The Heart Surgery Forum #2011-1126
15 (2), 2012 [Epub April 2012
doi: 10.1532/HSF98.20111126

Online address: http://cardenjennings.metapress.com

Quality of Life after Children Undergo a Radical Arterial Switch Operation at an Older Age

Jing-bin Huang, MD,[1] Yong-tao Cao[2], Jian Liang, MD,[1] Xiao-dong Lv, MD[3]

[1]Department of Cardiothoracic Surgery, Affiliated Ruikang Hospital of Guangxi Traditional Chinese Medical College, Nanning, Guangxi Zhuang Autonomous Region; [2]Department of Statistics, University of Wyoming, Laramie, Wyoming; [3]Pediatric Center of Cardiac Surgery, Cardiovascular Institute and Fuwai Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China

ABSTRACT

Objective: The goal of the study was to evaluate the quality of life of children after the older corrective arterial switch operation (ASO) by means of the Pediatric Quality of Life Inventory (PedsQL), version 4.0.

Methods: The records of 86 patients who had complete transposition of the great arteries plus a nonrestrictive ventricular septal defect, or a Taussig-Bing anomaly, and severe pulmonary arterial hypertension, and who underwent a corrective ASO at an older age (>6 months) between May 2000 and October 2008 were reviewed retrospectively. Eighty survivors were followed up, and the health-related quality of life of the survivors was evaluated with the PedsQL, version 4.0.

Results: There were 6 hospital deaths. The mean (SD) follow-up interval was 3.5 ± 2.3 years, and the mean age at last visit was 7.0 ± 1.2 years. Two late deaths occurred, and 8 patients were lost to follow-up. Patients who underwent a corrective ASO at an older age showed acceptable scores for all scales, and they were all comparable with those of a healthy population.

Conclusions: Our data suggest that the quality of life of children who undergo a corrective ASO at an older age (>6 months) is acceptable, compared with that of healthy children in China.

INTRODUCTION

The aim of surgical therapy has changed from providing only therapy for survival toward offering corrections that allow an almost normal life in terms of expectancy and quality. It is apparent that assessments of mortality and morbidity are too narrow to analyze the benefits of various therapies. The cardiac lesion itself is not always the major problem for these patients, because issues pertaining to quality of life and psychosocial aspects often predominate. Therefore, the interest in the quality of life of patients after repair is increasing [Loup 2009]. Global quality-of-life instruments have recently been developed for use in children. The Pediatric Quality of Life Inventory (PedsQL) is a modular instrument for measuring health-related quality of life dimensions in children and adolescents 2 to 18 years of age. The PedsQL version 4.0 Generic Core Scales are multidimensional (physical, emotional, social, school role functioning) child self-report and parent proxy-report scales developed as a generic measure to be integrated with the PedsQL disease-specific modules [Varni 2001, 2002; Uzark 2003]. The reliability and validity of the PedsQL Generic Core Scales have been demonstrated in healthy and patient populations [Varni 2001, 2002; Uzark 2003]. Previous studies used the Child Health Questionnaire to assess quality of life in children who survive a neonatal arterial switch operation (ASO) [Dunbar-Masterson 2001; Culbert 2003]. Most children with complete transposition of the great arteries or a Taussig-Bing anomaly have corrective surgery at as early an age as possible; however, medical, social, or economic reasons (particularly in developing countries) sometimes cause a delay in the timing of surgery. This study aimed to evaluate the quality of life of children after older (>6 months) corrective (not palliative) ASO by using the PedsQL 4.0 Generic Core Scales.

METHODS

Study Participants

All patients (n = 86) with complete transposition of the great arteries and a nonrestrictive ventricular septal defect (or a Taussig-Bing anomaly) plus severe pulmonary arterial hypertension (PAH) and who underwent corrective ASO at older age (>6 months) were enrolled at our institution between May 2000 and October 2008. The records of 86 patients included in the study were reviewed retrospectively. Severe PAH was defined as mean pulmonary artery pressure >50 mm Hg or a systolic pulmonary/systemic pressure ratio >0.8, as obtained from cardiac catheterization data of patients under general anesthesia. Eligibility criteria included (1) a diagnosis of complete transposition of the great arteries with a nonrestrictive ventricular septal defect, (2) operation at an older age (>6 months), (3) severe PAH (mean pulmonary artery pressure >50 mm Hg or a systolic pulmonary/systemic pressure ratio >0.8), and (4) an age between 2 and 18 years when the

Received August 28, 2011; accepted October 18, 2011.

