

**VENTILATOR USE FOR PATIENTS WITH DUCHENNE  
MUSCULAR DYSTROPHY: AN ETHICAL ANALYSIS  
OF PHYSICIANS' BELIEFS AND PRACTICES**

by

**Barbara Gibson**

A thesis submitted in conformity with the requirements for the degree of  
Master of Science  
Institute of Medical Sciences  
Collaborative Program in Bioethics  
University of Toronto

© Copyright by Barbara Gibson 1998



National Library  
of Canada

Acquisitions and  
Bibliographic Services

395 Wellington Street  
Ottawa ON K1A 0N4  
Canada

Bibliothèque nationale  
du Canada

Acquisitions et  
services bibliographiques

395, rue Wellington  
Ottawa ON K1A 0N4  
Canada

*Your file Votre référence*

*Our file Notre référence*

The author has granted a non-exclusive licence allowing the National Library of Canada to reproduce, loan, distribute or sell copies of this thesis in microform, paper or electronic formats.

The author retains ownership of the copyright in this thesis. Neither the thesis nor substantial extracts from it may be printed or otherwise reproduced without the author's permission.

L'auteur a accordé une licence non exclusive permettant à la Bibliothèque nationale du Canada de reproduire, prêter, distribuer ou vendre des copies de cette thèse sous la forme de microfiche/film, de reproduction sur papier ou sur format électronique.

L'auteur conserve la propriété du droit d'auteur qui protège cette thèse. Ni la thèse ni des extraits substantiels de celle-ci ne doivent être imprimés ou autrement reproduits sans son autorisation.

0-612-33948-3

**Canada**

VENTILATOR USE FOR PATIENTS WITH DUCHENNE MUSCULAR DYSTROPHY: AN  
ETHICAL ANALYSIS OF PHYSICIANS' BELIEFS AND PRACTICES

Barbara Gibson  
Institute of Medical Sciences  
Collaborative Program in Bioethics  
University of Toronto, 1998

**Abstract**

Long term ventilation (LTV) has been shown to extend the lives of individuals with Duchenne muscular dystrophy (DMD); however, initiating LTV is still considered controversial. This study examined Canadian practice from the perspective of bioethics. The purposes of the study were: 1) to describe current Canadian practice, 2) to defend a claim that all patients with DMD should be informed of the option of LTV and 3) to make recommendations designed to enhance the decision making process. The research included a physician survey, face-to-face interviews, and normative ethical analysis. LTV decisions are discussed within the context of the patient- physician relationship and a conceptual exploration of “quality of life” and “futility”. It is concluded that LTV information should be disclosed to all patients/families with DMD. A shared decision making model is proposed that emphasizes ongoing dialogue over time, addresses power inequities within the family-physician relationship and incorporates a family-centred approach to decision making.

## **Acknowledgments**

I wish to thank the many people who contributed to the development and completion of this thesis. I wish to first acknowledge the efforts of my committee members, Dr. Karen Yoshida (supervisor), Dr. Laura Shanner and Dr. William Harvey who facilitated my learning in a diverse area of topics and methodologies, provided constant positive feedback and helped me to find my “voice.” I also need to recognize the support of my two mentors, Dr. Dina Brooks and Chantal Graveline who so generously shared of their time and wisdom. As well, I would like to thank my family for their constant encouragement, support and humour.

I would also like to thank the Ontario Respiratory Care Society for providing me with fellowship funding without which this research would not have been possible.

Finally but most of all, I thank my partner in life, Richard Smith, first for his technical assistance, but mostly for his unwavering and selfless support, patience and encouragement throughout this journey.

## **TABLE OF CONTENTS**

<b>CHAPTER 1: INTRODUCTION AND BACKGROUND</b> .....	<b>1</b>
INTRODUCTION .....	1
LIMITATIONS OF THE STUDY AND FUTURE DIRECTIONS.....	3
RELEVANCE AND BACKGROUND.....	4
<b>CHAPTER 2: LITERATURE REVIEW</b> .....	<b>12</b>
INTRODUCTION .....	12
INFORMED CONSENT .....	13
THE PHYSICIAN-PATIENT RELATIONSHIP .....	24
QUALITY OF LIFE AND ATTITUDES TOWARDS DISABILITY.....	34
SUMMARY OF LITERATURE REVIEW .....	41
<b>CHAPTER 3: EMPIRICAL METHODOLOGY</b> .....	<b>43</b>
OVERVIEW OF EMPIRICAL METHODOLOGY .....	43
PART ONE: MAIL QUESTIONNAIRE .....	43
PART TWO: INTERVIEWS.....	45
<b>CHAPTER 4: RESULTS</b> .....	<b>49</b>
PART ONE: QUANTITATIVE DESCRIPTION OF PRACTICE AND ATTITUDES .....	49
PART TWO: QUALITATIVE DESCRIPTION OF PRACTICE AND ATTITUDES .....	70
SUMMARY AND DISCUSSION OF EMPIRICAL RESULTS.....	86
<b>CHAPTER 5: NORMATIVE ANALYSIS</b> .....	<b>92</b>
PART ONE: NONDISCLOSURE.....	93
PART TWO: SHARED DECISION MAKING: APPLICATIONS TO LONG TERM VENTILATION DECISIONS.....	109
<b>CHAPTER 6: CONCLUSIONS AND RECOMMENDATIONS</b> .....	<b>128</b>
RECOMMENDATIONS.....	131
<b>REFERENCES</b> .....	<b>134</b>

**APPENDICES.....144**

1. a) ENGLISH VERSION OF SURVEY.....144  
b) FRENCH VERSION OF SURVEY.....157

2. a) ENGLISH VERSION OF COVER LETTER .....171  
b) FRENCH VERSION OF COVER LETTER .....172

3. INTERVIEW GUIDE.....174

4. CONSENT FORM.....175

## List of Tables

TABLE 1: NUMBER OF PATIENTS WITH DMD FOLLOWED BY THE PHYSICIANS .....	51
TABLE 2: NUMBER OF VENTILATED PATIENTS FOLLOWED BY THE PHYSICIANS .....	52
TABLE 3a: NUMBER OF PHYSICIANS WHO ALWAYS GIVE THE <i>SAME</i> ADVICE REGARDING VENTILATION: TYPE OF ADVICE BY FREQUENCY .....	59
TABLE 3b: NUMBER OF PHYSICIANS WHO GIVE <i>VARYING</i> ADVICE REGARDING VENTILATION: TYPE OF ADVICE BY FREQUENCY.....	59
TABLE 4: REASONS FOR NOT OFFERING/RECOMMENDING VENTILATION .....	61
TABLE 5: REASONS FOR INFORMING PATIENTS/FAMILIES ABOUT VENTILATION .....	62
TABLE 6a: TIMING OF VENTILATION DISCUSSIONS: INITIAL DISCLOSURE .....	64
TABLE 6b: TIMING OF VENTILATION DISCUSSIONS: IN-DEPTH DISCUSSIONS.....	64
TABLE 7:DO YOU AGREE OR DISAGREE THAT SOCIETY CANNOT AFFORD VENTILATION FOR PATIENTS WITH DMD? PHYSICIANS' OPINIONS BY MEAN MIDS SCORE .....	67
TABLE 8: DISCLOSURE PRACTICE BY GENDER.....	69
TABLE 9: DEMOGRAPHIC DISTRIBUTION OF INTERVIEW PARTICIPANTS.....	71

## List of Figures

FIGURE 1A: TYPES OF FULLTIME VENTILATION.....	54
FIGURE 1B: TYPES OF PART TIME VENTILATION.....	55
FIGURE 1C: TYPES OF VENTILATION: COMBINED FULL AND PART TIME.....	56
FIGURE 2: FREQUENCY OF INFORMING PATIENTS ABOUT THE OPTION OF LTV.....	57
FIGURE 3: OVERVIEW OF PHYSICIANS' MANAGEMENT OF VENTILATION DECISIONS .....	73

# Chapter 1: Introduction and Background

## Introduction

As medical technology continues to advance at a rapid rate, the ethical problems that health care professionals face in day to day practice are becoming increasingly complex. Since the landmark case of Karen Quinlan,<sup>1</sup> there has been much discussion in the medical, ethical and public arenas regarding the appropriate use of mechanical ventilation. Much of this discussion focuses on the critically ill patient who is maintained on a positive pressure ventilator in an intensive care unit. This focus, while important, may not be relevant to the unique situation of those individuals who require ventilation as a progression of their rehabilitative care or ongoing maintenance. Ventilator users with neuromuscular diseases may be living in the community, holding jobs, and/or maintaining marital relationships. These life circumstances vary significantly enough from the situation of the intensive care patient that ethically appropriate care for one group may not necessarily apply to the other.

Whether or not to offer patients with neuromuscular diseases long term ventilation (LTV) still remains a subject of controversy.<sup>2,3</sup> In the past, individuals with Duchenne muscular dystrophy (DMD) could expect to survive into their late teens or early twenties. More recently, mechanical ventilation has been shown to prolong the lives of these individuals, but it does not offer a cure. Instead, the ventilator offers the continuation of a life that will grow even more dependent on third party care and electrical devices.<sup>2</sup> As Gilgoff asks,

The respirator: Is it the tool of the devil, entrapping the helpless patient and keeping him from the peaceful sleep of death, or is it the angel of mercy, saving a stricken young man from the menacing jaws of death?<sup>2</sup>

This question reveals some of the ethical controversy surrounding the practice of providing LTV for patients with Duchenne muscular dystrophy. Some physicians in the United States do not inform their patients of the option of ventilation or discourage this choice.<sup>4,5</sup> Reasons for not disclosing or discouraging LTV include unilateral value judgements regarding patient quality of life or amount of suffering.<sup>4,6</sup> Similar research examining physician attitudes and practices has not been previously undertaken in Canada until the present study.

The purposes of this thesis are: (A) to describe current Canadian practice, (B) to defend the claim that all individuals with Duchenne muscular dystrophy should be informed of the option of long term ventilation and (C) to make recommendations designed to enhance the decision making process. The first purpose was achieved by collecting and analysing data from a mail survey of Canadian physicians who follow patients with DMD and through qualitative interviews with a subsample of the physicians. The second purpose was achieved through normative ethical analysis. The third purpose was achieved through further normative analysis of the patient-physician relationship within the context of decisions regarding LTV. The descriptive information collected on Canadian practice informs this analysis.

This thesis considers the practices of physicians following patients with Duchenne muscular dystrophy, the most common type of neuromuscular disease. However, it is believed that the research will have implications for all individuals with severe neuromuscular diseases and for medical decision making in general.

### **Limitations of the Study and Future Directions**

Because one of the purposes of this research was to describe Canadian practice, a questionnaire was chosen as the appropriate methodology to collect information on practices across the country. However much information was gained from the qualitative interviews. A qualitative study that involved a larger sample size may provide further understanding of current practice.

The questionnaire is limited by response bias in that what clinicians *report* they do is not necessarily *what* they do. This issue is partially addressed by the anonymity of the survey. There also exists the potential for social desirability bias as clinicians may report what they believe is a socially desirable answer rather than their actual beliefs. Again this issue is partially addressed by anonymity.

The research examined the decision making processes of physicians only. While this information is relevant to practice, it does not provide complete information. Further research is needed to examine the experiences of patients and families, as well as the contributions and experiences of other members of the health care team. Participant observation of clinician - family discussions would contribute to this process.

In the thesis I limited my analysis to cognitively competent patients with DMD. Cognitively impaired individuals or younger children who have different diagnoses (for example Spinal Muscular Atrophy) will depend on their caregivers to make decisions on their behalf. Since a significant number of children with DMD may experience some degree of mental retardation, further exploration of the concept of competency in decision making is needed to determine the role of these individuals in LTV decisions. These sorts of cases have not been

considered here and the particulars of these cases may lead to a further refinement of the recommendations.

The thesis looked at LTV decisions from the perspective of the physician - patient/family relationship but not from a broader societal perspective. Political and economic issues also influence decision making. Some may believe that offering LTV constitutes an improper use of scarce public resources. Further study is needed to assess the normative validity of such beliefs and the potential effects on decision making.

## **Relevance and Background**

### *Duchenne Muscular Dystrophy: The Disease Process*

Duchenne muscular dystrophy (DMD) is a genetically inherited disease affecting male children which is characterised by progressive weakening of skeletal muscle. Weakness usually begins in the hip musculature and advances to include all the muscles of the legs, arms and trunk with a corresponding loss of physical function. By the ages of 12 to 16, most patients require a wheelchair for mobility.<sup>7</sup> As weakness progresses, the individual may take advantage of various technologies to assist with physical function. These include electric wheelchairs, robotic manipulators, voice synthesizers, environmental control systems, and adapted switches which can be interfaced with a personal computer. Use of these devices can help to increase the independence of the individual and increase his participation in recreational and social activities. Once the child is no longer walking, progressive spinal scoliosis begins. The progressing scoliosis, along with a marked weakening of the muscles of respiration, eventually leads to respiratory insufficiency. In time, the patient no longer has sufficient strength to breathe

independently. Signs of respiratory compromise include frequent pulmonary infections and hypercapnia. Clinically, individuals may present with nausea and vomiting, poor concentration, impaired intellectual functioning or changes in body weight.<sup>3</sup> Life expectancy without the assistance of ventilatory aids is 18 to 22 years with the usual causes of death being respiratory failure.

### *Ventilation*

As the technology has advanced, ventilator assistance has increasingly been offered as an option to extend the lives of patients with DMD.<sup>3,8</sup> Curran (1989) found that ventilatory assistance increased the average lifespan of a group of twenty-three DMD patients by six years.<sup>9</sup> At present there are individuals in the community whose lives have been extended by greater than 10 years with the assistance of mechanical ventilation.<sup>10</sup> Ventilatory assistance comes in many different forms. These are generally grouped according to 1) invasiveness - tracheotomy vs several non-invasive options, and 2) the use of positive vs negative pressure ventilation. As the patient begins to develop symptoms of respiratory failure, he may initially require night-time ventilation with a non-invasive mode of ventilation such as a nasal mask or lip seal.<sup>11</sup> As the disease progresses, the patient eventually requires respiratory support twenty four hours a day in order to survive. This can be accomplished with positive pressure ventilation delivered via a tracheostomy or with one of the non-invasive modes.<sup>3</sup> In many instances, particularly when ventilation is initiated in response to a crisis situation, ventilation via tracheostomy may be the individual's first and only form of ventilatory assistance used. The ventilator is portable and can be mounted directly onto the patient's wheelchair allowing mobility and community access.

Despite the proven success of ventilation in extending the lives of individuals with DMD, it appears that it is still not routinely offered or discussed with all patients. Colbert and Schock<sup>5</sup> surveyed Muscular Dystrophy Association (USA) clinic directors in 1985 and found that 24% never provided respiratory support systems while 43% provided respiratory support systems only under special circumstances. There appeared to be no standardized protocol for offering or not offering ventilation. Some directors only offered non-invasive means, while others said they wouldn't offer ventilation on a "chronic basis" or for "terminal cases." A more recent study was conducted by Bach in 1992.<sup>4</sup> Of 221 Muscular Dystrophy Association clinic directors surveyed, 33% never discussed ventilatory assistance with their patients, 31% discussed it with "at least a few patients" and only 36% discussed it with all or most patients. The most frequently cited reasons for not offering ventilation were: "poor patient quality of life", "lack of adequate financial resources", "inadequate home environment", "cognitive deficits", "excessive burden on the family" and "families uninterested or unwilling to discuss it". In their comments, the directors also made reference to the notion of futility with such statements as "it is only postponing death" or "it is inappropriate in terminal illness."

Several ethical concerns are raised by the results of these studies. In the literature review, I will outline the related concepts and review the relevant literature on each.

### *Theoretical Approach*

The proposed investigation approaches the issue of physician practice and long term ventilation from the perspective of bioethics. Bioethics is a multidisciplinary study of moral problems in medicine and the medical sciences. In this investigation, I wish to describe current practice as well as evaluate that practice and provide normative guidance.

Problems can be considered ethical in nature when they raise concerns about the rightness or wrongness of an action, policy, or personal characteristic.<sup>12</sup> Some notion of what constitutes "the good" or "humanity's highest good" is inherent in all ethical inquiry.<sup>13</sup> *Descriptive* ethics seeks to report what people believe and how they act while *normative* ethics seeks to evaluate whether an action is right or wrong.<sup>14</sup>

This research engages in both descriptive ethics (describing current practice and beliefs) as well as normative ethics (evaluating the appropriateness of those practices and beliefs). To achieve this end, a multi-method, multi-disciplinary approach is required. Empirical methodologies (survey methodology and qualitative interviews) are employed to collect information on current beliefs and practices. Philosophical inquiry is required to investigate which practices can be considered ethical and why. Further elaboration of what is meant by philosophical inquiry will help to clarify my intent.

Brody<sup>15</sup> discusses the philosophical approach as a form of qualitative analysis in which one attempts to find meaning using a variety of divergent sources of information. "Thick description" is employed to capture adequately the nature and complexity of the problem, while active reflection is necessary to examine assumptions and establish clear definitions of terms. Much of the normative work therefore involves concept clarification and refinement of

definitions. Integral to the process is the evaluation of competing arguments for internal consistency and coherence as well as logical validity.

As an example, the research examines the concept of "quality of life." The survey portion of the research demonstrates that some physicians advise against ventilation because they believe the patient's quality of life on a ventilator will be poor. To evaluate the moral appropriateness of this rationale, it is necessary to clarify what is meant by "quality of life," who should be making these judgements and whether or not predicted poor quality of life counts as a morally valid reason for advising against ventilation.

The work of values clarification and the examination of competing arguments for consistency and coherence employs the process of wide reflective equilibrium described by Daniels.<sup>16</sup> Wide reflective equilibrium generally involves identifying a set of considered moral judgements reached in a single case or a group of similar cases, formulating principles that account for them, comparing these principles to background philosophical theories generally believed to be true (e.g., the point of morality is to protect the interests or well being of individuals), testing the principles on similar cases with contextual differences, and then revising the considered judgements, principles and background theories until reaching equilibrium.<sup>17</sup> In this way moral reasoning involves both inductive and deductive reasoning, proceeding "up" from cases to theories as well as "down" from theories to cases.

Physician practice regarding long term ventilation is a complex phenomenon influenced by a variety of socio-cultural and moral elements. The proposed research therefore takes a multidisciplinary approach. Empirical methodology is employed to determine practices and normative analysis to determine the rightness or wrongness of those practices.<sup>15</sup> A variety of background theories and concepts from the social sciences and the field of bioethics will be

explored in the literature review. Once the data from the empirical portion of the research is presented, these concepts will be revisited with the purpose of analysing current practice and formulating normative guidelines.

### *Specification of philosophical approach*

To demonstrate that the methods of philosophical inquiry are indeed employed in the thesis, I provide here an outline of the process of wide reflective equilibrium as it applies to the work.

The thesis begins with a *considered judgment*: LTV information ought be provided to patients with DMD and their families. This judgment arises from my own clinical experiences as well as from the opinions presented in the medical literature, in particular the studies by Bach<sup>6</sup> and Colbert and Schock<sup>7</sup> referred to above. Nevertheless, this initial judgment is provisional and subject to revision. *Moral principles that account for this judgment* are then sought. The principle of respect for autonomy initially seems to account for this judgment. *Background theories commonly believed to be true* are then consulted. In medical practice, it is commonly held that a central purpose in medicine is to positively affect the patient's health (whether this be through prevention, cure, rehabilitation, or palliation). In addition, the maxim "one ought to respect persons" is generally held to be true in our common morality. *The moral principle that was thought to account for the initial considered judgment is then revised*. In addition to autonomy, beneficence and protection of the vulnerable also seem to be norms relevant to the practice of disclosing/withholding LTV information. The reflective work to this point (moving from an autonomy centred account of disclosure to an appeal to other principles) is done prior to the writing of the thesis such that, in the literature review, these principles are presented together

and defended as duties that arise from an overarching principle of respect for persons. A general exploration of the demands of beneficence and autonomy as applied to the patient-physician relationship is presented through the use of three models: paternalism, consumerism and shared decision making, each of which emphasizes these duties differently. These models provide a framework for discussing the particular actions that may be demanded by autonomy, beneficence and protection of the vulnerable and provide a richer description of the relationship. Shared decision making is defended as the model of choice for the majority of patient-physician relationships because it attempts to address these three duties without sacrificing one to the other(s). This provisional conclusion is then *tested against the case and similar cases*. This work appears in the normative analysis. To do this, the methods of philosophical inquiry are employed as follows. (1) Competing arguments are examined for consistency and clarification. For example, if LTV is futile for patients in a persistent vegetative state, does it follow that it is futile for patients with DMD? (2) Thick description is employed. What features of this case make it different or the same? What do we mean by 'futile' treatment? (3) A further *specification* of the shared decision making model is proposed based on the particular features of the case: the chronic nature of the illness, the relatively large amount of time available for decision making, and the need to include family members and the developing child as decision makers. In addition, a common feature of all patient-physician relationships is addressed: the issue of power imbalance and the effects on physician-family communication. The *initial considered judgement is then revised* as follows: One ought to disclose information regarding LTV to patients and families with DMD and assist the family to make decisions consistent with their collective and individual life plans. From this the *background theories are also revised*: Respect for persons is unchanged as a maxim but has now been specified to the case. Medical practice ought strive not

only to have a positive effect on the patient's health, but also to bring about an effect consistent with the family's life plan. Together physician, patient, and family work together to discover "the good" for a particular patient and family based on the family's goals and values and the options medicine can offer. The thesis concludes by offering normative guidance for how these objectives can be achieved in practice for LTV decisions. A shared decision making approach that borrows from Katz's "conversation model" is advanced.

