

70. Robin G, Brief L: Scoliosis in childhood muscular dystrophy. *J Bone Joint Surg* 53-A:466-476, 1971
71. Robin G: The scoliosis of muscular dystrophy. In Robin G (ed): *Scoliosis and Neurological Disease*. New York, John Wiley, 1975, pp 49-70
72. Robinson D, Galasko CS, Delaney C, et al: Scoliosis and lung function in spinal muscular atrophy. *Eur Spine J* 4(5):268-273, 1995
73. Rodillo E, Fernandez-Bermejo E, Heckmatt J, et al: Prevention of rapidly progressive scoliosis in Duchenne muscular dystrophy by prolongation of walking with orthoses. *J Child Neurol* 3:269-274, 1988
74. Sakai D, Hsu J, Bonnett C, et al: Stabilization of the collapsing spine in Duchenne muscular dystrophy. *Clin Orthop* 128:256-260, 1977
75. Sanyal S, Johnson W, Thapar M, et al: An ultrastructural basis for electrocardiographic alterations associated with Duchennes progressive muscular dystrophy. *Circulation* 57:1122-1129, 1978
76. Schwentker EP, Gibson DA: The orthopedic aspects of spinal muscular atrophy. *J Bone Joint Surg* 58-A:32-38, 1976
77. Seeger B, Sutherland A, Clark M: Orthotic management of scoliosis in Duchenne muscular dystrophy. *Arch Phys Med Rehabil* 65:83-86, 1984
78. Shapiro F, Sethna N, Colan S, et al: Spinal fusion in Duchenne muscular dystrophy: A multidisciplinary approach. *Muscle Nerve* 15:604-614, 1992
79. Shapiro F, Specht L: The diagnosis and orthopedic treatment of childhood spinal muscular atrophy, peripheral neuropathy, Friedreich ataxia, and arthrogryposis. *J Bone Joint Surg* 75-A:1699-1714, 1993
80. Smith A, Koreska J, Eng P, et al: Progression of scoliosis in Duchenne muscular dystrophy. *J Bone Joint Surg* 71-A:1066-1074, 1989
81. Sullivan M, Thompson W, Hill G: Succinylcholine-induced cardiac arrest in children with undiagnosed myopathy. *Can J Anesth* 41:497-501, 1994
82. Swank S, Brown J, Ralph E: Spinal fusion in Duchenne's muscular dystrophy. *Spine* 7:484-491, 1982
83. Toda T, Blake D, Roche A, et al: Localization of a gene for Fukuyama type congenital muscular dystrophy to chromosome 9q31-33. *Nat Genet* 5:2883-2886, 1993
84. Tome F, Evangelista T, Leclerc A, et al: Congenital muscular dystrophy with merosin deficiency. *Life Sci* 317:351-357, 1994
85. Wada N: A study on spinal deformities in patients with progressive muscular dystrophy. *Skikoku Acta Med* 32:185-202, 1976
86. Wijmenga C, Frants R, Brouwer O, et al: The facioscapulohumeral muscular dystrophy gene maps to chromosome 4. *Lancet* 2:651-653, 1990
87. Wijmenga C, Padberg G, Moerer P, et al: Mapping of facioscapulohumeral muscular dystrophy gene to chromosome 4q35-qter by multipoint linkage analysis and *in situ* hybridization. *Genomics* 9:570-575, 1991
88. Wijmenga C, Hewitt J, Sandkuijl L, et al: Chromosome 4q DNA rearrangements associated with facioscapulohumeral muscular dystrophy. *Nat Genet* 2:26-30, 1992
89. Wilkins K, Gibson D: The patterns of spinal deformity in Duchenne muscular dystrophy. *J Bone Joint Surg* 58-A:24-32, 1976
90. Winter R, Lovell W, Moe J: Excessive thoracic lordosis and loss of pulmonary function in patients with idiopathic scoliosis. *J Bone Joint Surg* 57-A:972-977, 1975
91. Wollinsky K, Weiss C, Gelowicz-Maurer M, et al: Preoperative risk assessment of children with Duchenne muscular dystrophy and relevance for anesthesia and intra- and postoperative course. *Med Klin* 91:34-37, 1996
92. Yanagisawa A, Miyagawa M, Yotsukura M, et al: The prevalence and prognostic significance of arrhythmias in Duchenne type muscular dystrophy. *Am Heart J* 124:1244-1250, 1992

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QUALITY OF LIFE

Issues for Persons with Neuromuscular Diseases

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The goal of rehabilitation is to improve the physical, mental, social, and vocational aptitudes of persons who are disabled, with the objective of preserving their ability to live happily and productively on the same level and with the same opportunities as their neighbors.