Jing-bin Huang, Yong-tao Cao, and Jian Liang contributed equally to this article.

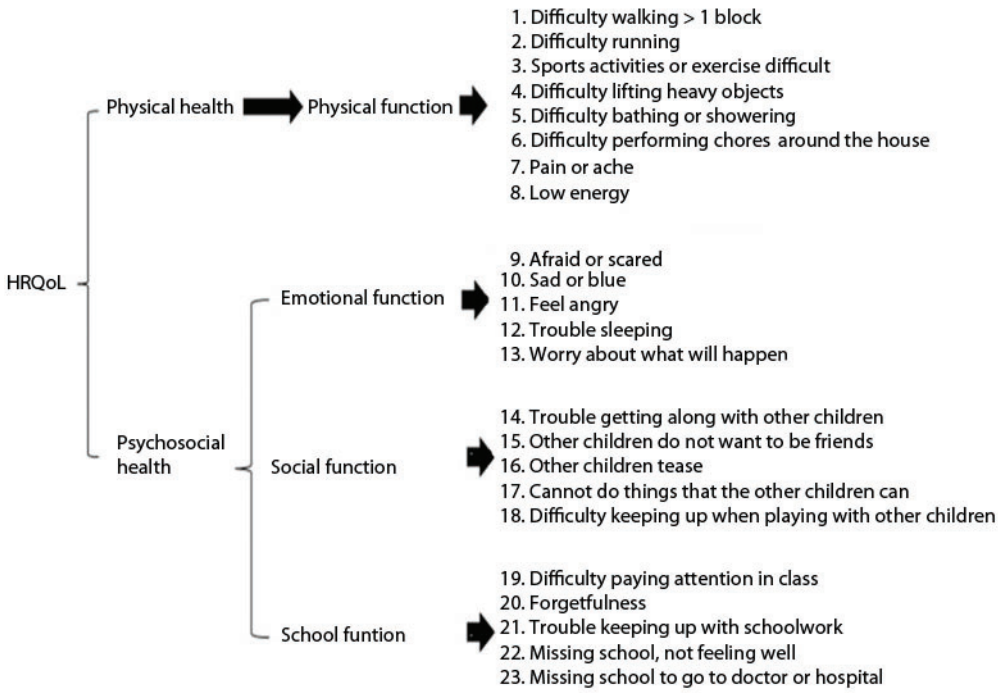
Correspondence: Jing-bin Huang, Department of Cardiothoracic Surgery, Affiliated Ruikang Hospital of Guangxi Traditional Chinese Medical College, Nanning 530011, Guangxi Zhuang Autonomous Region, China; 86-10-88398188; fax: 86-10-68332747 (e-mail: gz98039@hotmail.com).

PedsQL 4.0 Generic Core Scales evaluations were completed. Exclusion criteria included (1) an age at operation of less than 6 months, (2) trisomy 21, (3) associated extracardiac anomalies of more than minor severity, and (4) an age not in the range of 2 to 18 years when the PedsQL 4.0 Generic Core Scales evaluations were completed. Preoperative cardiac catheterization data were available for all patients (n = 86). Cardiac catheterization was conducted with the patient under general anesthesia. The preoperative pulmonary artery pressure and pulmonary vascular resistance were measured by the conventional cardiac catheterization protocol and the Fick method. The diagnosis was made on the basis of echocardiographic and cardiac angiographic findings and was confirmed during the operation.

Our study was approved by the Medical Ethics Committee of Fuwai Cardiovascular Disease Hospital, and the committee granted approval to waive the need for patient consent for publishing follow-up data about these patients.

