Neurofibromatosis type 1 (NF1) is an extremely variable, progressive neurocutaneous disorder that affects dermatological, skeletal, ophthalmological, and neurological functioning. The diagnosis of NF1 is based on clinical features including multiple café au lait spots, plexiform neurofibromas, Lisch nodules, and freckling in the inguinal and axillary areas. NF1 is a common autosomal dominant genetic disorder affecting approximately 1 in 3000 individuals with no predilection to race or gender. The natural history of this condition has been well-studied, but few studies have focused on the effects of NF1 on the quality of life (QOL) in individuals with this condition, with no studies specifically looking at children and adolescents with NF1. This study was undertaken to determine the overall QOL in children and adolescents with NF1. A total of 64 children between the ages of 5 and 18 years from 53 families with their respective parents took part in this study. Children with NF1 and their parents reported a total QOL score that was significantly lower than healthy controls, with parent’s perceiving QOL much lower than children. Parents of children with NF1 reported lower activities, fewer social interactions with peers, more problems in school, and more behavioral problems in children with NF1 than expected from normed data. These findings indicate that children with NF1 have a lower overall QOL due to multiple contributing problems. However, when determining the effect of various predictor variables on the overall QOL, severity and the number of friends were the only two predictors that were significantly associated with QOL, activities, school, and social functioning. SES was inversely related to depression. The results of this study indicate that children and adolescents with NF1 suffer from a lower QOL due to the disease process as a whole, with disease severity and the number of friends playing a role as major QOL predictors. While genetic counseling should be provided to all individuals with NF1, special attention should be focused on those with increased disease severity and fewer friends. This process will potentially help to increase quality of life and patient confidence, and decrease future psychological and psychosocial manifestations associated with NF1.