Journal of Adolescent Health 46 (2010) 70–76

JOURNAL OF
ADOLESCENT
HEALTH

Original article

Self-Perception of Quality of Life by Adolescents with Neuromuscular Diseases

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Manuscript received February 1, 2009; manuscript accepted May 18, 2009

Abstract

Purpose: Little is known about quality of life of adolescents with neuromuscular diseases or the factors that influence it. We searched whether physical impairment, physical disability, and medical complications were predictors of low quality of life.

Methods: Motor function, health, orthopedic status, and rehabilitation were assessed in 43 adolescents with neuromuscular diseases (mean age, 13.8 years, standard deviation 1.7 year; sex ratio 2.9/1). Quality of life was measured with the VSP-A (''Vécu Santé Perçu par l'Adolescent''; self-perceived health state in adolescents), a validated health-related quality of life self-perception test. A mixed linear regression related quality of life to impairment, disability, and respiratory status. Comparisons were made with results from an age/sex-matched nondisabled group.

Results: On the average, the VSP-A scores in physically disabled adolescents were: (1) similar to those of the nondisabled group with regard to vitality, body image, relationships with parents and friends, and physical and psychological well-being; (2) higher with regard to school performance (score 68 vs. 52.8) and relationships with teachers (67.4 vs. 43.2); and (3) lower with regard to leisure activities (43.9 vs. 60.9). Physical disability and physical impairment were not negatively associated with seven of the nine VSP-A dimensions, but physical impairment was negatively associated with leisure activities and vitality ($p < .001$ and $p < .01$, respectively). Adolescents with ventilatory support did not express lower scores than adolescents not requiring ventilatory support (67.7 ± 11 vs. 62.9 ± 15, $p = .39$).

Conclusions: These surprising results should lead us question our medical, educational, and rehabilitation practices. Already well-managed disabled adolescents should benefit from less compassionate but more daring and dynamic interpersonal contacts.

Keywords: Quality of life; Neuromuscular disease; Physical impairment; disability

The course of most neuromuscular diseases, whatever their cause, is progression toward decline of motor functions. In conditions such as spinal muscular atrophy or congenital myopathy, children have to cope with a lifelong disease.

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doi:10.1016/j.jadohealth.2009.05.005

The resulting pain and physical impairment may require intensive rehabilitation, ventilatory support, and surgical interventions. These alter the patients' everyday living and their attitude toward life, not to mention their grappling with challenging developmental tasks concerning self-identity [1].

Nevertheless, patients have often remarkable adaptation abilities, and their quality of life (QoL) has been reported to remain reasonably stable over time, whatever the disease progression [2]. Several reports on QoL, physical disability, and daily functioning concluded that, in adults with muscular dystrophy, increased disability does not always lead to decreased life satisfaction [3–5]. In adults with Duchenne muscular dystrophy (DMD), a large majority of severely disabled adults seemed to be satisfied with their lives [6], and medical complications did not seem to adversely affect their QoL [3]. Even adults with DMD receiving ventilatory support considered their QoL satisfactory [7]. However, the impact of altered health and initiation of ventilatory support in children and adolescents is still largely unknown. Studying noninvasive ventilation in children with neuromuscular diseases, Young et al [8] expected a deterioration of QoL with increasing age and disability but found that QoL remained stable despite disease progression and initiation of ventilation support.

To date, there is no cure for neuromuscular disorders. The goal of pediatric rehabilitation remains to help disabled children ''live happily and productively on the same level and with the same opportunities as their neighbours'' [9] or ''enjoy their lives to the fullest possible extent'' [2]. Thus it becomes important to determine how far the disease and its treatment compromise a child's QoL. However, self-reported QoL or life-satisfaction reports in adolescents with neuromuscular diseases are rare because the tools used in adults are obviously inadequate.

Interested in the way that adolescents cope with neuromuscular diseases, we explored their feelings about their lives, specifically, their subjective degree of life satisfaction [1], and we compared their indicators of life satisfaction to those of a matched group of nondisabled peers.

Methods

Patients

''Neuromuscular diseases'' encompassed ''all disorders caused by an abnormality of any component of the lower motor neuron system: anterior horn cell, peripheral nerve, neuromuscular junction, or muscle'' [10]. The diagnosis rested on characteristic clinical features or proven genetic abnormalities.

The study included all patients with neuromuscular diseases 10–17 years of age and followed-up from November 1, 2005, to May 1, 2007, in three specialized outpatient clinics at Lyon, Grenoble, and St-Etienne (France). The study excluded patient intellectually unable to fill out a questionnaire, those with programmed surgery within 3 months, and those whose status was expected to substantially change within 6 months. Adolescents' and parents' informed consents were obtained, and the protocol was approved by the Ethics Committee of Lyon A. Only two consecutive adolescents refused to participate.

Data collection

A physician collected personal and clinical information and questioned disabled children and their parents about digestive symptoms (e.g., gastrostomy, gastroesophageal reflux, constipation, abdominal pain) and urinary or fecal incontinence. Pain was assessed using a 10-cm visual analog scale [11].

A reduced forced vital capacity (<70% of expected value), a history of recurrent respiratory infections, or use of lung hyperinflation were considered as ''moderate respiratory involvement,'' while ''severe involvement'' corresponded to the use of noninvasive ventilation or to mechanical respiratory support via invasive tubes. In all institutions, adolescents who required mechanical ventilation received it.

Physical impairment was evaluated with the Motor Function Measure (MFM), a validated test applicable to the main neuromuscular diseases whatever the degrees of physical impairment [12]. A 100% score indicates normal motor function.

Physical disability (inability to perform daily living activities and dependence on others or on technical aids) was evaluated with a Functional Independence Measure for Children (MIF Mômes), designed in English [13] and translated in French [14]. A score of 126 corresponds with complete autonomy.

We counted the number of severe contractures, especially those with functional consequences such as knee or hip flexum >45°, the number of weekly rehabilitation training sessions (psychologist or physical, occupational, or speech therapist), and the number of orthoses worn at least 4 days per week.

QoL was assessed either immediately after or within 6 months of a follow-up consultation (two-third and one-third of cases, respectively) using the VSP-A ''Vécu Santé Perçu par l'Adolescent'' (self-perceived health state in adolescents), a French health-related QoL (HRQoL) questionnaire [15], for which the reliability and validity in adolescents were established in 2005 [16]. The VSP-A assesses the perceived well-being and satisfaction in 10 major dimensions (vitality, physical and psychological well-being, body image, relationship with friends, parents, teachers, and medical staff, school performance, and leisure activities) and uses a five-point Likert scale (''worse'' to ''better'') as subscore for each item. Because the VSP-A part concerning the relationships with the medical staff was lacking in an earliest version of the questionnaire, the VSP-A score retained for all participants was the mean of the nine scores relative to the nine shared parts.