The normative work therefore involves constant refinement and revision of considered judgements, principles and background theories until equilibrium is reached. While the process is both inductive and deductive, in the thesis it is largely presented in a deductive, linear fashion that proceeds from presentation of theory to application to the case at hand. This is done for purposes of organisational clarity.

## **Chapter 2: Literature Review**

### **Introduction**

Some of the possible reasons for the practice of not offering ventilatory assistance were investigated in Bach's 1992 American study discussed above.<sup>4</sup> These are: “poor patient quality of life”, “lack of adequate financial resources”, “excessive burden on the family” and “families unwilling or uninterested in discussing.” Other reasons cited included: “ventilation is prolonging death”, “clinician's personal philosophy” and “benefits not significant enough.”

Unilateral judgements made by physicians regarding a patient's potential quality of life or the possible burdens to his/her family raise questions about how medical decisions are made and the nature of the physician-patient relationship. Quality of life judgements may be subjective and value laden.<sup>18,19,20</sup> Furthermore, quality of life judgements and statements about prolonging death may be related to general attitudes towards disability. Reasons that deal with cost may be less prominent in Canada where LTV is covered by public health insurance. The reasons why physicians do offer ventilation may be explained by adherence to the ethical and legal demands of informed consent and positive attitudes towards persons with disabilities.

A conceptual and empirical overview of informed consent, the patient-physician relationship and quality of life will serve to illuminate some of the possible factors that impact on practice. These concepts are explored with regard to their meaning and scope and the principles and duties that form their ethical underpinnings.

## **Informed consent**

### *Overview of key concepts*

Informed consent is "the procedure whereby patients consent to - or refuse - a medical intervention based on information provided by a health care professional regarding the nature and potential consequences of the proposed medical intervention."<sup>21</sup> Valid informed consent contains at least the following elements: competency, voluntariness, disclosure of information and patient understanding of that information.<sup>22,23,24,25</sup> Consent is grounded in the ethical principle of respect for persons and is affirmed by Canadian law, professional policy and ethical guidelines. Although the core purpose of consent is to support autonomous decision making, the consent process is also informed by a duty of beneficence - an obligation to act in the patient's best interests. Obtaining consent is not a single *event* but rather a *process of decision making* that occurs within the context of the patient-physician relationship.<sup>26</sup>

### *Professional, legal and ethical support*

Virtually all prominent medical and institutional codes of ethics now include rules that demand health care professionals obtain the informed consent of the patient prior to implementing any intervention.<sup>27</sup> Informed consent has been required by Canadian common law since the 1980 Supreme Court of Canada's decision in the case of *Reibel v. Hughes*.<sup>28</sup> Since 1992, the province of Ontario has had specific provincial legislation governing consent procedures.<sup>29</sup> British Columbia and Prince Edward Island are in the process of enacting similar legislation.<sup>30,31</sup>

Support for the idea of informed consent is promulgated by the Royal College of

Physician's and Surgeons of Canada:

The physician is morally obliged to protect this right by providing patients with all reasonable data about diagnostic and therapeutic procedures, and possible alternatives and risks, and by allowing patients to make their decisions without coercion.<sup>32</sup>

The Code of Ethics of The Canadian Medical Association makes a similar statement:

Provide your patients with the information they need to make informed decisions about their medical care, and answer their questions to the best of your ability.<sup>33</sup>

Outside of Canada, in the U.S. the President's Commission for the Study of Ethical Problems in Medicine and Biomedical and Behavioral Research asserts that properly informed consent is a necessary element of responsible medical decision making.<sup>34</sup>

It seems then that sources from the disciplines of medicine, law and ethics all agree that patients have the right to make informed decisions about their own health care. Despite this agreement, it is sometimes not clear what is demanded by a policy of informed consent. Although the doctrine of informed consent has foundations in law, it is primarily an ethical imperative.<sup>34</sup> To understand the moral\* (as distinguished from purely legal) demands of consent it is necessary to examine the ethical principles on which the idea of informed consent is based.

### *Informed consent and decision making*

“Informed consent” is often criticised as being a limited concept that emphasizes disclosure of information rather than a true shared decision making process.<sup>35</sup> Because “informed

---

\* In this thesis the terms “moral” and “ethical” are used interchangeably

consent” is a term that is used in the context of medicine, law and ethics, it may hold different meanings to persons trained in each of these disciplines.<sup>36</sup> Dickens<sup>37</sup> has commented that the expression “informed consent” does not accurately describe the physician-patient interaction. Instead it implies that the purpose of giving information is to obtain consent rather than to reach a decision. Some scholars prefer to use terms that more accurately reflect the process such as *informed decision making* or simply *decision making*.<sup>37</sup> In this thesis I use the term “informed consent” to encompass the broader understanding of consent as a mutual process of decision making that occurs between a patient and his/her physician and that may also involve other members of the medical team and of the patient's family.<sup>26,34,36,38</sup>

#### *Value basis of informed consent*

The obligation to engage patients in informed decision making flows from an overarching principle of respect for persons.<sup>22,39,40,41</sup> The maxim “one ought to respect persons” has its foundations in religion<sup>42</sup> and in two broad secular ethical traditions: deontology and utilitarianism.<sup>36,42,43</sup> Before discussing the meaning and scope of the principle of respect for persons, these two traditions are briefly presented to demonstrate their contributions to a modern understanding of the principle.

Deontology is an obligation based theory of ethics.<sup>44</sup> This system of ethics is sometimes referred to as Kantian Ethics after its most influential exponent, Immanuel Kant (1724-1804). Essentially, Kantians purport that actions in and of themselves can be judged right or wrong and consequences are only relevant if the action is morally permissible. According to this view, persons must never be treated merely as a means to an end but always also as ends in themselves.<sup>45</sup> Kant believed that rational deliberation was the ultimate test of moral rules and

that all rational agents would produce the same maxims if employing reason alone. Morality, in his view, provided a rational basis for a system of rules that govern the actions of everyone, apart from their individual desires or intuitions.<sup>46</sup> The supreme moral rule for Kant is his categorical imperative which states: "Act only on the maxim that you can at the same time will that it would become a universal law." Thus Kant believed for a moral rule to be valid it must be universalizable, it must be derived from reason alone and it must not allow for persons to be treated merely as means to an end.

Applied to the modern medical encounter, Kantian ethics demands that physicians do not attempt to coerce patients into choosing one treatment option over another. Such coercion would violate a patient's status as a rational person capable of self-determination. In addition, because rational deliberation is presumably dependent on a foundation of relevant facts, Kantian ethics also demands that clinicians provide all the factual information necessary for their patients to make a reasoned choice.<sup>46</sup>

In contrast to deontological ethics, utilitarianism as advanced by John Stuart Mill (1806 - 1873) holds that consequences are the only relevant measure of the rightness or wrongness of an action.<sup>47</sup> Mill invoked the principle of *utility* where the utility of an action is the degree to which it produces or promotes happiness, or removes or avoids unhappiness. In deciding how to act, an agent ought choose that action which produces the best overall ratio of happiness and unhappiness for all affected. Mill believed that persons should be allowed the maximum freedom to live according to their own personal convictions as long as exercising this freedom did not interfere with the freedom of others to do the same.

### *Respect for persons*

The deontological and utilitarian traditions provide the moral foundations for the modern notion of informed consent. Consent is based primarily on the moral principle “one ought to respect persons”<sup>22,39,40,41</sup> The specific duties derived from this principle are not universally agreed upon. Two fundamental points are at issue: a) what constitutes *personhood* and b) what is meant by *respect*. For example, Kant emphasized that only rational beings belong to the community of persons. His definition would exclude not only animals but also young children and those who are cognitively impaired. *Respect* could potentially demand a number of negative and positive obligations depending on one's interpretation or ethical orientation. Respect might demand non-interference with a person's pursuit of life goals (a utilitarian/libertarian perspective) or a positive duty to assist those in need simply because they are persons (a deontological perspective).

As it relates to informed consent, at least three duties flow from the principle of respect for persons: respect for autonomy, protection of the vulnerable and promotion of the patient's wellbeing (beneficence).<sup>22,39,40,41,42</sup> It is generally accepted that autonomy, protection of the vulnerable and beneficence are guiding principles in health care and that health care professionals have a corollary *duty* to honour these principles (amongst other principles not immediately relevant to the thesis). These duties appear in current medical codes of ethics including the code of the Canadian Medical Association.<sup>33</sup>

The theoretical approach followed in this thesis (Chapter One) attempts to find equilibrium amongst competing duties by reflecting on cases, background theories, principles and considered judgements. Thus these duties are considered as *prima facie binding*; that is, they are morally obligatory unless overruled by another *prima facie* principle. *Prima facie* duties are not *absolutely* binding; that is, they are not duties to be followed always without exception,

because circumstances will arise where two or more duties may be in conflict. However, they are not rules that can be discarded if adherence to them does not produce the desired end but are rather *a priori* binding unless overridden by another duty. The duties can be said to be derived from our (society's) collective considered judgements, but need to be specified for particular cases. The "case" considered here is LTV decisions for patients with DMD. I turn now to a discussion of the meaning and scope of these duties and the normative implications for informed decision making.

### *Autonomy*

In a medical context, respect for autonomy grants primacy to the individual's unique ability to determine for his/herself what medical interventions are appropriate to his/her situation.<sup>48</sup> For Mill, autonomy translated into the right not to be interfered with while pursuing one's happiness. According to Kant, autonomy referred to the individual's ability to engage in rational deliberation free of any non-rational influences. When a person behaves according to universalizable rules that are based on reason alone, s/he can be considered to have autonomous will. Persons are not considered autonomous if their actions are guided by coercive force. These heteronomous influences include not only external coercive forces but also internal influences such as emotions and desires. Kant's version of autonomy differs significantly from at least one of its modern conceptualizations.

According to Beauchamp and Childress, autonomy consists of "personal rule of the self which is free from both the controlling interferences of others and from personal limitations that prevent meaningful choice."<sup>49</sup> Traces of Kant's version of autonomy can be seen here in that autonomy must be free of controlling influences, but Beauchamp and Childress' view does not

regard desires and emotions as limiting self determination. Furthermore, Kant would not accept this version because it is not based on pure reason and it does not provide one ultimate source of morality. The influence of Mill's view of autonomy is seen quite clearly in the modern definition, but does not wholly account for it. Contemporary writers in bioethics recognize that the function of autonomy in a medical context is not simply for doctors not to interfere with patients' choices, but rather to share their power thus enabling patients to be full (or at least fuller) partners in their health care and decision making.<sup>42</sup> I will return to this notion again later on.

The duty to promote a patient's autonomy will demand both acts of commission (for example, providing information) or omission (refraining from manipulating patients' decisions) in order to foster self determination. Beauchamp and Childress' interpretation provides a working definition of autonomy that will require further specification when considered alongside of other duties and in the context of LTV decisions. The normative implications of autonomy for informed consent are considered as an initial step in this process.

Clinicians often assert that autonomy and its derivative, informed consent, represent ideals that cannot be practically achieved.<sup>50</sup> Faden and others address these concerns by making important distinctions between autonomous persons and autonomous actions.<sup>51,52,53</sup> Having the capacity for self governance does not guarantee that this capacity has been exercised or that the conditions existed which enabled one to choose autonomously. For a choice to be autonomous it must be intentional, chosen with understanding and free from controlling influences.<sup>54</sup> In addition, an autonomous action can be said to lie on a continuum between *fully autonomous* actions and *fully nonautonomous* actions.<sup>55</sup> Faden argues that the doctrine of informed consent should not be rejected based on the argument that it is impossible for patients to make fully

autonomous decisions or to ever be completely free from the influence of others. Fully autonomous decision making represents an ideal that is seldom if ever possible to achieve. The goal of informed consent is thus better described as *substantially* autonomous decision making, with substantially autonomous decision making resting near the *fully autonomous* end of the continuum.

### *Protection of the Vulnerable*

Protection of the vulnerable demands that one must respect the inherent worth of all persons, even those whose autonomy may be diminished or absent.<sup>56,57</sup> For example, nonautonomous individuals, such as unconscious patients, receive the respect that is due to them when they are not treated merely as a means to an end (e.g., as research subjects without regard for their wellbeing) and when surrogates acting in their best interests make decisions on their behalf. While it is agreed that protection of the vulnerable is a central duty inherent to the principle of respect for persons, how this is best achieved or who need be considered vulnerable is the subject of debate. For example, Pellegrino and Thomasma emphasize the potential vulnerability of the *majority* of ill persons who experience fear, pain, stress, and the effects of treatment or disease.<sup>58</sup> They assert that a physician is justified in protecting a patient by overriding his/her autonomy if the patient is making clearly harmful choices<sup>59</sup> (This is a form of “strong paternalism” that I later discuss and reject ). Others equate respect for persons with respect for autonomy. On this view, protection of the vulnerable is limited to those substantially nonautonomous individuals who do not have the physical or cognitive capacity to participate in decision making (e.g., the unconscious patient, the newborn). These persons have their autonomy protected by having third parties act on their behalf.<sup>60</sup> Children and adolescents are said to have

developing autonomy and it is not readily apparent the extent to which their autonomous choices should be honoured or how much protection they require. These issues are addressed later in the normative analysis under the heading of *family-centred decision making*.

Who counts as vulnerable and in need of protection, and what kinds of protection are demanded, are difficult questions to answer. In a sense all persons are “vulnerable” in that the all of us are dependent, in some way, on others to function and thrive. In addition, all persons are governed by the elements of their internal makeup: their level of cognitive functioning, affect and conation. The actions demanded by a duty to protect the vulnerable will be specified differently depending on the nature and source of the vulnerability and the circumstances of the case. If, for example, a patient is vulnerable because of diminished autonomy, the appropriate action may be to appoint a surrogate to act as a substitute decision maker. Autonomous patients may also be considered vulnerable in that they are vulnerable to the power the physician holds within a patient-physician relationship. In this situation what is called for (I will argue) is a sharing of power through such techniques as disclosure and ongoing conversation structured to promote values sharing and clarification.

### *Beneficence*

Pellegrino and Thomasma and others have criticised the central role of autonomy as a guiding principle in the patient-physician relationship and decision making.<sup>61,62,63,64</sup> They argue that even substantially autonomous decision making is difficult to achieve and that physicians have other duties to their patients beyond a respect for autonomy. Central to the physician’s role is the principal of *beneficence*, wherein the physician is morally obligated to do what he/she can to positively influence the patient's health. Beneficence refers to a "moral obligation to act for the

benefit of others."<sup>65</sup> Frankena derives four obligations out of the principle of beneficence: 1) One ought not inflict evil or harm, 2) One ought to prevent evil or harm, 3) One ought to remove evil or harm and 4) One ought to do or promote good.<sup>66</sup> Historically, health care is based on a principle of beneficence: to help, or at least not harm, the sick and suffering.<sup>67</sup> According to Beauchamp and Childress, the physician's primary obligation has always been to act for the patient's benefit, not to support autonomous decision making.<sup>68</sup> Attempting to honour both of these duties can sometimes set up a situation of moral conflict for the physician whereby in order to respect the patient's autonomous wishes, s/he may be required to participate in care that s/he believes is not the optimum choice for the patient. This conflict is sometimes described as a central tension between two major competing moral visions of medicine: autonomy on one side and beneficence on the other.<sup>64,68,69</sup> This tension may be eased by an interpretation of the principle of beneficence that incorporates patient autonomy. In essence, such views claim that promoting autonomous choice serves the best interests of the patient by promoting his/her psychological wellbeing and healing. Furthermore, autonomy serves beneficence because patients generally know what is best for them.<sup>70,71,72</sup> Beauchamp and Childress criticise this view as "a dressed up defense of the autonomy model." Instead, they claim that beneficence "provides the primary goal and rationale of medicine and health care, whereas respect for autonomy... sets moral limits on the professional's actions in pursuit of this goal."<sup>73</sup> Churchill<sup>74</sup> asserts that the obligation to act beneficently is necessarily modified by a respect for autonomy which demands that the patient's, not the physician's notion of the good is definitive.

Honouring both autonomy and beneficence is a difficult task and represents an ongoing challenge for scholars and clinicians. While the idea of informed consent is supported by legal, professional and ethical sources, how best to translate this idea in to action remains the subject of

ongoing debate and discussion. The consent process is embedded in the patient-physician relationship with its inherent duties and obligations which may at times come into conflict. An examination of this relationship provides a richer description of the complexity of the decision making process.

## **The physician-patient relationship**

The process of decision making occurs within the context of the physician-patient relationship. Conditions such as the degree of trust between the patient and physician, the power dynamics within the relationship, and the individual approach to information sharing will serve to aid or hinder the decision making process. Several models of the patient-physician relationship appear in the literature. I have grouped them into the following three general models, each of which places a different emphasis on the principles of autonomy and beneficence: *paternalism* emphasizes beneficence towards patients, *consumerism* emphasizes patient autonomy and *shared decision making* attempts to balance the two principles.

### *Paternalism*

As Churchill points out, the history of medical ethics until recently is a history of paternalistic beneficence.<sup>74</sup> Only in the last thirty years has the concept of autonomy modified the physician's approach to the care of patients. Despite the recent criticisms of medical paternalism in the patient-physician relationship, some forms of paternalism are still prominent in modern medicine.

Paternalism is defined as " the intentional overriding of one person's known preferences by another person, where the person who overrides justifies the action by the goal of benefiting the person whose will is overridden."<sup>75</sup> In 1951, Parsons<sup>76</sup> described the physician-patient relationship as the functional interplay between two social roles with inherent duties and responsibilities. The patient temporarily takes on a "sick role" in which s/he is temporarily

exempt from the normal duties of life but is expected to comply with prescribed treatment. The physician's therapeutic role maintains a detachment within the relationship and through positive or negative reinforcements influenced the patient's behaviours to promote recovery.<sup>77</sup> The relationship as Parsons originally described it\* is often compared to one of parent and child, where the patient is temporarily exempt from his/her social responsibilities while s/he is being cared for.<sup>26</sup> While the patient always has the option to refuse treatment, s/he is not involved in the process of choosing amongst treatment options. The physician determines unilaterally the preferred option and then presents it to the patient. This paternalistic vision of the relationship is consistent with a beneficent approach to decision making untempered by an obligation to promote autonomy. The good of the patient is defined according to the perspective of medicine rather than the perspective of the patient.<sup>48</sup>

Two forms of paternalism have been described: strong and weak.<sup>78,79,80</sup> Strong paternalism consists of overriding the choices of another even though those choices are informed, voluntary and autonomous. Parsons' early view of the patient-physician relationship is consistent with strong paternalism. This model is now widely criticised and no longer considered acceptable when patients have the capacity to participate in decision about their care.<sup>80,81,82,83</sup> Strong paternalism effectively eliminates the type of decision making envisioned by the idea of informed consent, which emphasizes autonomy, not only best interests.<sup>26</sup>

---

\* Parsons later revised his description of the patient-physician relationship to acknowledge the patient's participation in decision making in the context of chronic illness.( Parsons T (1975) The sick role and the role of the physician reconsidered. *Millbank Memorial Fund Quarterly* 53 (3), pp. 257-278.)

Weak paternalism involves an action taken on the grounds of beneficence for a patient whose autonomy is substantially diminished. For example, patients with decreased levels of consciousness, severely impaired cognitive function, or those experiencing severe acute pain experience significantly compromised abilities to choose autonomously. In such cases, the physician may decide in advance which treatment s/he considers to be in the patient's best interests and present only that option. Alternatively, s/he may frame the information (present it in a biased fashion) such that this is the only choice the patient can make.<sup>84</sup> Overriding the wishes of a substantially nonautonomous patient in the interest of the patient's welfare is justified according to the standards of weak paternalism. This limited form of paternalism is widely accepted in law, medicine and ethics.<sup>75</sup>

### *Consumerism*

Paternalism, if and when it is considered justified, is grounded in the principle of beneficence. In stark contrast to this approach is the consumer model of the patient-physician relationship which is grounded almost exclusively in the principle of autonomy. This model, also called the engineering model or informative model,<sup>85</sup> firmly places decision making control exclusively in the hands of the patient.<sup>86</sup> The consumer model views physicians as service providers: the doctor outlines relevant information to the patient, the patient chooses the medical intervention s/he wants, and the doctor executes the chosen option. The consumer approach to medical care grew out of a growing resistance to medical paternalism. The origins of this resistance parallel other concurrent social movements in North America: the growing political emphasis on individual rights, the civil rights movement of the sixties and the general rise of consumerism in public life.<sup>87,88</sup>

Within this model a fairly clear distinction between facts and values is assumed. The patient's values are thought to be well defined and known; what s/he lacks are the facts. The physician's responsibility is to provide the facts and do what is asked. The physician's personal value judgements regarding the best course of care are not considered relevant. Even when the chosen intervention is against the better judgement of the physician, the physician still has a duty to perform it or else refer the patient elsewhere. There is little role for the physician's values in the process of decision making.<sup>85,89</sup>

The consumer model is in keeping with the basic philosophy of the Independent Living Movement and is often advocated for persons with disabilities and chronic illness.<sup>88</sup> Canadian disability organisations representing national, regional and local interests recently re-affirmed their commitment to the "Independent Living philosophy."<sup>90</sup> The movement views disability from a different perspective than the traditional medical model. Persons with disability should no longer be viewed as objects of pity that need the help of the medical professionals or government agencies. Instead, the environmental factors (structural, institutional and attitudinal) that limit the achievement of independent living are viewed as the locus of disability. Thus the movement advocates that persons with disabilities reclaim their lives in opposition to state controlled institutions.

