

# QUALITY OF LIFE AND PSYCHOLOGICAL ADJUSTMENT IN CHILDREN AND ADOLESCENTS WITH NEUROFIBROMATOSIS TYPE 1

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**Objective** To assess quality of life (QoL) and psychological adjustment in children and adolescents with neurofibromatosis type 1 (NF1).

**Study design** Forty-six patients with NF1 were investigated between the ages of 7 and 16 years (mean, 11.6 years), with children and parents used as informants. TNO-AZL Questionnaire for Children's Health-Related Quality of Life and Child Behavior Checklist scores were compared with healthy reference groups. Predictive values of sociodemographic variables, illness-related variables, and family-related variables for quality of life and psychological adjustment were assessed.

**Results** Most dimensions of QoL in NF1 children and adolescents were different from reference values. Deviations in the NF1 group were an impairment of motor, cognitive, and social functioning and a reduction of positive and negative emotions. Also, psychological adjustment in patients with NF1 was significantly impaired compared with normal subjects. Illness-related variables had a negative impact on the emotional domain of QoL. Good family relationships positively affected both QoL and psychological adjustment.

**Conclusions** QoL and psychological adjustment are impaired in children and adolescents with NF1. Illness-related variables and the quality of family relationships are important predictors. (*J Pediatr* 2006;149:348-53)

**N**eurofibromatosis type 1 (NF1) is a genetic disease of the central and peripheral nervous system caused by a mutation in a gene on chromosome 17. It is inherited in an autosomal dominant mode, with a high rate of new mutations and a prevalence of 1 in 3000 to 4000 individuals.<sup>1</sup> The symptoms of NF1 are progressive, unpredictable, and highly variable, ranging from differences undetectable by the untrained observer through mild esthetic disfigurement to life-threatening conditions. Defining features include café-au-lait spots, axillary freckling, Lisch nodules, cutaneous and plexiform neurofibromas, optic gliomas, pseudoarthrosis, and scoliosis or other skeletal abnormalities.<sup>2</sup> Complications can occur, including esthetic disfigurement, skeletal abnormalities, and a variety of benign and malignant tumors of the central nervous system.<sup>1</sup> The most common neurologic complications of NF1 in children and adolescents are cognitive impairments.<sup>3</sup> The diagnosis of attention deficit-hyperactivity disorder is common in children with NF1.<sup>4</sup>

The esthetic disfigurement and the physical and cognitive problems commonly associated with NF1 suggest a negative impact on the quality of life (QoL) and psychological adjustment of children and adolescents. One prior study confirms this suggestion for affected adults.<sup>5</sup> Little research, though, has focused on the psychological and social aspects of NF1 in children and adolescents. Although the self-concept of children and adolescents with NF1 was shown to be normal,<sup>6</sup> behavioral and social problems were reported.<sup>7</sup> Compared with unaffected siblings, children with NF1 showed more overall problem behaviors and internalizing and externalizing behavioral problems<sup>7</sup> as well as poorer social skills.<sup>8</sup> In a recent study assessing personality profiles, children and adolescents with NF1 are described as less conscientious, less emotionally stable, less open for new experience, less active, more extravert, more dependent, and more irritable.<sup>9</sup> Also, in comparison with normative data, a higher occurrence of sleep and behavioral problems is reported.<sup>10</sup> Although there is some knowledge about psychological adjustment of individuals with NF1, studies assessing QoL in children with NF1 are lacking.

The aim of this study was a comprehensive, standardized evaluation of QoL and psychological adjustment in children and adolescents with NF1 in comparison to healthy

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CBCL	Child Behavior Checklist	QoL	Quality of life
FRI	Family Relationship Index	TACQOL	TNO-AZL, Questionnaire for Children's Health-Related Quality of Life
NF1	Neurofibromatosis type 1		

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children. Based on clinical anecdotal evidence, we hypothesized that children and adolescents with NF1 would generally show a reduced health-related QoL as well as more psychological adjustment problems compared with healthy children.

## METHODS

### Subjects

Children and adolescents with NF1 meeting the diagnostic criteria of the National Institutes of Health Consensus Conference,<sup>2</sup> previously seen at the University Children's Hospital in Zurich or at the Children's Hospital in Aarau, and their parents were asked to participate in the study. For methodological reasons, the required age range at assessment was 7 to 16 years, and command of the German language was a prerequisite. Sixty-four children and adolescents met inclusion criteria. One patient with additional trisomy 21 was excluded from the study, and 17 patients refused to participate. The final sample comprised 46 participants (response rate, 72%).