Determining whether the Children Were Still Eligible for the Operation

The inhaled-oxygen test was completed in 48 of the patients. An inhaled-oxygen test result was deemed positive when the mean pulmonary arterial pressure deceased 20% when 100% oxygen was inhaled for 20 to 30 minutes. Inhaled-oxygen test results were positive for 15 patients (15/48, 31%) and negative for 33 patients (33/48, 69%). From January 2006 to October 2008, a strategy of diagnostic treatment and repair was used in 50 consecutive patients to evaluate whether PAH was reversible or not. All patients received advanced PAH treatment (10-20 ppm continuously inhaled nitrous oxide, 2-5 mg sildenafil 3 times per day, and 31.5 mg bosentan twice daily), which lasted a mean (SD) of 13 ≥ 5 days (range, 5-21 days). In our study, patients with complete transposition of the great arteries and a nonrestrictive ventricular septal defect (a Taussig-Bing anomaly) underwent a strategy of diagnostic treatment and repair. Patients who experienced an increase in the transcutaneous oxygen saturation of at least 5% (mean, 11% ± 6%) were deemed to have had an adequate response and then underwent corrective repair. Four patients (4/50, 8%) were excluded from the operation because the PAH was not shown to be reversible, and 46 patients underwent corrective repair. To examine the nature of the pulmonary arterial vessels in the patients selected to receive the diagnostic-treatment-and-repair protocol, we obtained lung specimens via open lung biopsy during the cardiac operation. The nature of the pulmonary arterial vessel was determined according to the Heath and Edwards classification. Seven of 11 patients showed a grade I change, 2 of 11 patients showed a grade II change, 1 of 11 patients showed a grade III change, and only 1 of 11 patients showed a grade IV change with a plexiform lesion. The measurements of postoperative pulmonary artery pressure were taken in the operation room at the end of the case. All survivors discharged from the hospital underwent chest radiography, electrocardiography, and echocardiography examinations. The patients were followed up in our outpatient clinic once every 3 months. The mean pulmonary arterial pressure was estimated by echocardiography.



The Pediatric Quality of Life Inventory (PedsQL) 4.0 Generic Core Scales. HRQoL indicates health-related quality of life.

Measurements

Health-related quality of life was measured with the PedsQL 4.0 Generic Core Scales. The proxy-report version of PedsQL 4.0 Generic Core Scales (Chinese version, limited agreement for use; Mapi Research Trust, Lyon, France) was sent to the parent or guardian, who completed the questionnaires. If no response was received within 6 weeks of the initial mailing, a reminder was sent, followed by 2 attempts to complete the follow-up by telephone. The completed PedsQL 4.0 Generic Core Scales were sent back to us. Quality of life for a control group of healthy children in China was also measured. Reference values for healthy children were taken from the publication in Chinese [Lu 2008].

PedsQL 4.0 Generic Core Scales

The PedsQL 4.0 Generic Core Scales consists of a 23-item generic core that measures children's health in the following 4 domains: (1) physical functioning (8 items); (2) emotional functioning (5 items); (3) social functioning (5 items); and (4) school functioning (5 items). The individual scales can be combined to yield 3 summary measures of physical (the same as the physical-functioning scale), psychosocial (emotional, social, and school-functioning scales), and total health (all 4 scales) (Figure). A growing number of studies have shown the reliability and validity of the PedsQL 4.0 as a measure of general physical and psychosocial health of children. Items are reverse-scored and linearly transformed to a scale of 0 to 100, so that higher scores indicate a better health-related quality of life. A physical health summary score (8 items) is the same as the physical-functioning subscale. To create a psychosocial health summary score, the mean is computed as the sum of the items divided by the number of items in the emotional, social, and school-functioning scales [Varni 2001, 2002; Uzark 2003]. PedsQL 4.0 mean scores of children who underwent corrective ASO at an older age (>6 months) were compared with those of the control group (healthy children in China, n = 291) [Lu 2008].

Table 1. Patient Characteristics*

Preoperative characteristics	
Female/male sex, n	23/47
Age at operation, y	2.0 ± 1.8
Age at last visit, y	7.0 ± 1.2
Weight, kg	9.0 ± 7.0
Preoperative mPAP, mm Hg	64.9 ± 13.0
PVR, dyn·s·cm^{-5}	324 ± 249
NYHA class	
II	80
III	6
Surgical data	
CPB time, min	233 ± 57
Aortic cross-clamp time, min	160 ± 40
Postoperative mPAP, mm Hg	30.5 ± 13.0

*Data are presented as the mean ± SD as indicated. mPAP indicates mean pulmonary artery pressure; PVR, pulmonary vascular resistance; NYHA, New York Heart Association; CPB, cardiopulmonary bypass.

Data Analysis

Data are presented as frequencies, medians with ranges, or means with SDs. Statistical analysis was performed with SPSS software (version 13.0; SPSS, Chicago, IL, USA). Analysis of continuous variables was performed with the Mann-Whitney test. A P value <.05 was considered statistically significant.