Two main criticisms of the consumer model are found in the literature. First is the assumption that there exists a body of value-free medical facts to which all patients have equal access. Second is an underlying assumption that the "consumer" is already in a position of equal power with the physician. Each is considered in turn.

A consumer model assumes that patients are aware of their values and that all they need are the facts in order to choose a treatment.<sup>85</sup> This confines the physician to dealing with the facts

but in actuality this is unrealistic. Doctors make value judgements all the time when they assess the risks of a procedure, interpret the clinical applications of medical research, or decide what is "significant" and what is "valuable" in treatment, results, risks and benefits.<sup>89</sup> The medical facts are always shaped by the physician's values, and therefore those values can never be completely removed from the medical encounter. Thus the practice of medicine is not value-free but value-laden.<sup>91</sup> At the same time the consumer does not always have well defined, fixed values and needs. In reality, people are often uncertain about what they want. Part of being an autonomous individual involves having the capacity to reflect on one's wishes and revise desires and preferences.<sup>85</sup> This will require a process of deliberation that entails a review of medical knowledge as it specifically relates to one's own situation. Such deliberation will be enhanced through discussion with another. It seems logical that the physician should be at least one of the individuals with whom the patient discusses options. The physician with his/her medical knowledge and experience can help to assimilate the patient's personal viewpoint with the available options. For fully informed consent to take place, clinicians have a responsibility to inform the patient of their opinion of what the best course of treatment is and why.<sup>38,92</sup> According to Hayry,

...concern for the patient's right to self-determination does not imply that the physician should psychologically abandon the patient: a doctor who merely spreads the options in front of the patient and then says 'go ahead and choose, its your life' is not anti-paternalistic but negligent. Genuine respect for autonomy requires that doctors make sure their patients understand the options open to them....<sup>93</sup>

Here Hayry reinforces the notion of optimizing autonomous choice through some form of shared decision making. If reasonable *understanding* is an element of informed consent, then it is enhanced through a model of decision making that involves ongoing sharing and clarifying of information.

The second criticism of the consumer model is that it already assumes that patients are in a position of equal power<sup>94</sup> with the physician.<sup>95</sup> It cannot be ignored that, at the outset, the physician possesses the power of knowledge over the patient. The patient often may not know where to begin to ask questions about possible options and outcomes. In this sense the model is easily open to abuse. The model presupposes the figure of the well educated, assertive consumer speaking to the doctor as an equal and carrying on a sophisticated conversation about medical options. While it is true that this situation can and does exist, it does not take into account disempowered patients, such as those who are elderly, uneducated, or from the lower socioeconomic classes. As Williams states in his critique of the independent living movement:

...there is plenty of evidence to suggest that those who most need help and services are the least effective in claiming and using them. Where the supply depends on effective market-place shopping, these people are likely to find themselves especially disadvantaged. ... a major feature of the social reality of disablement is the elderly female living... in an urban slum....<sup>95</sup>

Thus while the consumer model of decision making is worthy in its pursuit of respecting the patient's right to self determination, there exist at least two difficulties with the model that make it unsatisfactory for the majority of patients. Some evidence suggests that those who advocate a consumer model or an "independent living philosophy" are not advocating an abandonment of the physician's considered judgements but rather a sharing of power that moves away from a traditional paternalistic approach.<sup>96</sup> If this is so, then the consumer model begins to move away from its emphasis on autonomy to more closely resemble a shared decision making approach.

### *Shared decision making*

Shared decision making, as I have noted earlier, is sometimes equated with a broad definition of informed consent. This approach to medical decision making represents a middle ground between paternalism and consumerism which attempts to balance the values of beneficence and autonomy. Some versions of shared decision making accommodate the duty of protection of the vulnerable with a recognition of three internal sources of vulnerability: cognition, affect (emotions) and conation (will). Sharing involves not only sharing of information but sharing of values and emotions and willingness to act.

Several sources agree that mutual participation addresses both the duty to act in the patient's best interests and the duty to promote autonomous decision making.<sup>34,38,85,89,97</sup> The President's Commission for the Study of Ethical Problems bases consent on two values: "promotion of a patient's well-being" and "respect for a patient's self-determination."<sup>34</sup> Lidz<sup>26</sup> and others<sup>98</sup> assert that informed consent should not be an event but rather a process, recognizing that decisions are rarely made at one point in time. The advantage of ongoing dialogue is that it affords patient and physician the opportunity to share expertise. The physician's expertise lies in his/her medical knowledge while the patient's expertise lies in knowledge of the contextual facts of his own personal situation including his values, aspirations, and available supports. The physician's role in shared decision making is to assist the individual to make the choice that is most appropriate by fostering the patient's understanding of how the medical information relates to his/her unique perspective and situation. This is addressed through ongoing dialogue.<sup>26,38</sup>

Mutual participation may be difficult to achieve in practice. Power inequalities inherent in the relationship may hinder autonomous choice.<sup>99,100</sup> There may also be ethnic, class, economic and value differences between patients and physicians which undermine the assumption of a

commonality of interests between the two.<sup>89,101</sup> Genuine attempts at mutual participation can therefore easily be reduced to situations of paternalism.<sup>85</sup> The physician may unwittingly impose his/her own values onto the patient and the uncertain patient may too easily accept this imposition.

However, these criticisms do not necessarily support outright rejection of mutual participation. Several authors have outlined different models of shared decision making, each of which includes its own strategies for promoting patient participation and avoiding paternalism. Some prominent examples include the contractual model as outlined by Veatch,<sup>89</sup> Emanuel and Emanuel's deliberative model,<sup>85</sup> and the conversation model proposed by Katz.<sup>38</sup> An approach specific to LTV decisions for persons with DMD that incorporates Katz's model is defended in the normative analysis.

Empirical evidence suggests that shared decision making is not the norm in medical practice and elements of paternalism remain strong. Clinicians and particularly physicians, continue to make recommendations that patients follow without question.

Beausoleil et al.<sup>102</sup> studied the effects of education and experience on ethical attitudes of physicians in a U.S. neonatal intensive care. A survey was administered to medical students, residents and neonatologists. As part of the survey, the participants were asked to rate the relative importance of the parents, the paediatrician, the neonatologist, the ethics committee and the court in making "level of care" (treatment) decisions. The neonatologist was rated as the most important decision maker by the neonatologists and the residents, while the medical students believed the parents were most important.<sup>102</sup> In this study, physicians still believed they were the primary decision makers. Perry found similar results with a diverse group of health care professionals. When 143 medical residents, nurses, psychiatrists and psychiatric nurses were

asked who should make Do-Not -Resuscitate decisions, 32% said the physician, 40% said the patient, 20% said jointly by the physician and the patient and 8 percent said the family.<sup>103</sup>

In another study, Sullivan et al.<sup>104</sup> studied respirologists' approaches to mechanical ventilation decisions for patients with end-stage chronic lung disease. Through the analysis of fifteen qualitative interviews, they found that while the physicians advocated a shared decision making approach, they admitted to intentionally modifying information in order to influence the patient's choice. The authors concluded that the physicians followed a "physician driven" model of decision making rather than a shared decision making model. Miller and Lo<sup>105</sup> obtained similar findings. They observed 15 physicians discussing cardiopulmonary resuscitation with patients acting in a hypothetical encounter. Many of the physicians admitted to tailoring their discussions to "fit" the patient and approximately half admitted they did so to deliberately try to influence the patients' decisions.

Lidz et al.<sup>106</sup> conducted a qualitative inquiry using participant observation (on two inpatient wards and one surgical outpatient clinic) to record patient-staff interactions related to the decision making process. They supplemented their observations with semi-structured patient interviews. The authors found that, while patients did report wanting information about their treatments, less than 10% actually used that information to make a decision:

...patients believe that decisions about their treatment should be primarily or completely up to their physicians because of their technical expertise and commitment to the best interests of the patient.<sup>106</sup>

In addition, physicians did not normally view decision making as a patient's choice among alternative treatments. They usually told the patient what was going to be done, believing that they were in the best position to determine the superior treatment.

The results of these studies demonstrate the importance of examining the decision making approaches of clinicians. Physicians still retain much of the power in decision making and do not necessarily encourage mutual participation.

## Quality of Life and Attitudes Towards Disability

### *Quality of life*

Quality of life has been equated with personal happiness, well-being, satisfaction with life and contentment. Despite its common usage, the term remains difficult to define. According to Walter,<sup>107</sup> the word "life" can refer to either simple *biological life* or *personal life* which includes biological life but also other important capacities such as self awareness. "Quality" can possibly refer to excellence or be a property of biological or personal life. Some scholars define "quality of life" as relating to a single property. As an example, Walter refers to Richard McCormick's early view that the potential for human relationships is the minimum necessary for personal life. Others refer to several properties of physical, cognitive and social functions that jointly considered define one's quality of life.<sup>19,108</sup>

Faden and Leplège<sup>19</sup> point out two aspects of quality of life that are for the most part uncontested. The first is biologic life and the second is the minimum cognitive apparatus needed to attach some meaning to the biologic life. They also hold that there exists general agreement about the positive value of certain human experiences and capabilities such as mobility, being free of pain, being loved and having peace of mind. Important to defining individual quality of life, they say, is the understanding that individuals will vary as to the relative value they attach to each. This is of particular importance when individuals face a "trading-off" process in their lives, for example accepting some pain or discomfort in order to increase mobility.

Musschenga<sup>109</sup> observes that the overall purpose of evaluating quality of life in medical care is to attend to outcomes other than the biological when assessing the success or failure of treatment. Medical management of complex diseases (e.g., cancer, cardiovascular disease,

neuromuscular disease, and arthritis) rarely results in a complete restoration of health. Instead the effects of treatment can be partial, uncertain and/or temporary. "Quality of life" serves as a catch-all term for a diverse set of criteria that need to be considered beyond simple survival time.

Musschenga's conceptualization of "happiness," provides a working framework for an examination of the potential problems inherent in an assessment of third party quality of life. He contends that either subjective or objective measures may be used to measure quality of life depending on the purpose of evaluation. To distinguish these, he outlines three general *components* of happiness: enjoyment (positive mental states), satisfaction (the individual's evaluation of success in attaining life's goals) and excellence (the virtuousness or value of a person's activities, which can be further divided into self-assessed and ascribed). Each of these components is dependent upon three *conditions*: good fortune, material resources and person-dependent conditions (the individual's physical, cognitive and emotional capabilities). Musschenga argues that enjoyment and satisfaction are by their nature subjective and can only be measured definitively by the individual. Objective measures of quality of life are only appropriate when assessing the degree of normal physical or mental functioning, that is the person-dependent conditions for happiness.<sup>110</sup> What Musschenga is essentially saying is that objective measures may be used to assess a patient's level of physical or mental disability, but only the patient can tell us whether or not s/he enjoys or is satisfied with his/her life.

---

<sup>110</sup> Musschenga does not define precisely what he means by "objective assessment." My interpretation is that he is referring to standardized measures of function that are assessed by third parties. An example might be a six minute walk that assesses level of fitness. This test is carried out and scored by a third party rather than asking the patient his subjective response to "are you fit?" Musschenga's view is that this objective test would not measure quality of life per se but one of the person-dependent conditions that impact on quality of life.

“Quality of life” is thus a complex concept. Some elements of an individual’s quality of life are amenable to objective tests but other elements can only be evaluated by the individual. Because of this, it is not possible for third parties to make definitive unilateral judgements about an individual’s *overall* quality of life. When physicians attempt to make quality of life assessments that go beyond simply assessing a patient’s cognitive or physical function, their judgements will contain a subjective, value laden component that may vary from physician to physician. Health care professionals do not commonly use the term “quality of life” when simply referring to the results of objective tests of function. For example, if a patient demonstrates a forced vital capacity of 40% predicted, we do not say s/he has a poor quality of life, we say s/he has impaired lung function. Quality of life judgements refer to a subjective interpretation of those findings: “I think s/he must have diminished quality of life because of her impaired lung function.” However, a complete and valid assessment of a person’s quality of life would require the subjective input of that person.

In cases where the patient is incapable of participating in care decisions (e.g., an unconscious patient or a disabled newborn), whether or not it is permissible for physicians to base treatment decisions on unilateral assessments of the patient’s overall quality of life remains in dispute. There seems to be general agreement, however, that as long as persons are capable of judging the quality of their own lives, then judgements from third parties are neither required or desirable.<sup>111,112</sup> Hayry asserts that only when patients are unconscious or otherwise cognitively incapable are doctors or others justified in judging the quality of a patient's life. The demands made by autonomy make it suspect that physicians could legitimately decide about the treatment of competent patients based on a third party assessment of quality of life. Faden and Leplège agree with this position.

..the quality of any person's life is in important respects unique and personal to that individual. The impact of any event or health state on a person's quality of life is to a significant degree contingent on his or her personal projects, desires and history<sup>19</sup>

Despite these assertions, empirical evidence suggests that health care professionals may base treatment decisions on mistaken perceptions of the quality of life of their patients. Gerhart et al.<sup>113</sup> surveyed emergency care providers' (nurses, physicians and emergency technicians) attitudes toward quality of life after spinal cord injury. They compared their results to previously reported quality of life ratings of high level spinal cord injury survivors. Forty-five percent of the emergency care providers felt that if they personally sustained a spinal cord injury, they would want either nothing done or pain relief only. Only eighteen percent imagined they would be "glad to be alive," compared with ninety-two percent of the actual survivors. The investigators concluded that health care providers can influence patient and family decision making and therefore must be well apprised of "outcomes, well-being and life satisfaction following severe spinal cord injury." Schneiderman et al.<sup>114</sup> conducted a study that compared 1) patients' preferences for specific life saving procedures with physicians' predictions of the patients' preferences and 2) physician's preferences for themselves if they were in the same situation. They found that physicians often underestimate their patients' perceived quality of life. The study also demonstrated that physicians' predictions of what lifesaving procedures their patients would want more closely corresponded to their own personal preferences than the patient's actual preferences.

Professionals working with muscular dystrophy patients often refer to poor quality of life when justifying their reasons for not recommending ventilation.<sup>5</sup> Two studies by Bach demonstrate this point. In the first study,<sup>6</sup> the researchers surveyed health care professionals and muscular dystrophy clients receiving ventilation. Each group was asked to rate their own quality

of life. In addition, the health care professionals were asked to rate the quality of life of the individuals with muscular dystrophy. The professionals predicted that the disabled individuals would in general report having a poor quality of life. However, the disabled individuals on average reported having close to as good a quality of life as the professionals. The professionals had significantly underestimated the disabled individual's quality of life.

The second study (Bach 1992)<sup>4</sup> has been discussed in part above. An additional component to this study asked all clinic directors to estimate the satisfaction with life of muscular dystrophy patients receiving ventilation. Eighty ventilated patients were given the same life satisfaction survey to complete. The study revealed that:

The clinic directors significantly underestimated the users' reported life satisfaction. The directors who discouraged ventilator use more significantly underestimated the users' life satisfaction than those who recommend it. We conclude that the MDA clinic directors' estimation of ventilator users' quality of life and satisfaction with life is positively correlated with the likelihood of their discussing and recommending ventilator use to prolong life.

These studies indicate that, in practice, clinicians both underestimate the quality of life of their patients and may use these inaccurate assessments to formulate recommendations. Quality of life judgements are sometimes made unilaterally without consultation with competent patients.

### *Attitudes Towards Disability*

The empirical research described above also suggests that quality of life judgements and attitudes towards disability can be closely related. Further evidence of this claim is found in an examination of the case of Elizabeth Bouvia, a young woman with a severe physical disability secondary to cerebral palsy who petitioned a California court to mandate that the Riverside County Hospital cooperate with her plan to starve herself to death. The American Civil Liberties

Union, acting on Ms. Bouvia's behalf, wrote a brief that described her disability as "incurable and intolerable" and commented on "the indignity and humiliation of requiring someone to attend to her every bodily need".<sup>115</sup> The superior court of California in its judgement claimed "She as a patient lying helpless in her bed, unable to care for herself, may consider her existence meaningless." While Ms. Bouvia did express a desire to end her life, there were numerous other factors that contributed to her situation, including the recent death of a sibling, financial hardship, a dissolution of marriage and a miscarriage. The statements of the ACLU and the court suggest that something inherent in the disability itself, not Ms. Bouvia's particular situation, make her life "intolerable" and "meaningless". The underlying assumption is that a person with a severe disability such as hers could not possibly (or is at least is not likely to) enjoy a reasonably good quality of life.

Hahn suggests that two critical values prevalent in North American society contribute to negative attitudes towards persons with disability: personal appearance and autonomy. Building on the work of Goffman, he suggests that persons who fail to meet the prevailing standards of physical attractiveness and functional independence are assumed to be inferior.

Like other minorities who have been victims of discrimination, disabled persons have characteristics that permit them to be differentiated from the rest of the population. These characteristics, which may be identified by physical or behavioral clues or by verbal labels, are likely to arouse strong feelings in nondisabled observers about their own appearance or autonomy.<sup>116</sup>

The nondisabled person's unspoken reaction may be "I wouldn't want to live like that". Disability and poor quality of life become equated; disability is assumed to lead to a poor quality of life. Persons with disabilities are then viewed as objects of pity rather than as equal, valued members of the human community. When negative attitudes are held by health care professionals, these views can shape which treatment options are discussed with patients or how information is

framed. The empirical evidence presented above demonstrates that health care professionals, including physicians, may have negative attitudes towards disability which may bias their care and treatment recommendations.

## Summary of Literature Review

Bach's American survey of Muscular Dystrophy Association clinic directors raised concerns that some physicians are restricting access to ventilation for patients with DMD based on unilateral judgements of quality of life and the perceived burden to the family. Such practice raises ethical concerns that drive this thesis. Decision making is considered in relation to the physician's moral obligations to the patient. The patient with DMD is considered as part of a family unit, the members of which are interconnected and share common goals. The family is not considered as a separate source of the physician's moral obligations, and thus burdens to the family are considered as affecting the family unit as a whole, rather than as a potential source of conflict within the family (see Chapter 5).

The process of informed decision making is grounded in three prima facie duties inherent to the practice of medicine. These are autonomy, beneficence and protection of the vulnerable. These duties flow from an overarching principle of respect for persons which is part of the common morality and rooted in both the deontological and utilitarian traditions. The normative implications of these duties are sometimes unclear and require specification for given circumstances or groups of cases. A richer description of the process of informed decision making acknowledges that decisions are made within the context of the patient-physician relationship. Different models of the relationship place different emphasis on each of the physician's duties. A model of shared decision making is held to be the preferable model for the majority of cases because it addresses the normative requirements of autonomy, beneficence and protection of the vulnerable without sacrificing one to the other(s). Relationships that support

mutual participation in decision making will involve a sharing of information over time so that informed consent becomes a process rather than an event. While much of the literature supports shared decision making, the empirical evidence indicates that elements of paternalism remain strong.

Health care professionals' attitudes towards disability may influence the types of recommendations they make to their patients. It has been shown that clinicians often underestimate the quality of life of individuals with disabilities but may refer to poor quality of life when justifying their decisions. Physicians' choice of recommending or discouraging ventilation for patients with neuromuscular conditions may be influenced by their attitudes towards disability. Quality of life includes the subjective components of "enjoyment" and "satisfaction with life" that resist assessment by third parties. A shared decision making model emphasises sharing of information and values such that quality of life judgements would be shared and explored together rather determined unilaterally by physicians.

To date there has been no published literature examining the issue of accessibility to long term ventilation in the Canadian context. Differences in the health care systems and cultures between Canada and the United States do not allow for a generalizability of Bach's American study. Reasons that deal with cost may be less prominent in Canada where LTV is covered by public health insurance. In addition, seven years have passed since Bach's original study was conducted. Even if Canadian and American practices were comparable, enough time has passed that practices and attitudes may have changed significantly.

## **Chapter 3: Empirical Methodology**

### **Overview of empirical methodology**

A two-part empirical study was conducted to determine the practices and attitudes of Canadian physicians towards long term ventilation as a treatment option for patients with Duchenne muscular dystrophy. Part one consisted of a self-administered questionnaire sent to all physicians who follow DMD patients through neuromuscular clinics in Canada. The purpose of the questionnaire was to provide a breadth of information regarding practices and attitudes. Part two consisted of face to face interviews with six of the physicians who responded to the questionnaire. The interviews were conducted to obtain a richer description of practice that was not possible with the questionnaire.

### **Part One: Mail Questionnaire**

#### *Population*

The target population consisted of all physicians who were directors, co-directors or respirologists associated with a Canadian neuromuscular clinic. Potential respondents were identified through provincial Muscular Dystrophy Association offices. Physicians were included in the study if they follow any patients with DMD. Each identified physician was mailed a questionnaire to complete.

### *Questionnaire Development*

The questionnaire consisted of 67 closed-ended questions (appendix 1). The questions were developed based on the primary investigator's clinical experience, the relevant literature (notably the study by Bach<sup>4</sup> previously referred to), and three initial pilot interviews with physicians who follow patients with DMD. The draft questionnaire was then pilot tested on three additional physicians. A French version was also produced. The translation followed the procedures as outlined by Guillemin et. al.,<sup>117</sup> as follows. An initial professional translation was tested for semantic and idiomatic equivalence with two bilingual clinicians. The translation was then back-translated into English by a professional translator whose first language is English, to check for consistency. Finally, the corrected French version was checked by a bilingual physician who works in the field of neuromuscular diseases.

The survey included the Modified Issues in Disability Scale (MIDS) developed and previously validated by Makas,<sup>118,119</sup> which measures attitudes towards disability. The MIDS is a 37 item Likert-type scale in which subjects are asked to indicate their extent of agreement or disagreement with statements about persons with disabilities. This scale was chosen because it has been tested for reliability, has strong content validity and is easy to administer and score. The MIDS is also a unique scale in that persons with disabilities were actively involved in the scale construction/validation process.

