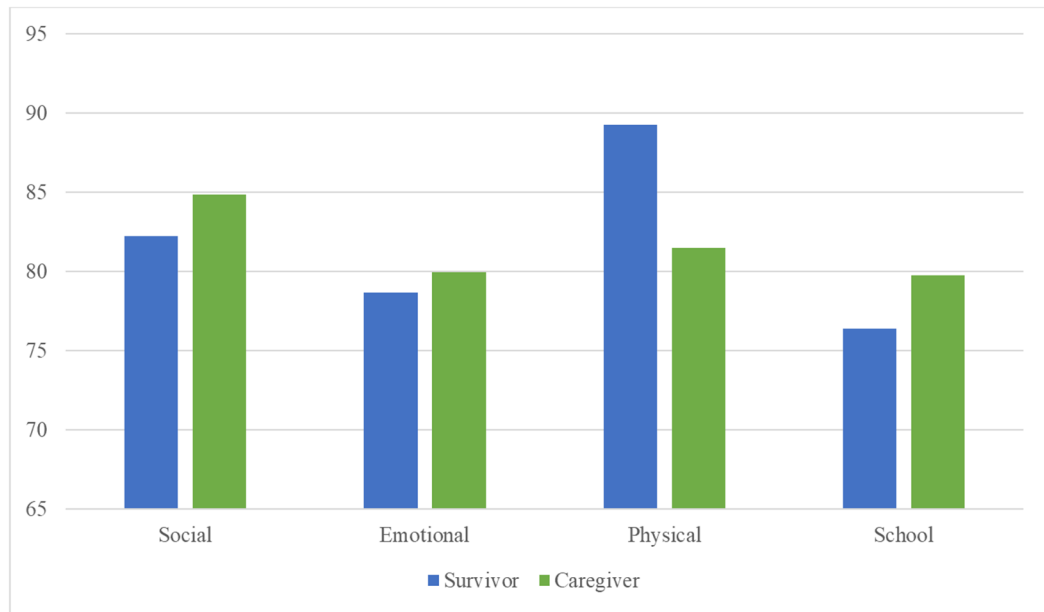


Figure 1. Mean survivor and caregiver scores across domains of QoL

Note. Survivors reported significantly greater physical QoL than caregivers. There were no significant differences between survivor- and caregiver-reported social, emotional, or school QoL.

Table 1.

Demographic and medical characteristics

	Range (M, SD) / N(%)
Age (years)	8.67 – 13.00 (10.89, 1.07)
Gender	
Male	34 (49.3)
Female	35 (50.7)
Race/Ethnicity	
White/Caucasian	43 (62.3)
African American	16 (23.2)
Multiracial	4 (5.8)
Other	5 (7.2)
Diagnosis	
Bilateral	30 (43.5)
Unilateral	39 (56.5)
Age at diagnosis (years)	0.3 – 5.94 (1.51, 1.37)
Time off treatment (years)	6.5 – 12.1 (9.37, 1.15)
Enucleation Status	
Yes	49 (71.0)
No	20 (29.0)

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

Table 2.