RESULTS

Surgery was performed with all patients on cardiopulmonary bypass at a low flow (50 mL·kg^{-1}·min^{-1}) and a rectal temperature of 18°C to 22°C. Of the 86 children who were enrolled in the study, 8 died; 78 were eligible for participation in the study. Of these patients, the families of 70 patients (90%) completed the PedsQL 4.0 Generic Core Scales. Eight patients were lost to follow-up. The characteristics of the 70 study individuals are summarized in Table 1.

The mean follow-up interval was 3.5 ± 2.3 years; the mean age at last visit was 7.0 ± 1.2 years. The 70 study participants survived with a significant regression of the pulmonary artery pressure from a mean preoperative value of 64.9 ± 13.0 mm Hg to a mean postoperative value of 30.5 ± 13.0 mm Hg (P < .001). The postoperative incidence of PAH was 42% (36 of 86 patients), the incidence of postoperative severe PAH was 6% (5 of 86 patients), and the incidence of residual severe PAH was 5% (4 of 80 patients). The latest follow-up data when the PedsQL 4.0 Generic Core Scales were completed showed that 4% (3/70) of the patients were in New York Heart Association (NYHA) class II and were receiving oral bosentan. Ninety-six percent of the patients (67/70) were in NYHA class I. There were no reoperations.

Follow-up data showed that the patients subjected to corrective ASO at an older age had significantly lower PedsQL 4.0 scores than healthy Chinese children (P < .05) for all scales

Table 2. Mean Pediatric Quality of Life Inventory (PedsQL) 4.0 Scores for Survivors Subjected to Corrective Arterial Switch Operation (ASO), Compared with Those of Healthy Children*

Scale	ASO Group (n = 70)	Reference Group (n = 291)	P
Total score	81.4 ± 13.6	84.9 ± 10.9	<.05
Physical health	85.7 ± 19.6	88.3 ± 11.4	<.05
Psychosocial health	80.1 ± 15.8	83.0 ± 12.7	<.05
Emotional functioning	76.3 ± 18.6	79.4 ± 17.7	<.05
Social functioning	81.1 ± 17.8	86.9 ± 15.3	<.05
School functioning	80.2 ± 19.7	82.7 ± 14.6	<.05

*Data are presented as the mean ± SD. Psychosocial health includes emotional functioning, social functioning, and school functioning.

(Table 2). In our study, only 2 patients had delayed neuropsychological development, and 97.2% (68/70) performed normally in school functioning. No correlation was found in the 70 patients between the age at operation and physical health ($P = .13$), psychosocial health ($P = .18$), emotional functioning ($P = .33$), social functioning ($P = .26$), or school functioning ($P = .46$).

DISCUSSION

The World Health Organization defines health as "a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity" [WHO 1948; Guyatt 1997]. The aim of surgical therapy has changed from providing only therapy for survival toward offering corrections that allow an almost normal life in terms of expectancy and quality. It is apparent that assessments of mortality and morbidity are too narrow to analyze the benefits of therapies; thus, additional outcomes of care are also important [Loup 2009]. Surgical advances have allowed increased longevity for many children with complex heart defects that were previously considered inoperable. Quality of life has emerged as an important outcome for these children. Therefore, the interest in the quality of life of patients after repair is increasing. The acceptable reliability and validity of the PedsQL 4.0 as a measure of general physical and psychosocial health of children has been demonstrated in a growing number of studies [Varni 2001, 2002; Uzark 2003]. Our study indicated that patients who had dextro-transposition of the great arteries and a nonrestrictive ventricular septal defect (or Taussig-Bing anomaly) and severe PAH, and who underwent a corrective ASO at an older age (>6 months) still showed acceptable scores measured by PedsQL 4.0 in all scales, including physical health, psychosocial health, emotional functioning, social functioning, and school functioning, all of which were comparable to those of the healthy population. Ninety-seven percent (68/70) of the patients were in NYHA class I. The health-related quality of life of this selected subset of patients who underwent a corrective ASO at an older age is comparable to that of the control group of healthy Chinese children.