### *Data Collection*

The survey was administered according to a modified Dillman procedure<sup>120</sup> which emphasizes a personalized approach followed by multiple mailings. At the initial mailing, each

potential respondent was sent a personalized cover letter (appendix two) with the questionnaire along with a stamped envelope for returning the survey. A reminder letter was sent three weeks later, followed by a second complete mailing three weeks after this. Finally, respondents who had not returned questionnaires were contacted by telephone. An honorarium of twenty dollars was offered to the physicians for completion of the questionnaire.

Because the survey was self-administered, respondents indicated their consent by completing and returning the survey. Respondents were advised that the information they provided is kept completely confidential.

### *Data Analysis*

The Statistical Package for the Social Sciences (SPSS)<sup>121</sup> computer based program was used for the analysis. Descriptive statistics (frequency distribution, range, means and standard deviations as appropriate) were generated for all variables. Bivariate analysis was conducted to determine possible associations between MIDS scores and the following variables: gender, age, years working in a neuromuscular clinic, clinical specialty, opinions on resource allocation, and disclosure practice. Bivariate analysis was conducted to determine possible associations between disclosure practice and the following variables: gender, age, number of children, years working in a neuromuscular clinic, clinical specialty, and MIDS score.

### **Part Two: Interviews**

Further exploration of physicians approach to LTV decisions was addressed using qualitative methods. Face to face interviews were conducted with six of the respondents after the questionnaires have been returned and analyzed. The physicians were asked to elaborate on the

decision making process they engaged in with patients and families. They were encouraged to describe actual cases as well as their overall philosophy and clinical approach to management of LTV decisions. The interviews allowed for the probing of specific responses that was not possible on the structured questionnaire, and therefore provide a richer description of the physicians' attitudes and practices.

### *Sample*

A convenience sample of six physicians was chosen from a list of sixteen who volunteered to be interviewed (through returning a postcard supplied with the questionnaire). While they represented a sample of convenience, selective sampling<sup>122</sup> was employed. Respondents were chosen based on four criteria: the respondents represented a range of geographical locations in Canada, the respondents' demographics approximated the demographic profile of the larger study sample, the respondents represented a range of attitudes/practices towards ventilation, and finally the respondents were accessible to the principle investigator.

### *Data collection*

All interviews were conducted by the principal investigator using open-ended semi-structured questions (see appendix 3 for interview guide). The interviews were audiotaped. The tapes were then transcribed by a medical transcriptionist and verified by the principal investigator prior to being used for qualitative analysis.

### *Analysis*

The interview transcripts were imported into the Ethnograph<sup>123</sup> qualitative analysis program. The data were then analyzed for recurring themes using grounded theory techniques as described by Strauss and Corbin<sup>124</sup> and briefly outlined here. Transcripts were segmented into small but meaningful sections of text ranging from 2-8 sentences long. Each section was then assigned one or more descriptive conceptual labels (open coding). All the segmented, labelled transcripts were then compared for similarities. Initial labels were then further refined to establish consistent codes. The purpose of the coding is to generate conceptual categories in the physicians attitudes, practices and decision making processes. Open coding was followed by axial coding which involved re-examining the initial codes to look for relationships between the categories and assigning new codes that consider context, causal conditions, and action/interactional strategies and consequences. Methods of memo writing, constant comparison and continued questioning<sup>125</sup> of the data were employed to conceptualize responses and then identify common themes within individual interviews and between respondents. Producing a complete "grounded theory" of the phenomenon was not the intention of the qualitative inquiry and would not be prudent based on the small number of interviews that were conducted. Analysis is limited to the themes which bear direct relevance the purposes of the study and complement the quantitative and normative inquiry.

### *Normative Evaluation*

The final stage of the analysis is the normative evaluation of the results obtained in the survey and interviews. Concepts and themes that emerge from the data require clarification and evaluation. New themes and concepts, along with material raised in the literature review, are

analyzed with the goal of defending which practices and attitudes are best justified. The methods of philosophical inquiry as outlined above (see Theoretical Approach) are employed.

## **Chapter 4: Results**

### **Part One: Quantitative Description of Practice and Attitudes**

#### **Response rate**

Administration of the questionnaire occurred between the months of February to April of 1997. The questionnaire was sent to all physicians in Canada who follow individuals with DMD through neuromuscular clinics. These individuals were identified through provincial Muscular Dystrophy Association offices. Eighty-three physicians were initially sent a questionnaire. Of these, 23 physicians were excluded from the study because they were no longer practicing in Canada or they were not following patients with DMD, leaving a total of 60 physicians from 34 clinics appropriate for the study. Forty-five questionnaires from 29 clinics were returned for a physician response rate of 75.0% and a clinic response rate of 85.3%.

#### **Demographic and clinical information**

The overall mean age of the respondents was 47.0  $\pm$  8.2 years (females, mean= 43.5 $\pm$  8.2, males, mean= 47.9 $\pm$ 8.1). The majority of the respondents were male (77.3%), married (88.9%) and had one or more children (93.3%). The sample consisted of 14 (31.1%) respirologists, 12 (26.7%) neurologists, 10 (22.2%) paediatricians, 6 (13.3%) physiatrists/rehabilitation specialists and 3 (6.7%) others. Twenty-two (48.9%) respondents worked in paediatric clinics, 13 (28.9%) in adult clinics and 10 (22.2%) in mixed clinics. The mean number of years the physicians had been working in a neuromuscular clinic was 11.61 $\pm$ 6.93 years. The size of the respondents' DMD

patient caseload is summarized in table 1. The majority of the physicians (63.6%) followed between one and 30 patients with DMD.

### **Clinical Practice**

Information was collected regarding the following aspects of the physicians' clinical practice: number of patients using ventilators, types of ventilation in use, disclosure of ventilation information, advice to patients, and practice rationale. The following results were obtained.

#### *Number of patients using ventilators*

Individual physicians were asked to report the number of patients they were following who used ventilatory support. These results are summarized in table 2. It was not possible to establish an accurate picture of the number of patients using ventilators grouped according to clinic because of a number of discrepancies in the reporting of individual physicians working at the same clinic. It seems likely that, in many cases, physicians working at the same clinic may be following different patients. However, it was possible to obtain the following general information. Of the twenty-nine clinics, 21(72.4%) were following patients using ventilators, the other 8 (27.6%) were not following any ventilated patients at the time of the survey. Of the clinics following ventilated patients, four (19.0%) reported only following patients receiving fulltime ventilatory support and an additional four clinics (19.0%) reported only following patients receiving part time ventilation. The remaining thirteen clinics (61.9%) had a mixture of fulltime and part time ventilator users on their caseload.

**Table 1: Number of patients with DMD followed by the physicians**

<b>Total number of patients</b>	<b>n (%)</b>
1 -10	10 (22.7%)
11-20	13 (29.5%)
21-30	5 (11.4%)
31-40	4 (9.1%)
41-50	5 (11.4%)
51-60	2 (4.5%)
>60	5 (11.4%)
	n=44 (100%)

missing cases = 1

**Table 2: Number of ventilated patients followed by the physicians (n=44)**

<b>Number of patients using ventilators</b>	<b>Number of physicians (%)</b>
0	12(26.7%)
1-5	29(64.4%)
6-10	1(2.2%)
11-15	2(4.4%)
>15	1(2.2%)*
	n=44 (100%)

missing cases = 1

median=2, mean=3.8±7.1

\* this respondent reported following 45 ventilated patients

### *Types of ventilation*

The physicians in the survey reported the use of seven different types of positive pressure ventilatory systems with their DMD patients. These are summarized in Figures 1a-c. None reported the use of negative pressure systems. Tracheotomy was the most common method of delivery used for fulltime ventilation. Nasal interfaces using either a bilevel ventilation (BIPAP) or a volume cycled delivery system were the most commonly used for part time ventilation. When part time and fulltime ventilation are considered together, the most common methods used were nasal BIPAP (n=23) and tracheotomy (n=20).

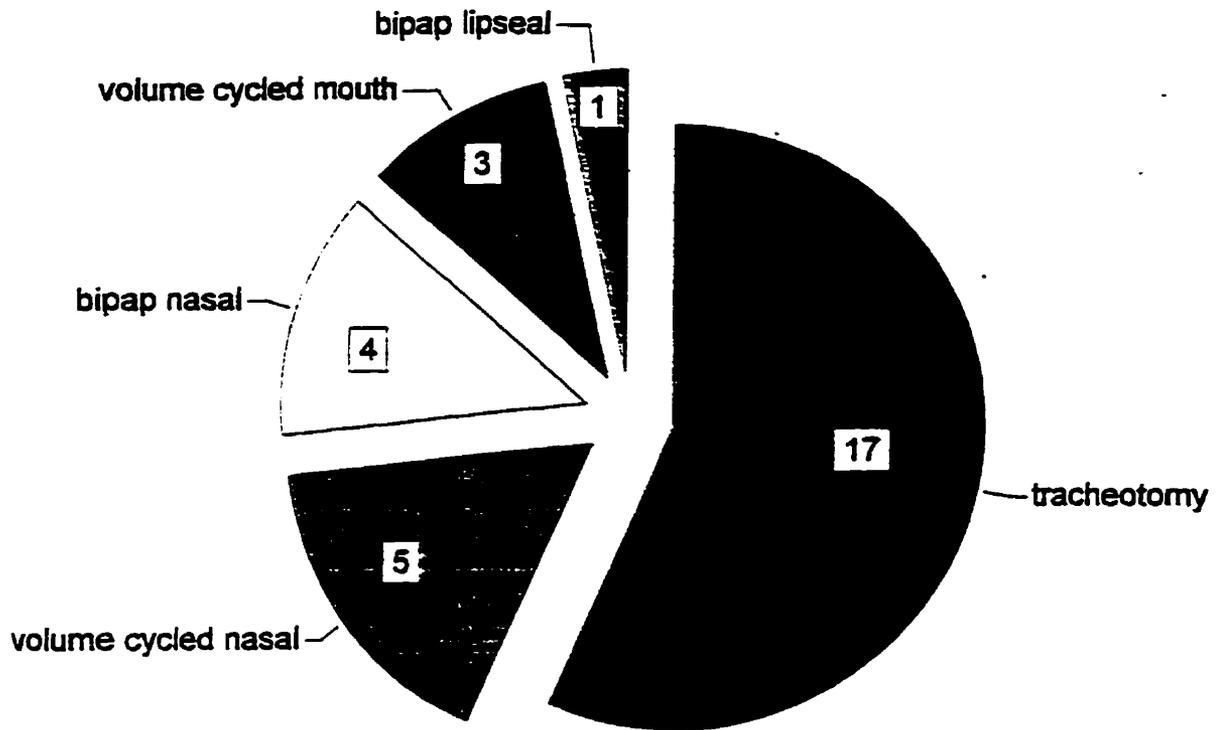
### *Disclosure*

When asked if they informed their DMD patients and families about the option of long term ventilation, of the 44 respondents who answered, 33 (75.0%) said they always inform their patients/families, 10 (22.7%) said they sometimes did not tell patients/families about ventilation and one respondent (2.3%) never informed patients/families about ventilation (Figure 2).

### *Advice to patients*

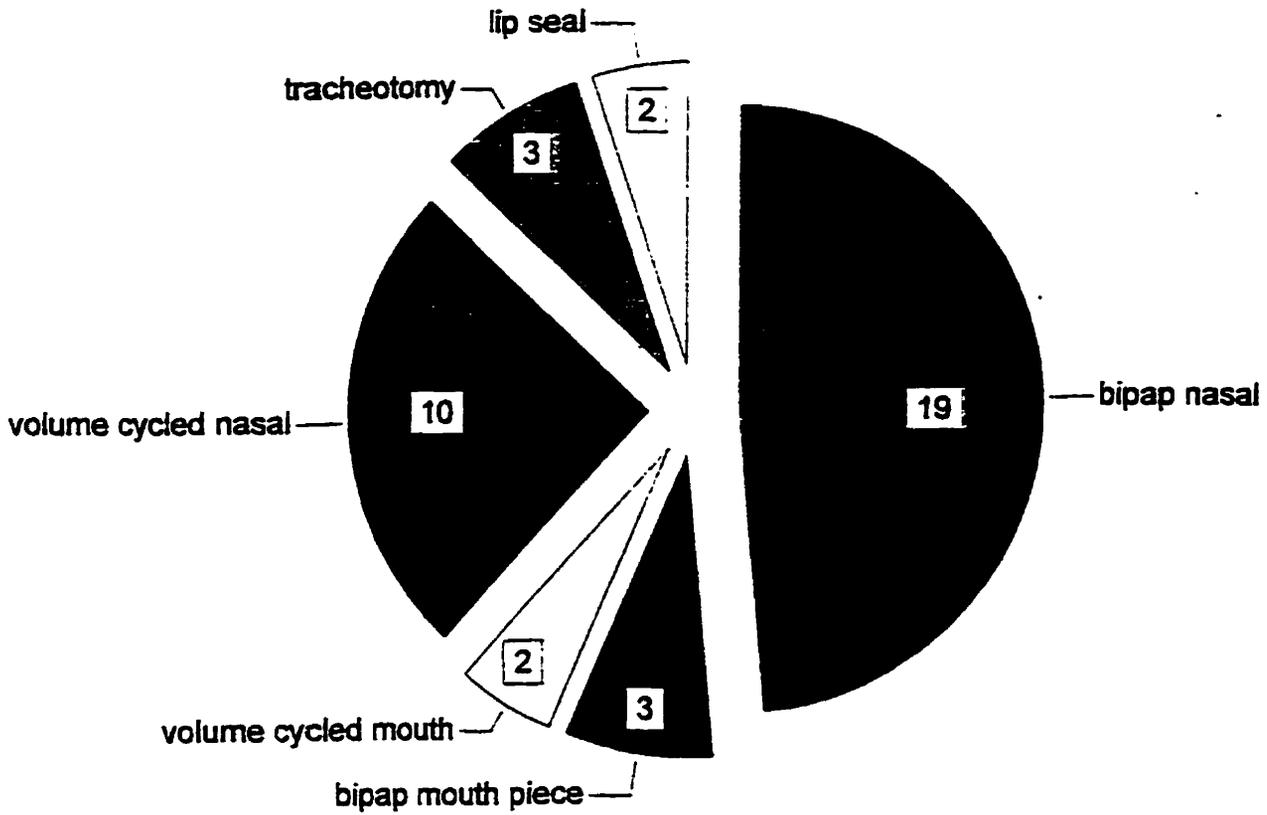
All of the physicians who disclosed information about ventilation (n=44) were asked whether or not they recommended ventilation to their patients. The physicians were asked to quantify, expressed as a percentage, how frequently they *recommend*, *advise against* or present ventilation information *impartially* (see appendix 1, question #16). Thus, the question was structured such that a physician might, for example, report s/he recommends ventilation to 20% of patients,

Figure 1a: Frequencies and types of fulltime ventilation



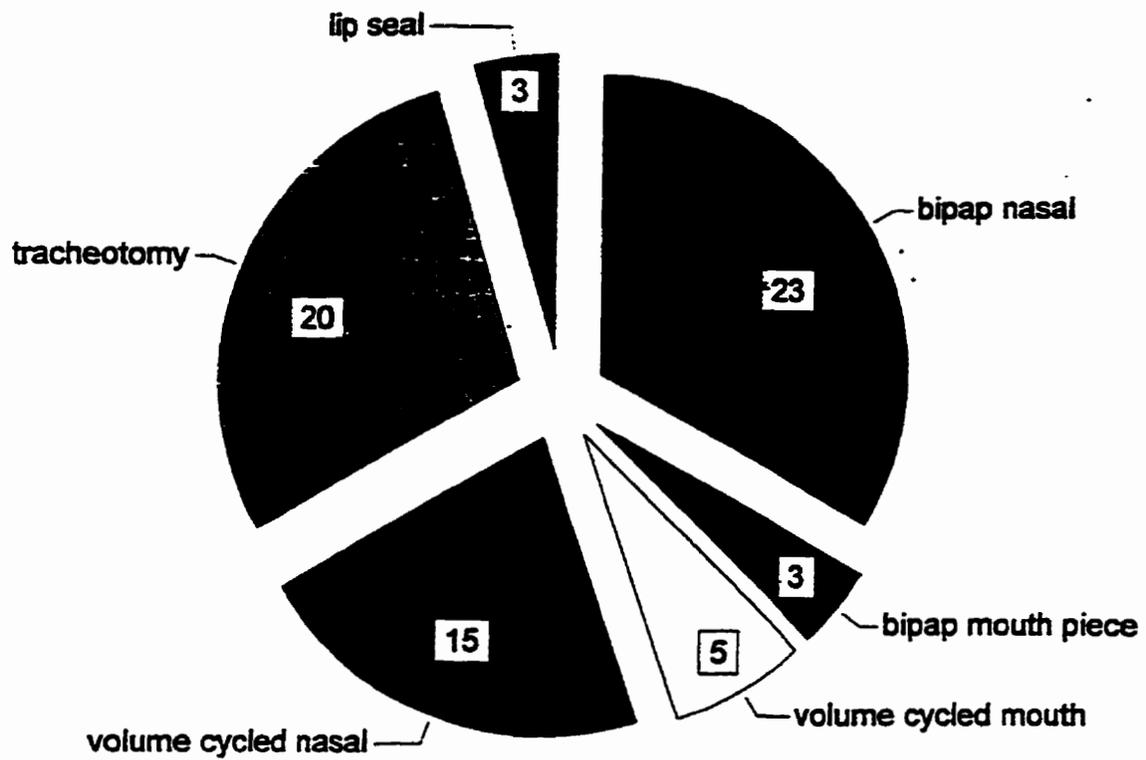
(Numbers represent number of patients using each type)

Figure 1b: Frequencies and types of part time ventilation



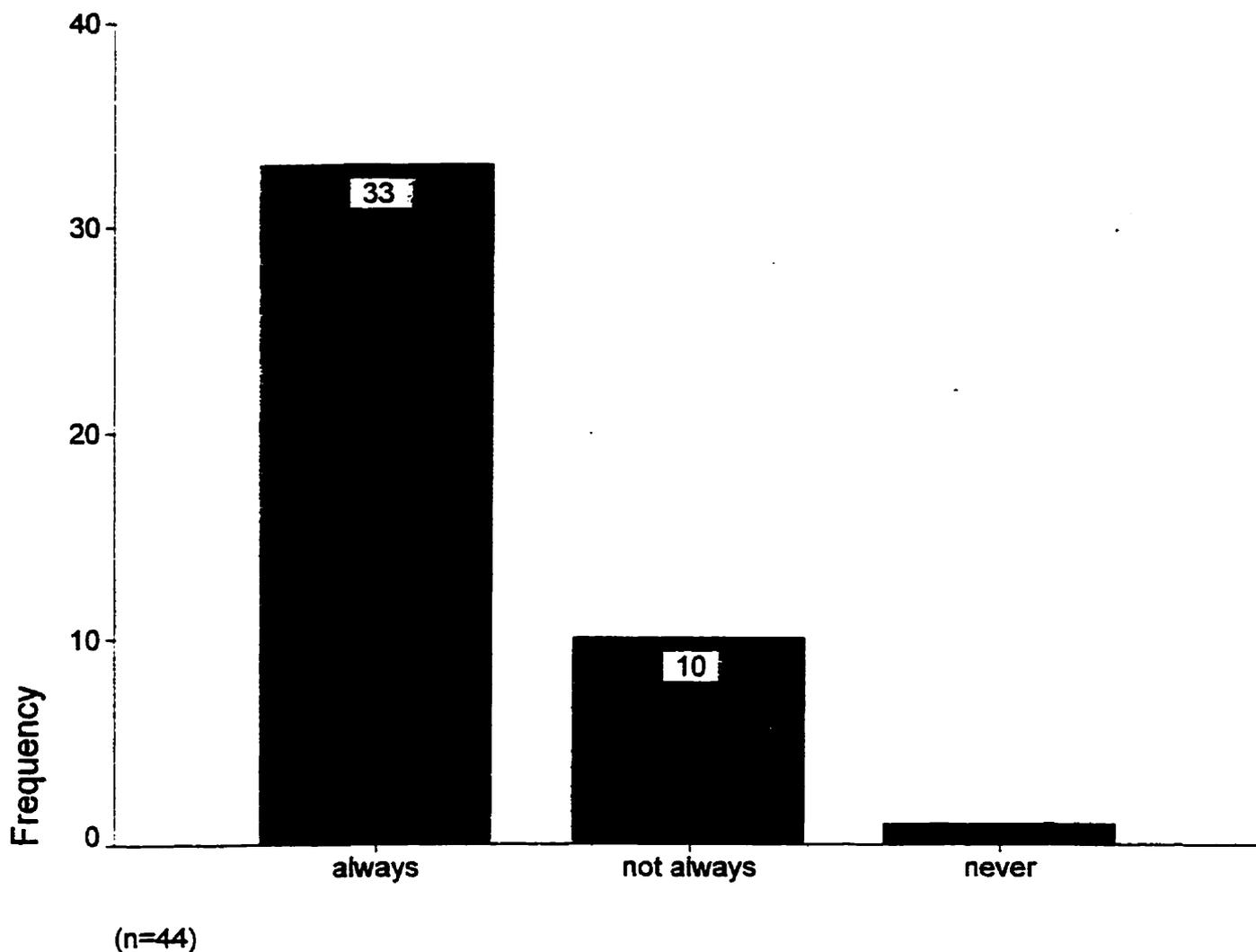
(Numbers represent number of patients using each type)

**Figure 1c: Frequencies and types ventilation:  
Combined full and part time**



(Numbers represent number of patients using each type)

Figure 2: Frequency of informing patients about the option of LTV



advises against to 10% of patients and discusses ventilation impartially with 70% of patients.