Summary of QoL scores

	Social	Emotional	School	Physical
<i>Respondent</i>				
Survivor	82.22 (21.44)	78.67 (18.51)	76.36 (19.46)	89.23 (13.38)
Caregiver	84.85 (18.63)	79.93 (18.87)	79.76 (17.78)	81.49 (22.42)
Survivor Report	Social	Emotional	School	Physical
<i>Laterality</i>				
Unilateral	83.30 (21.27)	77.24 (20.12)	73.95 (21.47)	87.18 (14.55)
Bilateral	80.71 (21.97)	80.54 (16.24)	79.46 (16.15)	92.08 (11.20)
<i>Enucleation Status</i>				
Yes	75.50 (23.39)	81.30 (18.27)	77.83 (20.62)	90.23 (13.14)
No	85.08 (20.14)	72.50 (18.03)	73.00 (16.50)	86.88 (14.00)
<i>Sex</i>				
Male	82.90 (20.58)	81.91 (21.10)	78.10 (16.42)	87.41 (14.84)
Female	81.52 (22.59)	75.17 (14.89)	74.53 (22.37)	91.10 (12.08)
Caregiver Report	Social	Emotional	School	Physical
<i>Laterality</i>				
Unilateral	86.28 (18.16)	80.00 (19.47)	79.84 (16.89)	73.34 (25.34)
Bilateral	82.93 (19.39)	79.83 (18.61)	79.66 (19.22)	87.07 (16.57)
<i>Enucleation Status</i>				
Yes	86.25 (17.52)	82.12 (19.32)	80.70 (17.21)	82.37 (22.34)
No	81.50 (21.16)	74.69 (17.45)	77.50 (19.36)	79.38 (23.28)
<i>Sex</i>				
Male	87.12 (17.28)	83.18 (18.87)	80.15 (17.96)	79.27 (26.16)
Female	82.71 (19.83)	76.86 (18.81)	79.39 (17.86)	83.57 (18.35)

All scores are listed as M (SD)

Table 3.Comparison of QoL scores by respondent to healthy comparison norms²¹

	M (SD)	Healthy Comparison M(SD)	One- sample t	P
<i>Survivor Report</i>				
Social	82.22 (21.44)	87.42 (17.18)	-1.99	0.051
Emotional	78.67 (18.51)	80.86 (19.64)	-.98	0.33
School	76.36 (19.46)	78.63 (20.53)	-.95	0.35
Physical	89.23 (13.38)	84.41 (17.26)	2.95	<.01
<i>Caregiver Report</i>				
Social	84.85 (18.63)	91.56 (14.20)	-2.97	<0.01
Emotional	79.93 (18.87)	82.64 (17.54)	-1.18	.242
School	79.76 (17.78)	85.47 (17.61)	-2.65	<.05
Physical	81.49 (22.42)	89.32 (16.35)	-2.88	<0.01

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

Table 4.Comparison of PROMIS scores to nationally representative median scores²⁴

	M (SD)	Median	One-sample t	P
<i>Domain</i>				
Anxiety	46.46 (11.82)	42.0	3.07	<0.01
Depression	43.13 (9.21)	45.0	-1.61	0.11
Peer Relations	47.71 (11.72)	48.0	-.201	0.84

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

Table 5.

Summary of survivor- and caregiver-reported behavioral and emotional health scores

	Survivor Report				Caregiver Report		
	Anxiety	Depression	Peer Relations	Internalizing	Externalizing	Behavioral Symptoms	Adaptive Skills
<i>Laterality</i>							
Unilateral	46.58 (12.87)	42.59 (9.00)	50.09 (10.15)	50.24 (10.93)	46.84 (9.39)	48.11 (9.35)	51.11 (9.56)
Bilateral	46.30 (10.45)	43.90 (9.64)	44.64 (13.02)	51.41 (13.40)	46.79 (9.29)	49.79 (10.14)	49.48 (9.84)
<i>Enucleation Status</i>							
Yes	46.04 (12.65)	42.67 (9.71)	49.56 (10.27)	49.13 (11.40)	47.19 (9.58)	48.46 (10.17)	50.46 (10.03)
No	47.50 (9.68)	43.39 (7.85)	43.33 (13.92)	54.84 (11.76)	45.89 (8.62)	49.79 (8.42)	50.26 (8.82)
<i>Sex</i>							
Male	44.08 (12.84)	41.35 (9.69)	47.39 (13.08)	47.88 (10.55)	45.71 (9.80)	47.12 (11.06)	58.88 (10.31)
Female	48.99 (10.22)	44.85 (8.52)	48.03 (10.38)	53.70 (12.56)	47.97 (8.71)	50.61 (7.75)	47.85 (8.30)

All scores are listed as M (SD)