Previous reports have shown that 96% of children are without symptoms or limitation of physical activity at up to 5 years after they undergo ASO during infancy [Hövels-Gürich 1997; Losay 2001]. Dunbar-Masterson et al [2001] used the Child Health Questionnaire to evaluate the general health status of 160 children at 8 years of age. They found that their overall physical and psychosocial health status after undergoing ASO during infancy was similar to that of a reference healthy population. These outcomes are reassuring in regard to the findings of our study, despite the fact that the patients in our study underwent their ASO at an older age (>6 months). Interestingly and amazingly, Culbert et al reported that the Child Health Questionnaire scores of children with dextro-transposition of the great arteries were significantly higher than those for the published reference population (indicating a better self-perceived health status) in all categories except self-esteem [Dunbar-Masterson 2001]. Loup et al [2009] also found that with regard to the overall quality of life, a group of adult patients (mean, 23 7 years) after ASO had similar or even higher scores in all SF-36 (36-item Short Form Health Survey Questionnaire) and HADS (Hospital Anxiety and Depression Scale) health dimensions, compared with a reference population matched by age and sex [Loup 2009].

Frequently, it is assumed that patients with congenital heart disease have a reduced quality of life. Surprisingly, the overall quality of life of the patients with congenital heart disease was acceptable, and sometimes was even better than that of the age- and sex-matched reference population. Given the assumption that these patients are exposed to an unusual level of anxiety about their future, their prognosis, and the possibility that their condition could get worse, these results are unexpected. Clearly, self-perception of disease is multifactorial. Quality of life depends on a variety of factors, such as the type and severity of the congenital heart defect; the attitude and support of family, friends, society, and the medical care team; the home and school environment; economic status; genetic or geographic factors; and so on. Another explanation is that the likelihood that these patients might feel grateful and lucky to be alive would certainly have a positive influence on their psychological attitude [Cella 1994; Kamphuis 2002; Hendry 2004; Immer 2005; Plummer 2009].

In China, because the prevention system for congenital heart disease has not been fully established, techniques of treating complex congenital heart disease have not been popular nationwide, and the documentation of treatment of people with congenital heart disease was not adequate, corrective surgery was delayed. Residual PAH after discharge affects the quality of life of children after a corrective ASO at an older age (>6 months); thus, regression of PAH is important [Nakjima 1996; Shafazand 2004; Cenedese 2006]. In our study, the 70 study patients survived with a significant regression of the pulmonary artery pressure from a mean preoperative value of 64.9 ± 13.0 mm Hg to a postoperative value of 30.5 ± 13.0 mm Hg ($P < .001$). Two patients with moderate residual PAH and in NYHA class II were receiving oral bosentan treatment at the last visit. Ninety-seven percent (68/70 patients) were in NYHA class I and free of medication. Recently, advanced therapies for PAH have become available and have been effective in reducing pulmonary vascular resistance and symptoms in patients with a near-systemic pulmonary arterial pressure, and orally administered endothelin antagonists (bosentan) and the application of sildenafil allow long-term administration [Lunze 2006; Apostolopoulou 2007].

Dunbar-Masterson et al [2001] reported that compared with the reference sample, patients with dextro-transposition of the great arteries were reported by their parents to have significantly more frequent attention problems, developmental delay/mental retardation, learning problems, and speech problems. Newburger et al [1984] also found that postponing repair of transposition of the great arteries was associated with a progressive impairment of cognitive function. A study by Oates et al [1995], however, showed that for children with cyanotic heart disease, there was no evidence of a detrimental effect of an older age at operation, as assessed by neuropsychological measures. In addition, delaying surgery for children with dextro-transposition of the great arteries

or tetralogy of Fallot did not appear to adversely affect their intellectual development. In our study, only 2 patients had delayed neuropsychological development, and 97% (68/70) performed normally in school functioning.

Study Limitations

First, we are sorry that neuropsychological testing and intelligence quotient (IQ) measurement was not done. Second, the long-term benefits of corrective ASO for these selected patients with severe PAH need to be demonstrated with a larger number of patients and a longer follow-up.

CONCLUSIONS

These data suggest that the quality of life of children after a corrective ASO at an older age (>6 months) is acceptable, compared with that of healthy children in China.

ACKNOWLEDGMENTS

We thank Professor Hui Chi for assistance with translating this article from Chinese into English.

REFERENCES

Apostolopoulou SC, Manginas A, Cokkinos DV, Rammos S. 2007. Long-term oral bosentan treatment in patients with pulmonary arterial hypertension related to congenital heart disease: a 2-year study. Heart 93:350-4.