The results are as follows. Not all results are represented in tabular form. Summaries of the numbers of physicians who always give the same advice and those who give varying advice appear in tables 3a and 3b. Overall, the majority of the respondents (78.5%, n=33) reported they were for the most part (i.e.,  $\geq 50\%$  of the time), impartial in their discussions. All but one of the physicians displayed a definite preference for one advice option over the others; that is, they gave one form of advice  $\geq 50\%$  of the time.

Of the 42 physicians who gave a response, 39 (92.8%) reported they were impartial at least part of the time. The mean percentage of time the physicians said they were impartial was  $86.21\% \pm 25.6\%$  with a range of 20-100%. Twenty-four (57.1%) physicians said they were impartial with all of their patients (i.e., 100% of the time).

Thirteen (31.0%) physicians reported they recommended ventilation at least part of the time (mean=  $49.3\% \pm 34.8\%$ , range =5-100%). Of these, 2 (4.8%) recommended ventilation 100% of the time.

Ten (23.4%) of the physicians reported that they advise against ventilation at least part of the time (mean=  $7.3\% \pm 7.3\%$ , range= 1-20%). None of the physicians said they always (100% of the time) advised against ventilation.

**Table 3a: Number of physicians who always give the *same* advice regarding ventilation:****Type of advice by frequency (n=26)**

Type of advice	Frequency
Always impartial	24
Always recommend	2
Always advise against	0

**Table 3b: Number of physicians who give *varying* advice regarding ventilation:****Type of advice by frequency (n=16)**

Type of advice	Frequency*
Sometimes impartial	15
Sometimes recommend	11
Sometimes advise against	10

\* total frequency is >16 because respondents could choose multiple responses (see text)

total missing cases for table 3a and 3b = 2

## **Practice Rationale**

### *Reasons for not informing/advising against ventilation*

Respondents were asked to report the reasons why they might not inform patients/families about ventilation (questions #11 and #13, appendix 1) or advise against this option (question #17, appendix 1). These questions were asked separately on the questionnaire, but the questions contain identical response options. Table 4 contains the response frequencies for both questions.

Respondents were allowed to choose as many reasons as might apply to their practice. Nineteen physicians gave a total of 58 responses. The most frequently cited reason for not recommending ventilation was poor patient quality of life, 9/17 (52.9%). The second most cited reasons were distributed equally between lack of home care support availability and inadequacy of the home environment with 7/17 (41.1%) choosing each of these reasons.

### *Reasons for informing patients/families about ventilation*

Respondents who said they disclose ventilation information were asked to give their reasons for this practice.\* The results are summarized in table 5. Forty-two physicians gave a total of 91 responses. The most commonly cited reason for informing patients/families about ventilation was

---

\* They were allowed to choose as many reasons as might apply to their practice. Respondents were also asked why they recommend or are impartial when presenting information regarding ventilation. For each of these questions, the response options were identical (question # 12 and #18, appendix 1). Parallel results were obtained for both and therefore to simplify the reporting, only the results of the first question are presented.

**Table 4: Reasons for not offering/recommending ventilation†**

<b>Reason</b>	<b>not offering (n=2)</b>	<b>not recommending (n=17)</b>
poor quality of life	1	9
inadequate home environment	-	7
inadequate home care support available	-	7
poor cognitive function	-	6
burden on the family	-	5
ventilation is prolonging suffering	-	5
family unwilling to direct their own care	-	5
ventilation is prolonging death	-	4
lack of family financial resources	-	3
institutional care too distant from family	-	2
other	2	1
inappropriate use of resources	1	-

† respondents could choose multiple responses

**Table 5: Reasons for informing patients/families about ventilation (n=42)**

<b>Reason</b>	<b>frequency†</b>
ethical obligation	41(97.6%)
patients can enjoy good quality of life	24 ( 57.1%)
patients should not be influenced by clinicians	13 (31.0%)
legal obligation	11(26.2%)
other	2(.5%)

† respondents could choose multiple responses  
missing cases = 3

"an ethical obligation to inform patients of all options" (41/42, 97.6%). The belief that it is possible for patients to enjoy a good quality of life on ventilation was the second most frequently cited reason (24/42, 57.1%).

### **Timing of Discussions and Clinical Review**

#### *Timing of information sharing*

Respondents who follow paediatric patients (n=32) were asked to respond to two questions regarding the timing of their discussions regarding ventilation with patients and families. The first question asked when the physician first tells the patient and/or family about the option of ventilation and the second question asked when more in-depth discussion was initiated. The responses are listed in tables 6a and 6b.

Of the 31 physicians who responded, 10 (32.3%) *first tell* patients/families about the option of ventilation when the patient begins to develop laboratory signs of respiratory compromise and 9 (29.0%) tell shortly after the initial diagnoses of DMD.

Thirty of the physicians who inform patients of the option of ventilation reported that they engage the patient and/or family in further "in depth" discussion. Thirteen (43.3%) of the 30 physicians who responded said they begin these discussions when the patient develops laboratory signs of respiratory compromise, while 5 (16.7%) said they began these discussions after the

**Table 6a: Timing of ventilation discussions: Initial disclosure**

<b>Timing of Initial Disclosure</b>	<b>n</b>	<b>(%)</b>
first visit	3	(9.7%)
shortly after diagnosis	9	(29.0%)
when patient admitted for elective surgery	2	(6.5%)
laboratory signs of respiratory compromise	10	(32.3%)
clinical symptoms of respiratory compromise	1	(3.2%)
when patient/family asks	1	(3.2%)
depends on family situation	2	(6.5%)
other	3	(9.7%)
	n=31	(100%)

**Table 6b: Timing of ventilation discussions: In-depth discussions**

<b>Timing of in-depth discussions</b>	<b>n</b>	<b>(%)</b>
first visit	2	(6.7%)
shortly after diagnosis	1	(3.3%)
when patient admitted for elective surgery	1	(3.3%)
laboratory signs of respiratory compromise	13	(43.3%)
clinical symptoms of respiratory compromise	5	(16.7%)
at time of respiratory failure	1	(3.3%)
when patient/family asks	2	(6.7%)
depends on family situation	1	(3.3%)
other	4	(13.3%)
	n=30	(100%)

missing cases = 1

patient developed clinical symptoms of respiratory compromise. One physician reported not discussing ventilation until the time of respiratory failure.

### *Frequency of review*

All respondents (n = 45) were asked how frequently patients are seen for routine clinical review. The mean response was every  $6.16 \pm 1.47$  months, range = 3-12 months. In addition, respondents were asked how frequently they reviewed patients with known or expected imminent respiratory compromise. The mean response was every  $3.62 \pm 2.05$  months, range = 1-12 months.

### *Progression of review*

The data were further analyzed to determine if individual physicians were increasing the frequency of follow-up for patients with imminent respiratory compromise. Physicians who only follow adults and all respirologists were not included in this analysis because presumably adult DMD patients and/or those followed by a respirologist may already be facing imminent respiratory compromise. Therefore progression of follow-up may not be applicable for these physicians. Once these individuals are eliminated, 25 physicians remain. Twenty (80.0%) of these reported increasing the frequency of follow-up for patients with imminent compromise. Five of the 25 (20.0%) did not increase their follow-up frequency (routine review: mean= $6.0 \pm 2.1$  months, review when expected respiratory compromise: mean= $6.6 \pm 1.3$  months). Three of these physicians reviewed patients every 6 months and continued this practice when compromise was imminent. One physician continued following patients every 9 months and one physician followed patients less frequently, initially following them every 3 months and then increasing the time to every 6 months when compromise was imminent.

### **Allocation of resources**

All respondents were asked if they believed society could not afford to provide long term ventilatory support for individuals with DMD. The majority of the physicians reported they either strongly disagreed (n=20, 44.4%) or disagreed (n=12, 26.7%) with the statement. Eleven (24.4%) were unsure and two respondents (4.4%) agreed that ventilation was too costly to provide. None of the physicians strongly agreed.

### **Attitudes towards disability**

The Modified Issues In Disability Scale (MIDS) score was calculated for each of the respondents. The MIDS score provides a measure of an individual's attitudes towards disability. The highest possible score (the most positive attitudes towards disability) is 259. The scale is designed to compare attitudes amongst individuals or groups, not to indicate who has “good” or “bad” attitudes. The mean score obtained was  $185.7 \pm 23.0$  (Median 186.5, bimodal: 187 and 212, range= 138-236).

The MIDS score was used to compare the attitudes towards disability of selected subgroups of physicians. The mean for females ( $196.9 \pm 18.1$ ) was greater than that for males ( $183.2 \pm 23.8$ ) (*t*-test  $p=.079$ ). The means for groups agreeing or disagreeing with allocating resources for ventilation can be seen in table 7. The trend indicates that those who believe ventilation is too costly to provide have the least positive attitudes.

**Table 7:****Do you agree or disagree that society cannot afford ventilation for patients with DMD?****Physicians' opinions by mean MIDS score**

<b>Opinion</b>	<b>mean MIDS Score</b>
agree	152.0
unsure	178.6
disagree	186.1
strongly disagree	192.4

This trend was not statistically significant (one-way ANOVA  $F(3,40)=2.59$ ,  $p=.067$ ). Based on one way analysis of variance, no significant differences in mean MIDS scores were found amongst groups based on the variables of age, years working in a neuromuscular clinic, or clinical specialty. Comparisons between disclosure practices and MIDS score are outlined below.

### **Comparison of disclosure practice to other variables**

For purposes of analysis, disclosure practice was divided into two groups, those physicians who always informed patients of the option of ventilation ( *always*,  $n=33$ ) and those physicians who sometimes didn't inform patients of the option ( *sometimes*,  $n=11$ ). The *Always* group had a mean MIDS score of 190.3 while the *Sometimes* group had a score of 175.5 ( $t$ -test  $p=.062$ ).

A statistically significant difference was found in the disclosure practices of male versus female physicians. Table 8 illustrates that female physicians are more likely to inform their patients about the option of ventilation (Chi-square=4.48,  $p=.034$ . Fisher's exact test  $p=.034$ ).

Using bivariate analysis and the Chi-square statistic, no significant difference in disclosure practices were found amongst groupings based on age, number of children, regional location of clinic, clinical specialty or years in neuromuscular practice.

**Table 8: Disclosure practice\* by gender**

	<b>Always disclose</b>	<b>Sometimes disclose</b>	<b>Row total</b>
<b>Female physicians</b>	<b>10</b>	<b>0</b>	10 23.3%
<b>Male physicians</b>	<b>22</b>	<b>11</b>	33 76.7%
<b>Column total</b>	32 74.4%	11 25.6%	43 100%

Chi-square = 4.48,  $p = .034$ . Fisher's exact test  $p = .034$

missing cases = 2

\* practice of informing patients/families about the option of long term ventilation

## **Part Two: Qualitative Description of Practice and Attitudes**

### *Demographics of the interview participants*

The interviews were conducted between May and July of 1996. A selected sample (criteria for sampling described on page 46) of six physicians were chosen from a list of sixteen who volunteered from the questionnaire sample population. The six physicians were situated in four different cities from three different provinces in Canada, with representatives from both eastern and western provinces. Five of the respondents were located in cities of population sizes greater than 500,000 and one respondent was located in a city of less than 100,000. Further information on location is not supplied to protect the confidentiality of the physicians. Table 9 shows the distribution of the respondents' demographic characteristics compared to that of the larger study sample.

### *Overview*

The core phenomenon examined is **physicians' management of long term ventilation decisions**. Here “management” refers to the specific actions that physicians undertake that lead to a decision for or against initiating LTV. For example, reflection, consultation, disclosure of information, referral and sharing of values are actions physicians might undertake as part of management. In many situations, these actions involve interactions with the patient and his family and occur over time. All physicians gave detailed accounts of their approach to LTV decisions. The analysis of these accounts suggest that the physicians engage in a general process of reflection and discussion prior to embarking on a course of action. This process was general as well as patient specific.

**Table 9: Demographic Distribution of Interview Participants**

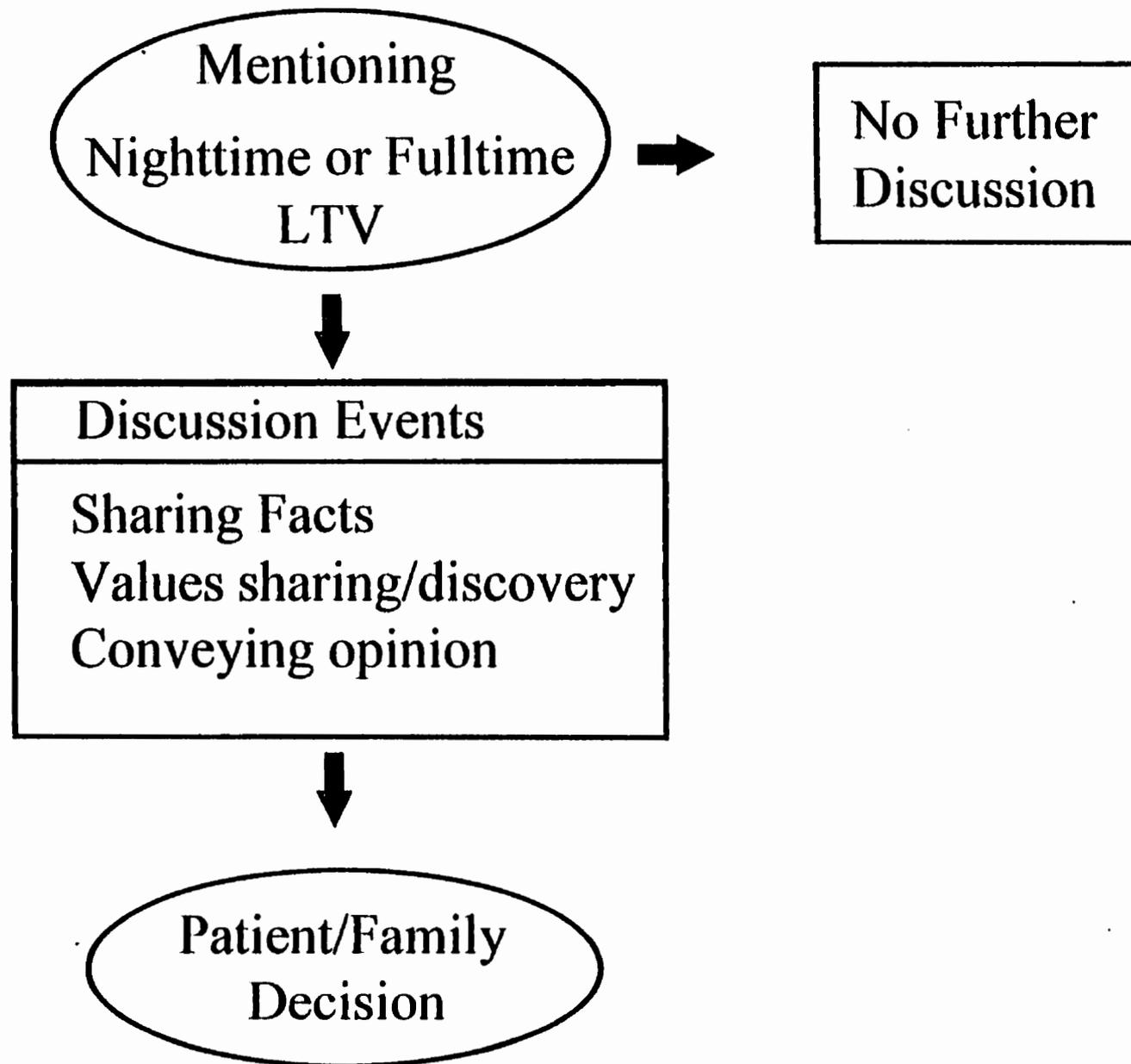
<b>Characteristic</b>	<b>Proportion of interview sample</b>	<b>Proportion in study sample</b>
Clinical specialty	Paediatrics 2      33.3%	13.3%
	Neurology 1      16.6%	26.7%
	Respirology 3      50.0%	31.1%
Clientele	adult 1      16.6%	28.9%
	paediatric 3      50.0%	48.9%
	mixed 2      33.3%	22.2%
Disclosure practice	always disclose 4      66.6%	75.0%
	not always 2      33.3%	22.7%
Advice practice	100% impartial 4      66.6%	57.1%
	mixed advice 2      33.3%	38.1%
Marital status	100% married	88.9% married
Children	100% had children	93.3% had children
Gender	100% males	75.6% male
Years of experience	range = 1-18 years mean = 10.9 years	range = 1-28 years mean = 11.6 years
Age	range = 40-53 years mean = 45.8 years	range 34-69 mean = 47.0
MIDS score	range = 152-220 mean = 178.7	range = 138-236 mean = 185.7

The physicians hold individual general opinions about the benefits and burdens of ventilation for patients with DMD and relate a personal history of how they came to hold these opinions. Analysis of the physicians' accounts revealed three shared major themes that were important to their management of LTV decisions. These are:

- 1) **Mentioning and discussing:** Mentioning and discussing LTV are discrete events with different purposes.
- 2) **Night-time/fulltime distinction:** Management may vary with the type of ventilation (night-time vs fulltime) being considered.
- 3) **Discussion and influence:** Physicians vary the quality and frequency of discussions with the effect of influencing outcome.

The three themes, while related to each other, also emerge as discrete ideas with different origins and consequences. They are presented below sequentially to demonstrate how each subsequent theme is influenced by the previous and contributes to the overall management of LTV decisions. An overview of the physicians' management of ventilation is presented in Figure 3. The overview is a schematic representation of the physicians' management as it relates to the three themes and emerged from the accounts of all the physicians. Each theme is discussed in detail with supporting quotes from the interviews. The quotes that are chosen are representative of all of the physicians (unless otherwise stated) not only the physician quoted.

**Figure 3: Overview of Physicians' Management of Ventilation Decisions**



## Themes

### **Mentioning and discussing:**

Physicians initially *mention* ventilation to patients and families as a way of introducing the topic. *Mentioning* used here refers to the action of imparting information without entering into a dialogue about that information. It is a transfer of information without searches for shared meaning or understanding. It does not usually involve questions and answers or anything beyond cursory attempts to assess patient/family understanding of that information. Mentioning is usually embedded in other information presented during a particular clinical encounter:

...now, ah, when the patient comes and, the muscular dystrophy patient, and he's six years old and you're talking with the family about the diagnosis, and you say, well, about the age of ten he may be in a wheelchair, in the next sentence you say, well, at the age of ... fifteen or twenty he may need some extra help with ventilation.

Mentioning may or may not be followed by *discussion* events. *Discussion* here is used in the sense of a conversation that involves one or more speakers who, through revealing information and questioning, consider the pros and cons of a subject or subjects. Discussion events may immediately follow mentioning or may occur days, weeks, or years later. The use of mentioning and discussing in relation to the physician's approach to LTV decisions is demonstrated in the following quote:

I think its something that should be brought up first of all at the time of diagnosis-just mention it in sort of a broad sense that these children could have- You know their muscles are going to get weaker and weaker including the respiratory muscles -in as simple terms as you can put that. Um , but I think its important, um, to mention that again as I told you in terms of select times, over the next several years mention it again...If the respirologist said to me in the last clinic I think we're getting close to the time we should talk to his parents about this kind of issue in more detail, then I see them...

The physician in the following quote expressed a similar approach:

(W)hen I talk about, you know, the respiratory failure, etc., I will mention that there is an option of ventilation that in due time we will discuss.

Whether or not to proceed from mentioning to discussion depends on the physician's and the team's general philosophy and their opinion on the best course of action for a particular family. From the data, there appear to be two distinct categories of action. One group of physicians discuss ventilation options with all of their patients/families regardless of individual family circumstances or type of LTV under consideration. This is demonstrated in the following quotes from two of the physicians:

Dr.: There is no wrong decision...My conviction is that I am not going to change the decision. We're just trying to tell them that they have to make a decision and have to live with their decisions. But I don't think there's one decision.

BG: Do you make any distinction in your own mind between ventilating a patient part-time as opposed to fulltime?

Dr.: No. But almost all of them start off part-time overnight, and again it's their decision...(to escalate to fulltime).

These physicians reported that they discuss LTV with all families because they believe it is the family who determines which option will be best for them. In the first quote, the physician emphasizes that there is no 'wrong' decision. Based on this belief, he approaches all families in a similar fashion by carrying out in depth discussions regarding LTV options. Both accounts reveal that the physicians believe they have no motivation to limit ventilation discussions because they view it as a personal choice. LTV was discussed with all families regardless of type of ventilation being considered or the families' particular circumstances.

Other physicians may refrain from discussing LTV if the team agrees that LTV would result in an unacceptable quality of life for the patient, particularly (but not only) if fulltime ventilation is being considered. This does not preclude the mentioning of ventilation, but rather,

in-depth discussion events are not normally pursued unless initiated by the families. For example, the following case was described by one physician:

Well, I guess there was one family that did decline ventilation, where the mother had recurrent psychiatric problems with anxiety and depression, and, ah, we just couldn't see, she could barely handle the boy as he was, you know, he was just a regular guy with Duchenne's, a well-adjusted kid. Even then, you know, every nine months we'd hear she was having a breakdown or something, so we said to ourselves, there's absolutely no way we can add to this...We did raise it though, but we didn't push at all. They went home and they talked and they said, we don't want it, and we said fine, I guess that's about as close as we would get to not introducing it.