### Measures

**TNO-AZL CHILD QUALITY OF LIFE QUESTIONNAIRE TACQOL<sup>11</sup>:** The TACQOL is a generic instrument designed for QoL assessment in medical research and clinical trials. As a multidimensional instrument, it can be compared with questionnaires such as the PedsQL,<sup>12</sup> which is widely used for health-related QoL studies in children and adolescents in North America. The TACQOL is available in a child form (CF) and a parent form (PF), both containing five health status scales of eight items: physical complaints, basic motor functioning, autonomy, cognitive, social, positive emotional, and negative emotional functioning. Children and parents are asked to note problems in any of the mentioned domains and whether the child seems to be bothered by these problems. Maximum domain scores are 32 for the first five scales and 16 for the emotional scales. Higher scores represent a better QoL. Normal values for the child and parent forms are provided from a community sample of healthy Dutch children (child form: 1048 children/parent form: 1618 children).<sup>11</sup> Internal and external validity of the TACQOL has been confirmed in previous studies with healthy and clinical samples.<sup>11</sup>

**CHILD BEHAVIOR CHECKLIST CBCL<sup>13,14</sup>:** The Child Behavior Checklist (CBCL) is a standardized measure with excellent psychometric properties providing parental reports of a child's behavior. It consists of 120 items assessing internalizing (social withdrawal, somatic complaints, anxiety/depression) and externalizing behavioral problems (dissocial and aggressive behavior). From these problem scales, an overall Total Behavioral Problems Score is calculated and compared with normative data (T-score). Reference values are provided by 2856 healthy German children and adolescents ages 4 to 18 years.<sup>14</sup>

**FAMILY RELATIONSHIP INDEX FRI<sup>15</sup>:** The FRI is a 27-item questionnaire consisting of three subscales of the Family Environment Scale, assessing expressiveness, cohesion, and conflict within a family. The FRI index is calculated as the sum of the subscales expressiveness and cohesion minus the subscale conflict. Higher scores indicated better family relationships. Reliability and validity of this scale have been confirmed.<sup>15</sup>

**ILLNESS-RELATED VARIABLES:** Illness-related variables such as family history of NF1 were obtained through review of the medical records for each child. The severity of NF1 was rated by using a modified version<sup>6</sup> of Riccardi's severity scale,<sup>16</sup> which has been widely used in previous studies.<sup>5,6,8</sup> The modified severity scale excludes cognitive features of NF1 and is based only on physical features. Four degrees of severity were distinguished: Minimal NF1 includes the presence of few features with no compromise of health or well-being. Mild NF1 means that there are enough features present to make the disease obvious and a source of concern, but without significant compromise of health. Moderate NF1 reflects the presence of significant compromise of health and well-being, though the compromise can be reasonably well managed. Severe NF1 indicates a serious compromise of health that is managed with difficulty, intractable, or associated with a shortened life span. The physical features of the modified severity scale were rated by the parents and confirmed with information from each child's medical record.

The visibility of NF1 was scored using a scale developed by Ablon<sup>17</sup> that has been used in previous studies.<sup>5</sup> Ratings are based on the appearance of a fully dressed person. Three different degrees of visibility were distinguished: Mild, moderate, and strong visibility. Ratings are based on cutaneous signs and features associated with gait and posture as well as ocular movements and orbital symmetry. Final scoring of severity and visibility was made by the first author, under the supervision of a neurologist with extensive experience in NF1.

**SOCIOECONOMIC STATUS:** Socioeconomic status (SES) was calculated by means of a six-point scale of both paternal occupation and maternal education. The three social classes were defined as follows: SES scores 2 to 5, lower class; SES scores 6 to 8, middle class; and SES scores 9 to 12, upper class. This measure has been used in previous studies and has been shown to be a reliable and valid indicator of SES in our community.<sup>18</sup>

### Procedure

The study was approved by the local research ethics committee. Parents were informed about the study by letter. Children and adolescents were assessed by means of a standardized interview. The interviews lasted between 30 and 60 minutes. Most interviews were conducted in the children's homes. Eight were done at the hospital. Parents were assessed at the same time point with questionnaires.

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