Cella DF. 1994. Quality of life: concepts and definition. J Pain Symptom Manage 9:186-92.

Cenedese E, Speich R, Dorschner L, et al. 2006. Measurement of quality of life in pulmonary hypertension and its significance. Eur Respir J 28:808-15.

Culbert EL, Ashburn DA, Cullen-Dean G, et al. 2003. Quality of life of children after repair of transposition of the great arteries. Circulation 108:857-62.

Dunbar-Masterson C, Wypij D, Bellinger DC, et al. 2001. General health status of children with D-transposition of the great arteries after the arterial switch operation. Circulation 104(suppl I):I138-42.

Guyatt GH, Juniper EF, Griffith LE, Feeny DH, Ferry PJ. 1997. Children and adult perception of childhood asthma. Pediatrics 99:165-8.

Hendry F, McVittie C. 2004. Is quality of life a healthy concept? Measuring and understanding life experiences of older people. Qual Health Res 14:961-75.

Hövels-Gürich HH, Seghaye MC, Däbritz S, Messmer BJ, von Bernuth G. 1997. Cardiological and general health status in preschool- and school-age children after arterial switch operation. Eur J Cardiothorac Surg 12:593-601.

Immer FF, Althaus SM, Berdat PA, Saner H, Carrel TP. 2005. Quality of life and specific problems after cardiac surgery in adolescents and adults with congenital heart diseases. Eur J Cardiovasc Prev Rehabil 12:138-43.

Kamphuis M, Ottenkamp J, Vliegen HW, et al. 2002. Health related quality of life and health status in adult survivors with previously operated complex congenital heart disease. Heart 87:356-62.

Losay J, Touchot A, Serraf A, et al. 2001. Late outcome after arterial switch operation for transposition of the great arteries. Circulation 104(suppl I):I121-6.

Loup O, von Weissenfluh C, Gahl B, Schwerzmann M, Carrel T, Kadner A. 2009. Quality of life of grown-up congenital heart disease patients after congenital cardiac surgery. Eur J Cardiothorac Surg 36:105-11.

Lu Y, Tian Q, Hao Y, Jing J, Lin Y, Huang D. 2008. Reliability and validity for Chinese version of Pediatric Quality of Life Inventory PedsQL4.0 [in Chinese]. J Sun Yat-Sen Univ (Med Sci) 29:229-331.

Lunze K, Gilbert N, Mebus S, et al. 2006. First experience with an oral combination therapy using bosentan and sildenafil for pulmonary arterial hypertension. Eur J Clin Invest 36(suppl):32-8.

Nakajima Y, Momma K, Seguchi M, Nakazawa M, Imai Y. 1996. Pulmonary hypertension in patients with complete transposition of the great arteries: midterm results after surgery. Pediatr Cardiol 17:104-7.

Newburger JW, Silbert AR, Buckley LP, Fyler DC. 1984. Cognitive function and age at repair of transposition of the great arteries in children. N Engl J Med 310:1495-9.

Oates RK, Simpson JM, Cartmill TB, Turnbull JA. 1995. Intellectual function and age of repair in cyanotic congenital heart disease. Arch Dis Child 72:298-301.

Plummer M, Molzahn AE. 2009. Quality of life in contemporary nursing theory: a concept analysis. Nurs Sci Q 22:134-40.

Shafazand S, Goldstein MK, Doyle RL, Hlatky MA, Gould MK. 2004. Health-related quality of life in patients with pulmonary arterial hypertension. Chest 126:1452-9.

Uzark K, Jones K, Burwinkle TM, Varni JW. 2003. The Pediatric Quality of Life Inventory in children with heart disease. Prog Pediatr Cardiol 18:141-8.

Varni JW, Burwinkle TM, Katz ER, Meeske K, Dickinson P. 2002. The PedsQL in pediatric cancer: reliability and validity of the Pediatric Quality of Life Inventory generic core scales, multidimensional fatigue scale, and cancer module. Cancer 94:2090-106.

Varni JW, Seid M, Murtin PS. 2001. PedsQL 4.0: reliability and validity of the Pediatric Quality of Life Inventory version 4.0 generic core scales in healthy and patient populations. Med Care 39:800-12.

[WHO]World Health Organization. 1948. Constitution of the World Health Organization. In: Basic Documents. Geneva, Switzerland: World Health Organization.