In this example, the team has informed the family about the option of LTV by mentioning it but not pursuing in-depth discussions with the family. Although the physician does not indicate exactly what he means by "we did raise it", his comment at the end, "that's as close as we might get to not introducing it" suggests that the option was not discussed in detail.

In all the interviews, physicians expressed concerns about the potential burdens to the family, as is demonstrated in the above quote. The team in that particular situation felt that the mother could not cope with the burden of caring for her son if LTV were commenced. Limiting discussion was only one way of managing this concern and was not common to all the physicians. Other strategies are discussed under the theme of *discussion and influence* below. Mentioning without proceeding to discussion was most frequently evident in the context of switching from night-time to fulltime LTV. I turn now to discussion of that theme.

#### **Night-time/fulltime distinction:**

While the data indicate that initiating night-time LTV has become standard practice in the last few years, fulltime ventilation is still not routinely discussed by all physicians

with all patients. Night-time ventilation may be viewed as a relief of symptoms improving day time quality of life. For example one physician put it this way:

Night-time ventilation to me is a quality of life issue. Okay? Night-time ventilation, the children, the kids or the adults whatever they are- in the mornings they don't wake up with headaches, miserable. Parents tell you, they say these kids are miserable in the morning. They're - Some of the parents are very crude- they say "listen, he's a bitch to live with in the morning." That's what they tell you. One mother came in a few months ago and I said "What do you mean by that." "Oh he's just crabby, you know, he's just barking at us all the time and he is miserable and I know he's not happy Dr.[name]" Okay, put them on night-time ventilation and the next morning they tell you- I feel better.

Quality of life was often referred to when physicians were discussing differences in their management of fulltime versus night-time LTV decisions. Fulltime ventilation is viewed as life extending, but physicians vary on their opinion as to whether or not this represents a benefit or a burden for all or specific patients. One physician expressed his belief that fulltime ventilation results in an unacceptable quality of life:

I guess I'm sort of, I'm sort of still having a bias in terms of ventilatory support for somebody who is, for somebody who has a terminal disease. Um, their quality of life is the pits. Once they get on chronic ventilation, you've now forgotten about quality of life as a positive thing and you're thinking you know, just survival.

When the physicians were asked to define what they meant by the term "quality of life", they tended to offer examples rather than general definitions as is apparent in the following quote:

BG: When we were talking earlier, you mentioned that you might not necessarily recommend or offer ventilation to a patient with a poor quality of life, and I wonder if you could tell me what you mean by quality of life, because people have different views.

Dr.: One of the first criteria that we put down was a patient flat on the back all the time, um, severe developmental delay, um. those would be two things.

Another physician quoted the literature to support his opinion that only patients can judge the quality of their own lives:

And if you have read the articles by Bach about the perception of quality of life from the physician's point of view and from the patient's point of view, we're talking two different things. Totally different. In fact, I am not the one to decide what is his quality of life. You decide what is

**your quality of life, and I decide about mine. And that's, you're talking about technology and we're talking about people that had to decide whether they wanted the ventilator or not, and that's their choice. And if you had read the questionnaire etc. that has been done with these people, they have a good quality of life.**

Some physicians associated fulltime LTV with tracheostomy while others saw it as the increased use of noninvasive means. This reflects current debate in the literature where some centres strongly advocate for noninvasive systems for the majority of patients<sup>3</sup> while others feel tracheostomy is more effective and therefore the treatment of choice.<sup>126</sup> All physicians agreed that noninvasive options were more easily introduced to and accepted by patients especially for use at night only. When full-time ventilation was associated with tracheostomy, some physicians were opposed to its implementation. In the following exchange the physician indicates that he is opposed to LTV by tracheostomy because of the risk of the child requiring institutionalization:

**BG: You don't like offering trach's to the MD patients?**

**Dr.: We don't do that.**

**BG: Why is that?**

**Dr.: We, because, ah, a few reasons. One is that it may result in them being trapped in an institution.**

**BG: Um-hum.**

**Dr.: If it appears to be unnecessary. If it's invasive and it interferes with their feeding and sometimes their speech. And not necessarily, except for very, very late stage disease, any more effective.**

**BG: So you're comfortable having patients on 24 hour ventilation nasally?**

**Dr.: Comfortable? It's not entirely desirable. But we've done it. A few times. And we have not encouraged people once they require that much ventilation to switch to tracheostomies.**

This physician has strong feelings about patients with tracheostomies ending up in an institution where he feels they will have an unacceptable quality of life (alluded to by his use of the word "trapped"). He made this point several times. This particular physician seemed to believe that tracheostomy usually resulted in institutionalization or at least that the risk of institutionalization was too great. This was a curious conclusion considering that programs for home ventilation existed in his community, some of which he described to me himself. However, he was not alone

in his concerns as other physicians expressed worry about institutionalisation. However, the opposite point of view was also expressed:

Dr.: (W)e can have some of them, young adults, that can go into independent living centres, [CENTRE NAME] that I was referring to, and we have a few patients at [NAME] admitted in the hospital.

BG: Oh you do.

R: Yah. We have three patients, one with a tracheostomy, 28, 29, something like that, which will be there for a long time. Still in good shape. Very alert and tenacious. So I think the only thing that we can do is to try to help people cope with this situation, and if it's not possible, to try and find solutions, that's what it is, I guess. But you cannot condemn the individual to die because his parents aren't as able to cope with this situation.

A physician's personal opinion about night-time ventilation means that his/her protocol may end with a discussion and implementation of night-time ventilation with no planned discussion beyond that. Referring to the model in figure 3., the physician may proceed "downwards" from mentioning night-time LTV to discussing it until a family reaches a decision. When the process needs to recommence to consider progressing to fulltime LTV, mentioning may be done but the process ends there with no further discussion and no progression of therapy. One physician reported that he can "sense" when patients do not want to talk about escalating therapy:

Then I say- well certainly there's other options at that point. You could go to the chronic ventilatory support. Usually once you start to talk about it you get the sensation that the majority of the parents aren't very happy about the fact that you need a trach and all this other stuff. Its just not, they've already got a g-tube and they have to have a trach now, and have had spinal surgery and all this stuff. A lot of them get- What I tend to do at that point is say well lets not talk about this right now.

Early in the interview, this physician revealed that he did not believe that it was possible for a patient to enjoy a good quality of life with fulltime LTV via a tracheotomy. None of his patients to date had progressed to LTV with a tracheotomy while under his care. Considered in this light, the quote suggests that a subsequent discussion will not take place or at least will not be initiated by the physician. What the physician 'senses' may be more indicative of his own feelings

surrounding fulltime tracheotomy ventilation rather than the family's. This is partially affirmed by the accounts of other physicians who did not report that "the majority of parents aren't very happy about the fact that you need a trach." Previous research<sup>14</sup> also indicates that physicians may unwittingly impose their own preferences for life sustaining treatments onto patients.

In contrast, some physicians manage the fulltime decision in the same manner as the night-time decision and the full gamut of discussion events occurs with each. The physician in the following quote discusses progression to fulltime LTV in relation to the patient's personal choices and needs:

And they often ask, well, themselves, is it fulltime or part-time? I say to them, it's your decision. You start at part-time, and it may go on just overnight for ten years, or five years, or one year. You may find you get tired after meals and want to rest for an hour. You may find that eventually you're going to be using it all the time, maybe twenty hours a day or so, but, ah, I'm not going to tell you how much to use it.

The night-time/fulltime distinction also becomes apparent in the discussion events reported on by the physicians and is described under the following theme.

### **Discussion and influence:**

Discussion involves an in-depth exploration of the option of ventilation which can occur over time but usually involves at least one lengthy discussion with one or more team members.

The events that trigger these discussions are related to lab results or patient symptoms. For example:

I may see them, say if they're muscular dystrophy, when their vital capacity falls to a certain level, which would be about, ah, two to four years before we think that they might need to be ventilated, and we discuss other therapies to help, positive pressure breathing, chest physiotherapy, things that they should know for the next step. And I also mention, well, ventilation seems, do you have any questions about it or whatever.

Discussion events centre around the sharing of **facts**, **values** sharing and discovery, and conveying **opinion**. From the data, strategies for relaying **facts** included verbal discussions that took place over several visits and involved several team members, educational aids such as videos and written materials, formal or informal introductions to ventilator users, and opportunities to try out the equipment. The following three quotes provide examples:

... we have the team approach to discuss assisted ventilation, for example the occupational therapist will be there, the physiotherapist will be there, the psychologist will be there, social worker will be there, as well as the nurse and the physician.

I give them a hand out or an article or a book, whichever one I feel they are able to handle. Um sometimes we use a video here, we show them that as well. That just kind of describes a bit about this disorder. And then uh, as they keep coming back, I usually-what I use to do before I came here was, I use to um, we use to meet with the kids at intervals twice a year and when they came back and they were starting, you know, as they were showing some difficulties with their gait and that kind of thing and we'd start bringing up these issues again.

...to discuss what are the options. And the option exists to use or not to use the assisted ventilation. And for that we show them the video. Most of the time they see the video with either the psychologist or social worker.

**Values** sharing tended to focus around sharing positive health related values such as the relief of symptoms. Facts and values were sometimes presented together, possibly leaving the impression that all the material was of a factual nature and not open to interpretation. For example:

We approach it in a very concrete or practical way... (T)here are treatments that you might consider which include, you know, are this. Usually we always introduce night-time ventilation, but we don't use invasive ventilation....We don't really see it as a formal process of consent. We don't imbue it with a weighty atmosphere. We explained it to them as a symptom and a treatment, and we feel comfortable with it as that and that's how we transmit it.

This physician has engaged in a prior process of reflection about ventilation and sees non-invasive ventilation as the treatment of choice. While his view may be well founded, it is still based on a value judgement of the relative merits of this and other options (invasive ventilation,

palliation) but appears to be presented to the family simply as a set of facts. He presents information about non-invasive night-time ventilation as a treatment for a symptom, but avoids discussions of invasive ventilation even though invasive ventilation could also be characterised in such a manner.

Physicians may be selective about which of their personal values they share with patients. The following two quotes from the same physician show his willingness to share his values regarding the potential burdens to the mother but not his values regarding the patient's quality of life:

Dr.: Ya, I tell them. I say I worry because your going to find yourself, your own health is going to suffer and I say "health"- I mean physical as well as emotional because you're doing something which you're not trained to do as a mother. You may think you can do it because you feel its your responsibility but people here who are trained to do this. That's their job. I don't think emotionally or physically its going to be good for you. The last thing you want for your child right now is to get you sick so then you can't be of any help.

Later on the same physician offered the following:

Dr.: just being alive uh, with no quality of life-just being on a ventilator to me, uh - My own personal feelings about that are not as positive.  
 BG: Do you share that with the families?  
 Dr.: I guess if they asked me I'd probably do that.

The physician took the initiative to express his concerns about the mother in order to try to persuade her to accept homecare assistance. In the second quote the physician was not comfortable sharing his values regarding quality of life and chronic ventilation with the patient and family but would (probably) share these values if asked. In contrast, one physician used clinic visits to share some information with families but used special appointments, often on weekends, to engage in deeper discussions and values exploration. As he put it:

We talk about real things. We talk about life. We talk about death. We talk about ventilation. We talk about philosophy.

Conveying of **opinion** can be either intentional or unintentional. For example giving a clear recommendation or urging a patient to "give it a try" are examples of intentional conveying of opinion:

On the night-time ventilation, I sort of am positive about it and say " I really believe you might benefit from this. Give it a try."

Or:

You know I've never actually encouraged them to go for the option of, um, continuous ventilation. I encourage them to go for the night-time ventilation.

All the physicians expressed the belief that ultimately it was the family's decision whether to accept or reject LTV. Framing information negatively or positively or not initiating further discussion are examples of unintentional conveying of opinion:

Well, it's not that I don't recommend it. I say look, there's, here are the facts, and the care and the length of time and the benefits and the side effects you're going to get from all of this, side effects in terms of time and social structure, costs to your families that are considerable, but hey, you know. it's your decision in the end, and there you go. So we all tend to sway the families, there's no doubt about that.

Several issues are reflected in this quote. The physician does not appear to want to give a recommendation, presumably because he does not want to direct the patient/family's behaviour. He feels it is the family's decision and relays this to the family. Instead of giving a recommendation he presents some "facts" and "side effects" of ventilation which, as they are described here, are all quite negative. He then admits this information will sway the patient. Not wanting to give an explicit positive or negative recommendation was common amongst the interview participants. Another physician had this to say:

We would frame it in such a way as to lean a little bit against it. We don't tell them it's wrong. And we wouldn't refuse to do it. We tell them that in that type of situation. We would make it clear that it would be a challenge and that it's possible that in the long run it might be very hard. If they insisted, we'd do it.

Again the physician indicates he believes the final decision is up to the family, but only if they "insisted". Information is framed in a negative light to make it appear unattractive. One of the physicians explained that he believes there really is "no wrong answer" for these patients but he felt he influenced them nevertheless:

This is difficult. It's like discussing politics. If I discuss politics with you I'll try to influence you. But I don't think I try to influence them, because I think there's not any wrong decision or good decision. People have a choice to die and people have the choice to decide to go on with living, and it's hard, but there's no, not a bad decision from an ethical or religious or medical or human point of view. So basically, the message I was trying to give when I was in the initial meeting was, our goal is to help you with whatever you choose.

This quote is also representative of the caring attitude expressed by all the physicians towards the families they see. Throughout the interviews it became apparent that they all dealt with difficult issues on a day to day basis. Many of them had been through a process of securing funding for their LTV programs or advocating for LTV despite strong differences of opinion encountered from their colleagues in acute care:

Believe me, we fought hard. (We felt) this is our obligation as a team and as a hospital...But it was a fight all the way from about 1982 to 1988/1989. Many in [PROVINCE], neurologists, thought that we were crazy and we were out of our mind, to do that. (They said) "That's something that you shouldn't let these people have."

What most came out in the interviews was the physicians dedication to providing the best care possible to their patients and families.

### *Summary*

The three themes presented, *mentioning and discussing*, *night-time/fulltime distinction* and *discussion and influence* are part of the larger phenomenon of *physicians' management of LTV decisions*. Figure 3 provides a schematic representation of the phenomenon and its elements. The physicians' management of LTV decisions is integral to the decision making process that

involves physicians, team members, patients and their families. The themes represent commonalities amongst the interviews. Physicians initially mention ventilation prior to engaging in discussions with families. Mentioning precedes discussing and serves to prepare the family for the discussion later on. Alternatively, mentioning may not proceed to further discussion, especially in the context of fulltime ventilation decisions involving tracheostomy. Here two distinct groups of physicians could be identified, those who encourage fulltime ventilation regardless of method of delivery and those who were opposed to tracheostomies. All of the physicians admit to having influence over the families but believed that the final decision was the family's. Physicians convey their opinions to patients intentionally but also appear to convey opinion unintentionally by framing or emphasizing some information and de-emphasizing other. A further discussion and analysis of the physician-patient-family relationship, the duty to disclose as part of a valid informed consent and the effects of physician power on communication (as they relate to LTV decisions) is the focus of the Normative Analysis, Chapter 5.

## **Summary and discussion of empirical results**

In this section I summarize and discuss the key findings of the empirical research and identify ethical concerns that will be addressed in the normative analysis.

### *Disclosure*

The results indicate that the majority of physicians (75.0 %) disclose information regarding LTV to all their patients. However, a significant minority (22.7 %) reported they do not disclose to all patients and one physician (2.3 %) never disclosed LTV information. The results raise questions about whether or not nondisclosure can be morally justified and if so, in what kinds of cases. If disclosure is not always appropriate, then the practices of the majority of the physicians may be flawed. Alternatively, if disclosure to all patients is morally demanded, then the practices of a significant number of physicians may be misguided. In the normative analysis, I will argue that disclosure of LTV information is morally required of physicians for every patient with DMD (with some very limited exceptions) regardless of patient/family individual circumstances or the type of ventilation being considered (night-time/fulltime, invasive/noninvasive). Furthermore, I will argue that the practice of *mentioning* LTV without proceeding to further discussion satisfies disclosure requirements only in a very narrow sense and does not adequately fulfill the duty of the physician to assist families in understanding the potential benefits and burdens of all options and to make choices based on the family's values and goals.

### *Advice and influence*

Recommendation practices and other forms of conveying of opinion are important to study because physicians are in a position to influence patient/family choice. The physician's approach to decision making can potentially help or hinder the goal of choosing care options that are most consistent with patient/family values. The majority of the physicians (57.1% ) reported that they always present information to patients and families in an impartial manner rather than recommend or advise against ventilation. While it may be true that these physicians are attempting to eliminate bias and trying not to influence families, whether or not they can present information completely impartially is debatable and not necessarily desirable. Impartiality suggests a laying out of facts without the advantage of the physician's input to guide the discussion and assist the patient in values clarification. A questionnaire does not allow for exploration of the personal meaning each physician attaches to the concept of impartiality and therefore these results taken by themselves need to be interpreted with caution. The interviews offer further clarification. The physicians all voiced a belief that the final decision to accept or reject LTV rested with the patient and family, but admitted to having significant influence on that decision. Most physicians stopped short of giving an explicit recommendation but conveyed opinion to families using a less direct approach. Some forms of influence were overt, such as urging a patient to "give it a try". Others were more subtle, such as framing information in order to discourage a choice. Behaviours that influence choice could be deliberate, but behaviours that may unintentionally sway patient/family choice were also identified. For example, actions that were identified included: mentioning without proceeding to discussion, delaying discussions until (when and if) the patient asks about LTV and unintentional framing of information. In the normative analysis, I outline an approach to decision making that stresses values sharing and

'transparency'. Rather than attempting to eliminate physician influence, which is neither possible or desirable, an open sharing of values and personal deliberations through ongoing conversation is advocated. As part of this approach, the physician and family members engage in self reflection as well as open discussion in an attempt to clarify expectations and the underlying assumptions that can guide behaviour and result in unintentional influence.

The number of discussions related to LTV may serve to emphasize the relative importance a physician places on the option. The majority of the physicians in the survey disclosed and discussed LTV information at least several months before it would be indicated. However, some physicians (16.7%) were still not initiating discussions until the time that the patients imminently required ventilation, that is, when they developed clinical symptoms (Table 6b). While symptoms do not necessarily indicate an emergency situation, an immediate decision is required if symptoms are to be alleviated and to reduce the risk of respiratory failure. Other physicians waited until the family asked about LTV or waited until the patient was already in respiratory failure before initiating discussion. I will argue that the practice of disclosing early and discussing LTV over time is the preferred approach because it allows families time for reflective deliberation and allows for appropriately involving the developing child in decision making as he matures.

### *Quality of life*

In the questionnaire "quality of life" was stated as a reason both for offering or recommending LTV and for *not* offering or recommending LTV. *Poor* patient quality of life was the most common reason (10/19, 52.6%) for *not* offering/recommending LTV (Table 4). The belief that patients can enjoy a *good* quality of life was also a common reason (57.1%) for offering/recommending ventilation. This may be explained by general attitudinal differences amongst the physicians but is also at least partially explained by patient-specific information. A particular physician may feel that one of his or her patients will enjoy a good quality of life with LTV while another patient may not. Two general distinctions that physicians made were revealed in the interviews. Some physicians believe that (1) institutionalisation and/or (2) fulltime ventilation with a tracheotomy would result in unacceptable quality of life. The meaning that physicians ascribed to the term "quality of life" were not readily apparent. The physicians tended to refer to examples when attempting to define quality of life such as "in an institution" or "flat on his back." As I have discussed in the literature review, quality of life judgements, whether positive or negative include value based subjective assessments that may vary from individual to individual. What one physician considers an acceptable quality of life for a patient, another may not, and the patient may in turn also have a different view. I will argue that rather than making unilateral determinations of quality of life, physicians would do better to engage in open exploration of goals and expected outcomes with the patient and his family.

*Attitudes towards disability*

The MIDS score provides a measure of the individual physicians' attitudes towards disability. The mean score obtained was 185.7 ● 23.0 out of a possible score of 259. (A higher score is indicative of more positive attitudes.) This mean can be considered relatively high when compared to other population studies but lower than those reported for other groups of physicians. Makas,<sup>127</sup> who developed the scale, has collected data on college students, staff, and faculty in two studies and obtained mean scores of 177.61 and 176.87. The mean obtained in the present study is comparable to that obtained by Pfeiffer<sup>128</sup> in his sample of medical/educational and social services professionals, (mean = 183.97), and direct medical care professionals (mean= 187.97). However the results are lower than his sample of physicians (mean= 194.36) and Gutierrez's<sup>129</sup> sample of physicians (mean= 191.45).

A possible association between disclosure practice and attitudes towards disability was identified; those physicians with higher scores on the MIDS were more likely to disclose LTV information. This finding is consistent with other empirical research<sup>4,6,113,114</sup> that demonstrates correlations between negative attitudes towards disability and clinician's lesser likelihood of discussing life saving treatments with disabled individuals. Macro-level initiatives, such as education campaigns or institutional policy changes to address negative attitudes in the general public and amongst health care professionals, are important but go beyond the scope of this thesis. For the influence of negative attitudes to be addressed at the micro-level of the patient-clinician relationship, each health care professional who works with disabled individuals must be willing to examine his or her personal biases through self reflection and open communication.

Apparent throughout the results is the commitment physicians have towards providing quality care for patients and families. The normative analysis that follows is intended to provide

assistance to clinicians in these endeavours, to explore elements of informed consent and decision making and to elucidate the possible pitfalls that occur in practice that may interfere with the process of shared decision making and the goal of choosing appropriate care options.

