Quality of life of adolescents with neuromuscular diseases

Here what they say

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Introduction

The course of many neuromuscular diseases, whatever their cause, is progression towards decline of motor function. To date, there is no cure for neuromuscular disorders. The goal of paediatric rehabilitation remains to help children with disability « live happily and productively on the same level and with the same opportunities as their neighbours », Krause FM 1987 or « enjoy their lives to the fullest extent » Brown I 2002. Thus, it becomes important to determine how far the disease and its therapies compromise their life-satisfaction in a way to influence our medical, educative and rehabilitation practices. As a Quality of life (QoL) approach, we are interested in how pleased adolescents with neuromuscular diseases are and their feelings about the particular circumstances of their life. That represents the more subjective perspective of QoL also called individual’s degree of life satisfaction Remark R 1998.

Methods

The study included all patients with neuromuscular disease aged 10-17 and followed-up from November 1, 2005 to May 1, 2007. QoL was assessed using the VSP-A « Vecu Santé Perçu par l’Adolescent », a validated health-related QoL self-perception test. Comparisons were made with results from an age/sex-matched healthy group.

Function Measure = MFM), physical disability (Functional Independence Measure for children=MIFMôme were collected. A mixed linear regression related QoL to impairment and disability.

Conclusion

In our sample, a vast majority of disabled adolescents considered their lives worthwhile and rather enjoyable. The present astonishing results question our concepts of QoL and our current medical, educational, and rehabilitation practices. First, they show that it is very difficult to measure QoL without individual interviews that clarify what QoL is and what detracts from that quality. Second, they promote a shift towards more interest in the feelings, perceptions, and perspectives of these adolescents and more audacious timetables of care.

Discussion

To our knowledge, this is one of the few studies on self-perceived HRQoL in adolescents with neuromuscular diseases. For the first time, QoL scores were, on average, equal or higher to those of children in the general population in all domains except leisure activities where they were lower. This recalls the conclusion of a recent European Study on children with cerebral palsy Robinson MC 2007.

With few items on motor function, the VSP-A, we used assessed well-being, life satisfaction but not functional abilities. Diseased teenagers are used to cope with disability, medical complications, and, early in life, change their bearings and adjust their dreams to the reality of the disease. Thus, they could be satisfied with their lives despite impairment and disability.

Finally, physically disabled adolescents have specific priorities not addressed in the VSP-A; these should be included in any specific assessment of QoL in this population.

Results

43 adolescents (age 13.8±1.7, sexe ratio 2.9/1) were included: 34 used wheelchairs; 39 had respiratory involvement while 11 were receiving mechanical ventilation. Impairment MFM scores ranged from 0 to 98 (Mean 47.7, SD 25.9). Disability FIM scores ranged from 50 to 126 (Mean 79.4, SD 22.7).

In most domains of QoL, patients reported equal or even higher VSP-A scores than the age/sex-matched healthy group.

Significant effects of physical impairment (MFM) on dimensions of QoL

<table>
<thead>
<tr>
<th>Physical impairment</th>
<th>Total MFM [0-36] (n=42)</th>
<th>MFM [37-55] (n=14)</th>
<th>MFM &gt;55 (n=14)</th>
<th>p*</th>
</tr>
</thead>
<tbody>
<tr>
<td>School performance</td>
<td>67.8 ± 27.7</td>
<td>67.3 ± 22.6</td>
<td>62.7 ± 18.8</td>
<td>33.1</td>
</tr>
<tr>
<td>Body Image</td>
<td>70.3 ± 26.7</td>
<td>83.0 ± 11.6</td>
<td>67.7 ± 36.3</td>
<td>24.1</td>
</tr>
</tbody>
</table>

The most severely physically impaired adolescents [MFM<36] have expressed a higher QoL concerning body image than the least impaired adolescents [MFM>55].

QoL and mechanical ventilation

The 11 adolescents with mechanical ventilation had significantly more severe physical impairment and more physical disability than the 32 others (25.4 vs. 54.5, p=0.002 and 65.8 vs. 83.7, p=0.01, respectively). They also suffered more frequently from medical complications. However, the total VSP-A score in these severely disabled adolescents was not significantly different from that of the less impaired patients without mechanical ventilation (67.7 ± 11.3 vs. 62.9 ± 15.1, p=0.39).