KRUSEN IN HANDBOOK OF PHYSICAL MEDICINE AND REHABILITATION²¹

Although the goal of rehabilitation has been to help people with disabilities "live happily and productively on the same level and with the same opportunities as their neighbors," few studies have examined systematically what determines the quality of life of individuals with neuromuscular diseases. Lack of understanding and appreciation of such factors can significantly affect the perceived needs of these individuals, which often go beyond the medical care of their physical needs. Recent studies have documented that health care providers may underestimate the quality of life of individuals with severe neuromuscular diseases and that this may affect the level of service they receive. Our goals for this article are to document the subjective assessment of life satisfaction/quality of life by affected individuals and health care providers and to examine the relationship between these factors and life satisfaction. It is

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PHYSICAL MEDICINE AND REHABILITATION CLINICS OF NORTH AMERICA

understandable that the physician wants first to address the physical needs of the patients with neuromuscular diseases. However, to optimize care, physicians need: (1) to determine whether their own attitudes and expectations restrict potential outcomes; (2) to be proactive and provide information about the social and service needs that influence the quality of life of individuals; and (3) to work in partnership with patients to better understand the issues that affect their quality of life.

Historically, physicians often have made decisions for the good of the patient without the patient's consent and/or without the patient's knowledge. However, over the past four decades there has been an evolution from a medically based model of care to a model that incorporates consumer control and autonomy. Consumers have demanded and appropriately have been given more control over their care. The idea of personal rights has been expanded and has resulted in the development of a patient's Bill of Rights. The principle of autonomy encompasses the patient's right to know, the patient's right to consent to treatment, the patient's right to refuse treatment, and the patient's right to confidentiality and privacy. This situation has resulted in patients taking much more responsibility for their medical care. Health-related quality of life concepts emerged that included areas of health outside of physical needs.

Although the principle of autonomy is now embodied in law,²⁷ people with disabilities frequently do not receive adequate information to make informed decisions to guide their care and live independently. This can happen when physicians and other health care providers unwittingly project their own negative feelings about the potential quality of life of an individual with a disability when informing patients about potential outcomes. These negatively biased attitudes limit activities that could improve health outcomes and community integration. In the most egregious cases, these inadvertent negative attitudes may even prevent life-saving procedures.

Recent studies have shown that misconceptions regarding the potential quality of life of an individual who requires ventilatory support has prevented physicians from prescribing these life-sustaining treatments. Often these decisions are made without regard to the patient's desires. For example, Dracup⁹ has recommended that physicians rely on their own assessment of the patient's quality of life, and that this be done "independent of the patient's feeling" when making the decision to institute mechanical ventilation. In another study, Bach used the Index of Domain Satisfaction to assess the quality of life reported by Duchenne muscular dystrophy ventilator users.¹ The reports of these individuals with this progressive neuromuscular disease were compared with the assessment of the quality of life of the same patients by their physicians. Not only did these physicians underestimate the quality of life of the Duchenne muscular dystrophy ventilator users, but the physicians who discouraged ventilator use underestimated the quality of life to a greater degree than did the physicians who recommended ventilator aids. In addition, the physicians who discouraged ventilator use cited

poor quality of life as the most common reason for not recommending its use. As stated by Bach,¹ "the physicians' judgment of the patients' quality of life and their failure to accurately evaluate their patients' satisfaction with life influenced whether or not life-sustaining ventilatory assistance was used."

Although these cases represent only one end of the spectrum, other studies also have shown that health care professionals can significantly underestimate the life satisfaction of individuals with disabilities. Gerhart et al¹⁵ examined the attitude of 153 emergency care providers toward quality of life after spinal cord injury (SCI) and compared it to the quality of life perceived by 128 individuals with tetraplegia. Whereas 92% of those with tetraplegia reported that they were glad to be alive, only 18% of the emergency room technicians imagined that they would be glad to be alive after tetraplegia. Forty-five percent of these health care workers reported that if they did have a serious SCI, they would want nothing more than pain relief and would prefer death over life with a disability. The emergency care providers who had the most negative attitudes toward quality of life after SCI were least likely to favor aggressive life-saving techniques performed on them if they were to sustain a spinal cord injury. Similar negative attitudes toward quality of life after disability have been reported by nurses,³ medical students,²⁶ and resident physicians.¹⁰

These negative attitudes toward disability are not limited to nondisabled individuals. Disabled people, like nondisabled people, overestimate the negative effect that disabilities have on other disabled individuals. In a study by Campbell et al,⁶ disabled subjects who had experienced blindness, amputation, diabetes mellitus, cerebral palsy, or schizophrenia of long standing (at least 2 years for adaptation) ranked the impact of their disability on a fortunate-unfortunate scale. They were also asked to rank their perception of the effect of disability on individuals from each of the other groups. Subjects in each of the disability categories ranked themselves as being more fortunate than subjects with other types of disabilities. The only exception to this case was for the group with cerebral palsy, who ranked themselves as second most fortunate to people with diabetes. Regardless, they still ranked themselves as being more fortunate than individuals with blindness, amputation and schizophrenia. This research suggests that persons adapt to their disability and regain a measure of control over their environment. This contrasts sharply with newly disabled individuals, who are known to feel despair over their circumstances and who frequently feel unsympathetic toward others with disabilities.⁶ These studies demonstrate that most people instinctively react negatively to things they do not understand and that the only way to truly understand the impact of a disability is to experience it.

Several other studies also have shown that individuals with long-standing disabilities generally accept their functional limitations. Weinberg and Williams²⁹ queried 88 physically disabled people about the significance of their disability. Only 7% considered it to be "the worst