## **Chapter 5: Normative analysis**

My intent in this chapter is to examine LTV decisions from an ethical standpoint. The physician's moral obligations to the patient as set out in the literature review are now examined in the context of the case. Competing arguments and potentially conflicting duties are here considered and specified according to the particular features of LTV decisions for patients with DMD. The focus in this thesis is limited to the physician's duty to the patient; potentially conflicting duties to other family members are not considered in any depth. Nevertheless families are considered here as integral to the decision making process as the majority of patients with DMD are deliberating on choices as part of a family unit, the members of which are assumed to love and care for each other and share common life goals.

First I will look at the question of whether or not nondisclosure of the option of LTV can be morally justified. To do this I offer a positive argument based on the duty to promote autonomous choice and examine competing arguments based on therapeutic privilege, futility and legally accepted standards of disclosure. Having demonstrated that nondisclosure is not justified based on any of these arguments, I will then examine the decision making process in more detail, and propose a model of decision making that is appropriate for LTV decisions for patients with DMD. Borrowing heavily from Katz, I will advance an approach that stresses mutual participation through conversation, incorporates a family-centred approach, and attends to communicative factors that can interfere with shared decision making.

## **Part one: Nondisclosure**

It is generally agreed that a valid informed consent must contain at least the following elements: competency, voluntariness, disclosure of information and patient understanding of that information.<sup>22,23,24,25</sup> Disclosure of information is the cornerstone of virtually all descriptions of informed consent.<sup>23</sup> The survey revealed that 25% of the physicians did not disclose the option of LTV to some of their patients with DMD. In addition, some of the interviewed physicians avoided discussion of progression from night-time to fulltime ventilation.

The duty to promote autonomous choice places a positive demand on health care professional to disclose information regarding treatment options and the potential risks and benefits associated with each. This duty can only be overridden if disclosure would violate a competing moral duty. Thus the physician is morally obligated to reveal LTV information, both night-time and fulltime, to all patients unless it could be demonstrated that doing so would violate another prima facie duty such as beneficence. Other duties to the patient may at times override autonomy making disclosure inappropriate. Below I consider the possible arguments supporting intentional nondisclosure of LTV information, beginning with a discussion of the standards of disclosure.

### *Standards of Disclosure*

A duty to disclose does not necessarily require physicians to reveal all possible information about all possible procedures and the inherent risks and benefits of each.<sup>24,130</sup> As Faden and Beauchamp<sup>131</sup> have observed, such a standard would require inordinately lengthy, often confusing interactions with patients in order to outline all conceivable risks, benefits,

consequences and eventualities for even minor decisions about simple procedures. In such cases overdisclosure is as likely to lead to inadequate understanding as is underdisclosure.

The question of how much information is enough is not always clear. Determining an acceptable standard of disclosure has been debated by the courts and is reflected in current debate in medical ethics.<sup>130</sup> Though these standards though have their origins in law, they are based on ethical considerations and are subject to normative evaluation. I present them here not to determine whether or not it is *legal* to withhold LTV information but to determine if it is *ethical*. The legal categories provide the framework for the discussion.

Traditionally, the courts have relied on a *professional practice standard* in which acceptable disclosure is based on what the community of physicians dealing with similar patients would disclose.<sup>37,130</sup> This standard is based on the belief that physicians, by virtue of their professional qualifications, are in the best position to know which information needs to be disclosed (or withheld) in order to protect or promote a patient's health. Physicians as a professional community thus establish norms of disclosure just as they establish norms of care because no other person is qualified to make such judgements.<sup>132</sup> However, this view has been criticised because it does not allow for a patient's perspective to enter into decision making. Without considering what information the patient may require, informed choice can be undermined rather than supported. The professional practice standard can also be criticised because it provides a description of current practice without any normative evaluation of the medical or moral appropriateness of that practice. It is conceivable that the usual practice that all or most physicians engage in is not the best practice. The common medical practice, in some countries, of turning away patients who cannot pay doesn't necessarily mean that such practice is

morally preferable or best. Some other standard is needed to judge the rightness or wrongness of such actions.

To attempt to address these issues, the professional practice standard is frequently replaced by a *reasonable person standard* in which the physician is obliged to disclose whatever material information a hypothetical reasonable person would wish to know.<sup>37</sup> In Canadian law, this is the prevailing standard of disclosure.<sup>133</sup> The reasonable person standard is thought to be an improvement in that it does not depend solely on accepted practice amongst physicians. The physician is asked to look beyond what options s/he thinks is in the patient's best interests and consider what a reasonable patient in similar circumstances would need to know. Thus a reasonable person standard is sometimes referred to as an objective standard because the criteria for disclosure are not set by the physician's subjective judgements, nor by the patient's subjective judgements, but by objective reference to a hypothetical reasonable person. The reasonable person standard acknowledges that persons require adequate information to be able to participate in decision making around their own care and thus goes further towards facilitating autonomous choice than would the professional practice standard. For the practicing health care professional, the main difficulty with this standard is defining the information needs of the hypothetical reasonable person.

To avoid this problem, a *subjective standard* is proposed as the morally ideal solution.<sup>134</sup> According to this standard, the physician reveals what material information s/he thinks a particular patient would need to know based on that particular patient's values and goals. It has been argued that in reality this is seldom attainable.<sup>134</sup> Rarely do physicians have the opportunity to get to know their patients intimately enough to ascertain their personal value systems well enough to definitively *know* what information they would need or not need in order to make a

decision. It is also questionable whether any one person can know another's needs and thoughts this well without specifically discussing the options at hand. Therefore, the reasonable person standard is often the best that can be expected.

The best overall standard to employ in practice remains the subject of debate. Legally, the reasonable person standard is defended as preferable<sup>37</sup> because it avoids the paternalism inherent in the professional practice standard while at the same time avoiding the practical problems of a subjective standard such as exposing physicians to the "bitter hindsight" of litigious patients. Morally, Beauchamp and Childress<sup>135</sup> argue that the subjective standard is preferable because disclosure is particularized to the individual patient, decreasing the potential for mistaken assumptions inherent in both the reasonable person and the professional practice standards. My purpose is not to debate these points but to examine if *any* of these standards provide a *moral* justification for nondisclosure of LTV information to patients with DMD.

It is doubtful that a professional practice standard justifies withholding ventilation information from patients with DMD. The results of the survey indicate that 75.0% of the physicians disclose LTV information to *all* of their patients. Therefore the prevailing standard of practice seems to be to tell all patients about ventilation regardless of the particulars of a patient's circumstance. Attempts to justify the physicians' nondisclosure based on a professional practice standard are thus dubious at best. Furthermore, the determination that disclosure of LTV information is common practice tells us nothing about the rightness or wrongness of that practice and carries no normative weight. This standard fails to take into account the information needs of patients which may diverge from current professional norms.<sup>136</sup> Thus, even if the current medical standard of practice in Canada were not to disclose LTV information, moral objections could still be raised concerning the appropriateness of such practice.

Is nondisclosure of LTV information morally justified according to a reasonable person standard? To answer, one needs to ascertain whether or not a reasonable person with DMD would need to know about the option of ventilation. Perhaps the case could be made that no reasonable person would want to live life on a ventilator and therefore it need not be mentioned as an option. However, even if a physician believed that the preferred, reasonable *choice* is to accept death rather than life on a ventilator, this still does not preclude *informing* patients that the option to extend life exists. A reasonable person would likely wish to be informed of the option whether or not he would choose that option. Many individuals in the community have chosen ventilation rather than death. Unless we assume that all these individuals (and their physicians) are incompetent, unreasonable people, it seems clear that ventilation is an option about which reasonable persons need to know. Nondisclosure is therefore not supported by the reasonable person standard.

Of the three standards of disclosure discussed, the subjective standard seems the only one where nondisclosure may be justified. Perhaps it may be possible that a physician knows a patient well enough to be sure that the patient would not want or need to know about LTV. If so, not only would the physician have to be convinced that the patient would not choose ventilation, but also that he would not even want to know about the option. However it seems very unlikely that any one person can claim to know another well enough to justify not even *mentioning* a life-saving therapy. Faced with the monumental consequences of an incorrect assumption, it makes intuitive sense to err on the side of caution (life) and disclose. The duty to promote autonomous choice supports this assertion. The best way to ascertain whether or not the patient needs to know about the option of LTV will always be to ask the patient. The patient is thus rightfully given the

responsibility (with the physician's and others' assistance) to choose amongst options that will affect his personal life and health.

We have seen that nondisclosure of LTV is not justified according to the reasonable person or the professional practice standard. In addition, a subjective standard seems ill advised as the consequences of incorrect interpretation of the patient's values are too great. The question of *why* the physician would withhold LTV information is still to be addressed. Two arguments could be advanced to support intentional nondisclosure; the first is based on therapeutic privilege and the second on futility. Each is discussed in turn.

### *Therapeutic privilege*

It is sometimes argued that intentional nondisclosure is justified if revealing information will cause harm to the patient. This practice is referred to as therapeutic privilege.<sup>137</sup> Applebaum uses the example of a patient with an unstable cardiac arrhythmia being asked to consent to its treatment, the explanation of which might heighten anxiety and increase the risk of death.<sup>138</sup> Here strong paternalism is defended as protecting the patient's best interests; avoidance of harm presumably justifies overriding patient autonomy. Information that would be countertherapeutic for a particular patient to receive in a particular circumstance may be justly withheld. Whether or not therapeutic privilege is *ever* morally justified is the subject of debate. Some scholars reject the principle outright as too vague and open to abuse, while others allow that therapeutic privilege may be justified in certain narrowly defined circumstances.<sup>22,37,139,140,141</sup> Relatively minor harms such as causing the patient anxiety are not great enough to justify nondisclosure. As Dickens concludes: "The emotional condition or apprehension of the patient (for the invocation of therapeutic privilege to be justified) must be of an exceptional order and be of a severe if not actual pathological extent."<sup>37</sup>

It seems unlikely that revealing information regarding LTV to patients with DMD fits into this definition. Therapeutic privilege, if ever appropriate, applies only to a narrow range of cases where information sharing *per se* causes significant harm to the patient. There is no conceivable harm related to the disclosure of LTV information that would outweigh the benefit of autonomously choosing whether one lives or dies when one's muscles of respiration fail. While a variety of factors may influence the timing of disclosure, complete avoidance of these discussions is not justified based on therapeutic privilege.

### *Futility*

Nondisclosure of the option of ventilation may be justified if it could be demonstrated that ventilation represents a futile intervention for persons with DMD. Recent considerations of futility in medical practice have arisen out of a concern that some patients are insisting on useless treatments and physicians' decisional authority is being eroded.<sup>142,143</sup> Early in the history of the discussion, Murphy advocated that physicians exercise a strong paternalism to promote patient and family well being by unilaterally withholding information about futile treatments.<sup>144</sup> More recently, Schneiderman et al. have asserted that the emphasis on patient autonomy has "blinded" clinicians and scholars to the validity of other moral obligations to patients.<sup>145,146</sup> They claim "...the power to make judgements about futility is a necessary part of the physician duty of beneficence, that is the duty to use only treatments that provide therapeutic benefits."

The function of the concept of futility is thus to give physicians the moral authority to unilaterally limit life prolonging interventions in some cases, while preserving the rights of patients to decide in other cases.<sup>146,143</sup> Futile therapies need not be offered to patients or provided on patient request. Taylor lists six factors that contribute to the popularity of the concept of medical futility among health care professionals: the conviction that aggressive care is not always in the best interests of the patient and may even be cruel; the concern to preserve the moral integrity of the professionals; the concern to preserve the moral integrity of individual caregivers; the concern for escalating costs; the conviction that harm to a patient constitutes a justifiable limitation to patient or surrogate autonomy; and the conviction that new standards of care are needed for certain groups of patients (such as those in a persistent vegetative state) that would limit life saving interventions.<sup>147</sup>

Solomon has demonstrated that physicians tend to use the term “futility” in multiple and contradictory ways.<sup>148</sup> She found that futility was used both descriptively to denote the efficacy of a treatment and normatively to evaluate the wisdom of pursuing treatment. If futility is to be used normatively to determine where ends the physician's obligation to disclose, defining “futility” is the necessary first step. Futile therapy is therapy that is “useless” or provides no benefit to the patient. Normatively, this means that for any treatment in a given situation, if it is believed that the treatment would provide no or negligible benefit for the patient, the treatment could be considered futile and not offered. There are two problems with this statement. First, “if it is believed” refers to the physician’s judgement of the possible outcomes. This judgement will often involve speculation and will not always be based only on medical facts but may rely on a value judgement. Alternatively, one could substitute the phrase “if it is known that the treatment would provide no or negligible benefit...” but as I will discuss below, it is seldom definitively *known* which interventions will provide benefit. The second problem is what constitutes benefit. Benefits can be assessed objectively (the patient will live for another week) or subjectively (living for another week is a positive outcome). Again, the physician’s value judgements will often come into play. Determining what constitutes a benefit will often involve an evaluative judgement beyond medical or physiological improvement. Does living in pain for one more week constitute a benefit? For a physician to make this decision without consultation with the patient involves unilateral value judgements not only “medical” judgements. These two criticisms are explored in more detail below.

Several authors have offered categorizations of futility that attempt to delineate what constitutes benefit, or more precisely what does *not* count as a benefit (e.g., prolonging the life of a patient in a persistent vegetative state is not beneficial to the patient). These categories describe

criteria for labelling a proposed intervention as futile. They are also normative in that they prescribe action; the physician does not have to offer the intervention if it falls into one of the categories. Baruch Brody and Amir Halevy describe four categories that appear in the literature: 1) physiological, 2) imminent demise, 3) lethal condition and 4) qualitative. *Physiological* futility refers to those interventions where the desired physical effect of treatment is not produced (e.g., prescribing vitamins will not unite a broken bone.) Futility based on *imminent demise* refers to those patients who, regardless of intervention, are only expected to live for a short period, such as days or weeks, with no reversal or improvement in their condition expected with treatment. *Lethal condition* is similar to imminent demise except it refers to those circumstances in which a patient has a lethal incurable condition, and while therapy may prolong life for a short time, (e.g., hours or days), it will not ultimately affect prognosis. According to this criterion, prolonging life for a short time does not constitute a benefit and therefore intervention is futile. Finally *qualitative futility* relies on judgements regarding the quality of life the patient can expect after initiation of the life saving therapy. If predicted quality of life with therapy is thought to be unacceptable, then the physician has no obligation to disclose information regarding that therapy. The terminology here is a little misleading in that it implies that the other types of futility are not “qualitative” or value laden but purely descriptive. I would argue that this is not the case and will expand upon this below. I use the term “qualitative futility” here for convenience, recognizing that it refers to quality of life judgements.

An examination of these four categories reveals that none of them offer purely objective criteria for assessing the “futility” of an intervention. Some sources have suggested that only physiological futility constitutes an acceptable criterion for nondisclosure because it is the only version of futility that is value free (objective), and any other criteria are necessarily value laden

(subjective).<sup>149</sup> However as Griener<sup>143</sup> points out, even physiological futility involves using value judgements. In the majority of situations, there is a lack of definitive objective data that clearly establishes the expected outcome of a proposed intervention. Physicians have to rely on their clinical experience which is shaped in part by their own values. Similar concerns are raised with the clinical application of imminent demise and lethal condition criteria of futility.

In addition, what counts as “imminent” or even “lethal” is problematic. In the qualitative results, a quote was presented wherein a physician referred to DMD as a “terminal disease” (p.77). I would submit that DMD is “lethal but treatable.” A definite benefit arises from treatment, which is the prolongation of life. Individuals with DMD have a progressive neuromuscular disease that is “lethal” at about the age of 20 if it is not treated with LTV. However, once treatment is commenced, barring complications, that individual may live for another ten years or longer. Death cannot be considered “imminent” if the treatment results in several years of life extension. Pneumonia is also a lethal condition if not treated, but it would be absurd to suggest that treatment of pneumonia is futile because the patient might only live another ten years. LTV for DMD cannot be considered a futile therapy based on categories that use the criteria of imminent demise or lethal condition. Furthermore, inherent problems within these criteria make it suspect they can be used to justify routine nondisclosure.

Poor predicted patient quality of life is sometimes used as a justification for nondisclosure of treatment options. If a life-sustaining therapy is known to prolong life (such as LTV for DMD patients) but that life is “believed” to be without meaning or quality, then some would argue that such a therapy does not provide a benefit and is therefore futile. However, as was discussed in the literature review, individual “quality of life” consists of components (enjoyment and satisfaction with life) that cannot be unilaterally assessed by third parties because

they are subjective by nature. For this reason, few professional bodies or scholars of medical ethics support a qualitative test of “futility.”<sup>142</sup> Youngner contends that while physicians may be well placed to initiate and guide discussions about prognosis or potential quality of life, these decisions cannot be made unilaterally. For one patient, a life of extreme disability or pain might be quite tolerable, for another, totally unacceptable.<sup>149</sup> Physicians should not have the authority to decide which lives are not worth living because it is possible that they may inadvertently base decisions on their own values or unconsciously discriminate against the severely physically or mentally disabled. Furthermore whether or not a patient is offered a life sustaining therapy may depend on who their doctor is rather than their individual circumstances.<sup>143</sup>

However, Schneiderman and his colleagues argue that some treatments with qualitatively poor results (treatments that result in a “poor quality of life”) should not be offered to patients. Rather than defining these treatments and circumstances they offer some examples. They contend that the clearest case is that of continued biological life without any conscious autonomy. This position is also advanced in the American Thoracic Society's statement on Withholding and Withdrawing Life-Sustaining Therapy, which states that an intervention is futile if it is unlikely to result in “meaningful survival.” A state of permanent unconsciousness is described as having no value because it lacks any “cognitive or sentient capacity.”<sup>150</sup> Schneiderman et al. give other examples of qualitatively poor results such as conditions requiring constant monitoring, ventilatory support and intensive care nursing. However, they exclude those patients “for whom such care offers the opportunity to achieve life goals, however limited.”<sup>145</sup> Patients who fall into this latter group, they assert, should receive full disclosure about life sustaining therapies.

A qualitative test of futility will necessarily involve a physician making a unilateral judgement of the patient's future quality of life. Because of the value-laden component of such

judgements, qualitative evaluations are either rejected outright or can apply to only a very narrow range of cases such as the patient in a permanent vegetative state. Empirical data presented in the literature review demonstrated that physicians do not always inform patients with DMD about the option of LTV based on unilateral judgements of poor quality of life, beliefs that ventilation is prolonging death or suffering, or beliefs that ventilation is “futile.” In the present study, some physicians have stated they would not offer ventilation because of perceived poor patient quality of life. All of these statements are in essence statements of “qualitative” futility. Physicians are using their unilateral perceptions of patient quality of life as normative criteria for limiting their disclosure of LTV options. However, we have seen in the literature review that many patients report good quality of life even with permanent LTV. In addition, the findings of the present study show that the majority of physicians believe ventilator users with DMD can enjoy good quality of life, although on the questionnaire they were not asked to distinguish between fulltime and night-time LTV. The problem of intersubjective variability inherent in a qualitative evaluation of futility (and quality of life judgements in general) is obvious here. Because quality of life assessments are, at least in part, value dependent, different physicians will have different determinations of what constitutes an acceptable quality of life. It is somewhat controversial in the medical community whether or not patients with DMD can enjoy quality lives while dependent on a ventilator. Nondisclosure based on a determination of futility may seem legitimate to one physician and absurd to another. Even those who would accept qualitative evaluations of futility need to carefully consider Schneiderman's exclusion to this view: Life saving therapies should be offered and provided to those patients for whom the intervention

offers the opportunity to pursue life's goals. I would argue this is a possibility for all\* DMD patients.

Furthermore in cases where this may not be clear, I see no better way to explore the potential quality of life of a patient than to discuss it with him. Once this is acknowledged, there is no need to refer to a concept of futility, as the function of invoking futility, as I have outlined above, was to support the unilateral decisions of physicians. I submit there is no room for such judgements in LTV decisions for patients with DMD and that nondisclosure is not ethically supported by an argument from futility.

Does futility ever justify the limitation of life prolonging interventions? All of the criteria described above rely at least in part on physicians' subjective evaluations. While I am not claiming that there may not be a place for the concept of futility in medical decision making, it appears that any attempt to provide morally acceptable criteria is fraught with difficulties. Careful consideration is needed before establishing any guidelines for unilaterally limiting life prolonging treatments, and the burden of proof lies with those who would favour such a conception.<sup>142</sup>

I have considered futility only from the perspective of the patient-physician relationship and have not considered one of the other cited reasons for invoking futility, which is the proper allocation of resources. It is not my intent in this thesis to prove or disprove that society can afford to provide ventilation for patients with DMD. These decisions will have to be made alongside consideration of other valuable community resources and framed within a general discussion of distributive justice. Two points, however, may help to dispel concerns. First, the

---

\* Except possibly in unusual circumstances. For example, an individual with DMD who is also in a persistent vegetative state as the result of a severe head injury.

physicians in the study did not report limiting access to ventilation based on concern for the proper allocation of resources (only one physician reported this). Second, at present all provincial health plans cover the basic costs of ventilation for patients with DMD. Cost is therefore not a driving issue in the Canadian physicians' practice at present. An examination of LTV in the context of the proper allocation of resources is therefore not considered here but would likely be important to examine in a future inquiry, especially considering the present Canadian health care climate and the drive to cut costs. Long term ventilation would have to be considered alongside of other expensive therapies in an overall analysis of just resource allocation.

### *Conclusion*

The benefit of ventilation is that it prolongs life.<sup>3,9,10</sup> The potential burdens to the family and the quality of life of the patient are likely best determined by the patient and family.<sup>151</sup> Such decisions are value dependent and do not specifically fall within the realm of the medical knowledge. It is unlikely that physicians have the appropriate knowledge and insight to make judgements about the option of ventilation on behalf of families without first discussing it with them at length. It therefore seems unlikely that intentional nondisclosure of the option of ventilation can be supported with an argument based on futility, therapeutic privilege or by any accepted standard of disclosure. Patients with DMD and their families should be informed of the option of ventilation in order to make an informed choice. The clinician's personal opinions regarding quality of life or potential burdens to the family should not preclude disclosure and discussion of the available options. This conclusion is in keeping with the position of The President's Commission for the Study of Ethical Problems in Medicine and Biomedical and Behavioral Research.

Sometimes, however, a patient, though competent, is excluded from the decision making process. This is unjustifiable, since it demeans the patient by barring self determination and allows others to shorten the patients life or to establish a burden under which it will be lived without the assurance (which could be obtained) that the patient concurs in the judgement.<sup>34</sup>

Patients and families need to have information available to them in order to make informed decisions. I have attempted to demonstrate that withholding information about ventilation from individuals with DMD is not morally defensible. However, disclosure alone is not sufficient to support informed choice. Below I consider the decision making process in more detail and defend an approach to LTV decisions that incorporates family centred care and mutual participation through conversation.

## **Part Two: Shared Decision Making: Applications to Long Term Ventilation Decisions**

### *Introduction*

In the literature review, three general models of the patient physician relationship were described: paternalism, consumerism and shared decision making. Each model places different emphasis on the principles of respect for autonomy and beneficence. Paternalism is mainly grounded in the principle of beneficence while consumerism emphasizes patient autonomy. Shared decision making is widely advocated as the model of choice for the majority<sup>152</sup> of patient-physician relationships because it acknowledges and addresses the competing demands placed on a physician to act in the patient's best interests and to promote autonomous choice<sup>34,85,89,153</sup> How to balance these competing duties in practice is a challenge that has been much debated. Several authors have now outlined different models of shared decision making each of which includes its own strategies for balancing autonomy with beneficence. While it may be possible to argue that one of these models is generally preferable to the others and should therefore serve as a paradigmatic reference,<sup>85</sup> I propose instead to take the position that each model may or may not be preferable depending upon the context of the decision making.<sup>154</sup> While mutual participation is the morally preferable *general* approach to decision making in the majority of cases, choosing an appropriate model will vary from situation to situation. The particular characteristics of the physician, the patient, how long they have known one another, the type of decision in question and a range of other factors including religion, culture, and specific family dynamics will influence how shared decision making is approached.<sup>101</sup>

My intent here is to outline an approach to decision making that is appropriate for the majority of individuals with DMD who are facing LTV decisions. To do so I will be borrowing

heavily from Jay Katz's "conversation" model.<sup>153</sup> Katz's model is chosen because his approach, as I will demonstrate, represents a plausible model of decision making for persons with *chronic conditions* who are faced with decisions that can be made *over time*. I will also be augmenting his approach somewhat to include the family in the decision making process and to address potential barriers to communication.

The approach outlined is intended to pertain to the majority of families who are faced with these decisions. However, as I have stated above, a single model will not apply to all cases. In particular, the issue of competency has not been specifically addressed. Throughout the discussion I have assumed that the individual decision makers have the capacity to participate in decision making. Competency is an important factor in the patient-physician relationship and is considered a threshold element of the process of informed consent.<sup>155</sup> I briefly touch on the subject in reference to the child as an emerging decision maker, however with the assumption that the child does not have any cognitive limitations which will prevent him from developing "normally" and achieve competency as an adult. I have not addressed the issue further because to do so in any meaningful way would require a thorough examination that is beyond the purposes of this thesis. It may be helpful to recall that when families are deliberating on decisions together, we can usually assume that at least one of the family decision makers is capable of meaningful participation in the consent process. How to appropriately involve an incompetent family member in decision making is not explored in detail. The approach I outline thus is intended to apply to competent patients and/or parents and may require modification or further specification to be generalized to cases of incompetency.

I will proceed by first elaborating on two general contextual points: the chronic illness experience and the role of the family in decision making. I will then outline and defend Katz's

general approach to decision making. The model will be expanded to address some of the barriers to communication that appear in the literature and are specific to LTV decisions for patients with DMD. Recommendations are then presented in the following chapter.

### *Acute versus chronic illness experiences*

Individuals living with chronic conditions do not approach medical interventions in the same manner as individuals who are acutely ill.<sup>106,156</sup> The acutely ill individual experiences disease as a temporary interruption of one's life story. S/he typically assumes a patient role with concurrent exemption from previous obligations for the duration of the illness.<sup>156</sup> Conversely, the "chronic patient" experiences illness as an integral part of one's identity, affecting life's decisions and goals both on a day to day basis and as part of planning for the future.<sup>157,158,159</sup> The disability is thus an integral part of the life's trajectory rather than the temporary interruption it represents for the acutely ill individual.<sup>106,156</sup> For patients with DMD, we can expand this view of chronic illness to include the family. Not only does the maturing child experience his muscular dystrophy as an integral part of his identity, but living with a chronic condition is integral to the family identity and function as a whole. I will return to this point below.

Since the illness experiences of individuals with acute versus chronic conditions differ fundamentally, it stands to reason that the interactions between physicians and each group of patients may also differ considerably. Many possible future medical interventions can be known to the individual with a chronic illness far in advance of any immediate need to act. This is certainly applicable to ventilation decisions for individuals with DMD. The physician and family literally have years to share their values and preferences, discuss the benefits and burdens of

ventilation, community support, new technological developments, and the patient's evolving values and preferences as he matures.

### *Decision making and the role of the family*

Discussions regarding LTV can occur over time as part of a process of informed consent. However, the final decision to commence or forgo LTV will be imminently required by the time the individual with DMD is between the ages of 16-19.<sup>3</sup> Because informed consent is a process that occurs over time and because ventilation decisions are a part of the family's overall management of the child's illness, a dichotomous view of the patient-physician relationship and informed decision making is inappropriate. The teenager with DMD is unlikely to be considering LTV in isolation from his parents who have been the primary decision makers for most of his life and will potentially bear a large amount of the work of continuing care. Furthermore, in preparation for the relatively momentous LTV decision, the child can be appropriately involved in less consequential decision making throughout his development. These concerns are addressed within a family centred approach that considers the interests of the family as a whole and appropriately involves children in decision making over time.

*Developing decision making capacity.* Fostering the development of the child as a self-determining decision maker is a recognized goal of parenting.<sup>160</sup> "Self determination" here refers to a person's interest in making important decisions based on his own values.<sup>161</sup> As a child grows, he gradually develops the abilities necessary to exercise his increasing independence, first in small ways such as dressing and feeding himself and later to the point of making significant life choices. Through the ongoing process of exercising his self determination, the child begins to establish his own identity and psychological separation from his parents.<sup>162</sup> This is not to suggest

that separation implies a severing of the intimate bonds of familial relationships. The child develops his own identity and life plan but these are also embedded within the identity and goals of the family as a whole. I discuss this point in greater detail below.

King and Cross<sup>160</sup> and others<sup>162,163</sup> take the position that physicians and parents can work together towards the common goal of facilitating the child's self determination through escalating his participation in decision making. They advocate giving children of all ages appropriate information and choices that correspond with their stage of development. Very young children are given inconsequential choices to experience the freedom and responsibility that comes with decision making. As the child matures, the complexity of the information provided is increased as well as his power to decide. Fostering this development seems particularly relevant to children with progressive illnesses who will unavoidably be participating in grave decisions about their health care at a relatively early age.

Appropriately involving the child in decision making requires a careful assessment of his capacity to participate in informed consent. King and Cross<sup>160</sup> describe four interrelated factors: reasoning, understanding, voluntariness and the nature of the decision. The ability to *reason* is dependent on the child's stage of cognitive development.<sup>161,164</sup> Very young children (< 7 years old) will not have the cognitive capacity to weigh more than one alternative simultaneously or engage in abstract forms of reasoning. By the age of twelve, children have reached the final stages of cognitive development (formal operations).<sup>164</sup> This means adolescents (10-12 years old) and teenagers (13-19 years old) are able to engage in the same formal reasoning processes as adults (e.g., the ability to perform complex mental operations such as hypothetical deductive reasoning). Despite this, other necessary abilities may not be fully developed. *Understanding* of illness and the consequences of one's actions depends on the ability to reason, but is also

influenced by the child's past experiences. Children with chronic illnesses are often described as developing insight and understanding at an earlier age than their peers.<sup>160, 165</sup> Any child whose upbringing afforded them a say in family or personal decisions will be better prepared to participate in his own medical decisions compared to a relatively sheltered child. Children with disabilities may develop insight earlier than their peers but they may also have lived more protected and sheltered lives. For an informed choice to be considered valid it must be made *voluntarily*, that is free of coercion or undue manipulation by others. It is characteristic for children to submit to their parents' wishes out of a desire to please and to be viewed favourably. As the child matures, parental influence begins to wane and the child begins to assert his independence. Leiken<sup>162</sup> points out that while older teenagers may be able to make authentic free choices, empirical evidence suggests that adolescents tend to acquiesce to the wishes of authority figures such as physicians, parents and teachers. Finally *the nature of the decision* at hand will dictate the extent of the child's input. Children and teenagers faced with decisions about life-prolonging therapies need to have a mature understanding of life and death and be able to formulate views on their own quality versus quantity of life.<sup>162</sup>

Each child's capacity to participate in informed decision making will need to be assessed in context of the particular family, the child's emerging abilities, and the decision at hand. Harrison and colleagues<sup>163</sup> provide some general guidelines. Primary school aged children can participate by assenting (agreeing to) or dissenting to (refusing) a proposed intervention despite not fully understanding its consequences or being fully capable of making a voluntary choice. The child's input is considered seriously but the parents retain the final authority of consent. Some adolescents and teens may have the decision-making capacity of an adult and can often provide their own consent, but again this will vary from child to child and the nature of the

decision at hand. The adolescent's participation may require some facilitation by team members skilled in communication with children.

*Family-centred decision making.* The potential burdens that parents face when caring for a technology-dependent child are significant.<sup>166</sup> Several sources agree that these decisions which affect the entire family must be considered using a "family-centred approach"<sup>163</sup> which acknowledges the interconnectedness of family members, the potential benefits and burdens for all, and the centrality of the child's special interests.<sup>154,165,166</sup> Christie and Hoffmaster<sup>154</sup> have noted that the recent emphasis on autonomy in medical care may lead to an atomistic moral view that is not appropriate for family decision making. Nelson and Nelson<sup>165</sup> describe the family as having collective mutual goals rather than a conglomeration of individual goals. They conceptualize the family as an ongoing story. Medical decisions are relevant not only in the individual's life plan but also to the broader story of the family as a whole. Because of the interconnectedness of family members, the child's interests and also the interests of all members need to be considered. This is a somewhat natural process for most families who love and care for one another and at times willfully sacrifice their own personal desires for the good of another member or the good of the family as a whole.

Conflict amongst family members may nevertheless occur. Hardwig<sup>167</sup> argues that the interests of the patient in most cases will carry greater weight than others' interests but the burdens to the family must be acknowledged and addressed rather than dismissed as irrelevant. In conflict situations, the physician can assist families by acting as a facilitator who guides the family by emphasizing their interconnectedness, not their individual rights.<sup>165</sup> Rather than only advocate for the patient and possibly entrenching family divisions, the physician works with the family to consider mutually satisfactory options. Arras and Dubler<sup>166</sup> suggest that *accommodation*

and *mediation* be used as strategies to resolve conflict. For example in situations of high tech home care, other care options can be explored if parents feel they cannot manage the child's care at home. Such choices might include care through an appropriate institution, group home setting or independent living with attendant services.

Children with DMD who are appropriately involved with their medical and other decisions as they develop will be better prepared to make choices about LTV in their teenage years. The family's interests in ventilation decisions must also be taken into consideration. An individual with DMD who uses a ventilator requires round the clock care that can impose tremendous burdens on family members. The teenager with DMD who has the capacity to make an autonomous choice about LTV should have his choices respected. In most cases his choice will have been made in consultation with his family and physician and the choice will represent a consensus of the overall interests and desires of the family. Physicians (and others) assist families by presenting a comprehensive picture of the options and the concomitant responsibilities as well as assisting all family members with values clarification. In situations of conflict, physicians may need to assist families by facilitating the exploration of care options.

#### *Katz's approach to shared decision making*

In this section, I discuss Jay Katz's approach to decision making and its application to LTV decisions for individuals with DMD. Katz's model is chosen because it can support the family centred approach previously outlined and lends itself well to any clinical relationship where decision making is an ongoing process that occurs over time. In *The Silent World of Doctor and Patient*, Katz<sup>153</sup> emphasizes the need for *conversation* between physicians and patients. His central theme is summarized in his introduction:

In the absence of any one clear road to well-being, identity of interests cannot be assumed, and consensus on goals, let alone on which paths to follow, can only be accomplished through *conversation* (my italics). Professional considerations, personal value judgements, and self interest decisively influence physicians' pronouncements. Personal values, considerations of life style and other competing preferences influence patients' choices. Conflict within and between parties is inevitable.<sup>168</sup>

Katz takes a Freudian view of self determination, emphasizing the fact that the patient's values are not necessarily known to the patient because of unconscious influences. The physician enters into a dialogue with the patient (or polylogue with the family as appropriate) to help elucidate their values and make treatment choices that are consistent with these. Physicians who are motivated to act in the patient's best interests still cannot know how to meet the patient/family needs without first discussing with the patient and family what those needs are. Put succinctly, "altruism cannot demand dumb trust."<sup>169</sup> He advocates a new definition of "trust", in which identity of interests is not presumed but rather established through conversation. In addition, both parties must acknowledge what they can and cannot contribute to the encounter. Family members bring knowledge about their own needs and physicians bring medical knowledge. Therefore the two relate to each other "as equals and unequals," with the equalities and inequalities complementing each other.

Katz repeatedly asserts that one of the responsibilities of the physician in decision making is to examine her own biases through self reflection. Physicians as well as patients do not make decisions based solely on rational or conscious deliberations. He asserts that while we cannot know all the unconscious, internalized forces that guide us, open discussion can serve to identify underlying assumptions that may not be self evident. Katz stresses that he is not calling for an exploration of the psychological roots of the patient's and physician's motivations, but rather only a "bona fide attempt" to clarify any misconceptions they may have of each others' expectations

and needs.<sup>170</sup> Howard Brody expands on this notion and suggests that what is required of the physician is "transparency."<sup>171</sup> Physicians should essentially make transparent the basis on which a proposed treatment has been chosen. The physician thus reveals not only factual information but the values he attaches to each. As an example related to LTV decisions, the physician may reveal that she does not recommend LTV because she does not feel the patient will be able to enjoy a reasonable quality of life. She may add that she feels strongly about this because she is worried that the family will not be able to cope financially or emotionally with care for the patient at home and that he will eventually end up in an institution. The family members then have the opportunity to question the physician's reasoning, to explore their own values to see if they are in agreement, and to query or confirm the physician's assumption about their coping abilities or the unacceptability of life in an institution. This seems particularly appropriate for the families of individuals with chronic illnesses facing long term decisions who have the unique opportunity of having a series of discussions with their doctors over time.

This model therefore requires explication and interpretation of the values primarily of the patient but also of the physician. Reflection and sharing are required of both parties. The patient's autonomy is supported through a process of clarifying and specifying his values in order to achieve self understanding and come to a decision about medical choices. Families similarly identify their collective and individual goals and values and negotiate (where necessary) between competing values and desires.

*Criticism of Katz's model.* Emanuel and Emanuel have criticized this type of model (which they call an *interpretative model* ) on three fronts.<sup>45</sup> They question whether physicians have 1) the time or 2) the interpretative skills necessary to assist their patients in this task. In addition, 3)

they object to the exclusion of the physician's evaluative judgement as a part of the process. The Emanuels believe Katz's model constrains the guidance that the physician can offer by confining the physician to recommending treatments consistent with the patient's values rather than encouraging the patient to adopt other values. They assert that the moral duty of a caring physician is to encourage some health related behaviours and discourage others in order to promote the patient's overall wellbeing. The authors use examples from preventive medicine to support their argument, pointing out that in practice physicians frequently urge patients to engage in behaviours that lower their risk of developing certain conditions, such as quitting smoking to reduce the risk of heart disease.

Perhaps all three of the Emanuels' criticisms are valid in certain cases. However, as I have stated above, it is not my intention to show that this model is preferable for all patient-physician relationships, but rather that it is preferable for patients with DMD making decisions regarding ventilation. We can assess whether or not these criticisms preclude the use of Katz's model for this particular decision. I have already shown that time should not be a constraint if discussions are initiated early on, so we can quickly dispense with the first criticism.

Some physicians may not have the interpretative skills to help a patient and his family articulate or develop their values surrounding living with a ventilator. However, this does not represent a deficiency with the model but perhaps a deficiency with the physician. If an interpretive model is ethically sound and developing interpretive skills is achievable, should physicians not be encouraged to develop these skills? Again, perhaps it is not necessary or indicated for all physicians, but surely interpretive skills are invaluable to physicians who work with patients with chronic illnesses where lifelong relationships are formed and decision making is an ongoing process. Even if a given physician is unwilling or unable to acquire the necessary

skills, the nurses, social workers, physical therapists and respiratory therapists associated with the clinic are all in a position to help the family clarify their goals. As long as the physician is willing to accept the family's decision, he does not need to be the only professional facilitating decision making or doing the necessary interpretive work. Therefore a lack of interpretive skill on the part of the physicians does not necessitate abandoning Katz's approach.

Finally, the Emanuel's third criticism does not appear to be valid. Katz's model does not exclude physicians' evaluative judgements, nor does it exclude encouraging patients to adopt certain health related values. Indeed, he asserts that it is important for physicians to give specific recommendations, but only in conjunction with the interpretive work outlined above. This approach is demonstrated in the following excerpt from a dialogue between Katz and a patient:

Of course I shall eventually give you my recommendation, but I prefer not to do so yet. Since there are a number of alternatives available, each with its risks and benefits, I would like to hear first what your preferences are. After all it is your body that I intend to treat and can do so in a variety of ways. Since you will have to live with your body for a long time to come, you must have some opinions about which consequences would be easier or more difficult for you to tolerate. Once I have a better idea of your preferences and needs, I can make a recommendation....<sup>172</sup>

Therefore Katz's approach does not exclude encouraging patients to adopt certain behaviours that the physician feels are in the best interest of the patient. For LTV decisions however, there is no clearly right or wrong health related value that the physician should strongly support. The decision being made is less a medical choice and more of a values choice. If the physician gives a recommendation to reject ventilation and states her reasons or values supporting that recommendation, then her actions are consistent with Katz's model.

In addition, Katz' delaying of the recommendation in the above example serves several purposes. First, it properly avoids the misplaced assumption that the doctor can unilaterally know what the best option for the patient and family would be without first hearing what their needs

are. Second, it does not abandon the family to try to choose independently without the advantage of a dialogue in order to discover the best option. Third, it helps to avoid what may be an unconscious tendency of the physician to slip into (strong) paternalism by asserting her own beliefs before the patient speaks. Finally, it insists that the patient and family participate in the decision making process.

A model of decision making that stresses conversation and transparency can serve physicians and families in the pursuit of choosing care options that are consistent with the family's values and individual or collective life plans. Katz provides a feasible framework for decision making but fails to address how the imbalance of power<sup>173</sup> inherent in the physician-patient/family relationship may impede conversation.<sup>174</sup> Because of their power, physicians may either intentionally or unwittingly control conversation through communicative verbal and nonverbal acts with the effect of suppressing the family and patient's input.<sup>175</sup> Katz advocates conversation but does not examine how the discourse itself can unduly influence decisions causing shared decision making to potentially lapse into paternalism. Instead of abandoning the conversation model, further specification is required to address the power differential and its manifestations in communication.

*Power and communication.* Physicians by virtue of their profession have what Brody describes as three kinds of power.<sup>174</sup> *Aesculapian power* is the power of knowledge and medical skill; *social power* is the authority granted to the medical profession by society to control and evaluate its own work. *Charismatic power* is based on the physician's personal qualities such as courage or kindness. These three types of power are employed by the ethical physician to benefit particular patients and/or society as a whole. For example, physicians can use their Aesculapian power to

effect a cure or conduct research, their social power to advocate on a patient's behalf and their charismatic power to convince a child to cooperate with physical examination. Clearly, however, there is also potential for this power to be misused. Physicians who intentionally use their power to control the choices of their competent patients are engaging in a form of strong paternalism. This paternalism can be covert rather than explicit. For example, information can be intentionally framed and presented in such a biased fashion that the patient is left with no real option but to concur with the physician's recommendation. The patient is left with an illusion of being a participant in the process when in actuality he has had little meaningful input. I have already argued above that strong paternalism is ethically inappropriate for patients who have the ability to participate in their decision making and will not pursue it here (see Chapter Two). I wish to focus instead on the physician who believes herself to be involving the family in shared decision making but because of the power discrepancy, unknowingly or unintentionally influences choices and suppresses the participation of family members. Physicians who are dedicated to compassionate care and are committed to the family's comprehension of medical information may not fully apprehend the power they have to sway patient and family choices and how that power can manifest itself in relatively subtle ways. For a conversation model to be effective, the particulars of a conversation must be addressed. Adequate and effective communication will require a true conversation that moves to share power.<sup>176</sup> Communicative factors can influence how patients perceive information and can help or hinder the goal of mutual participation through conversation.

Before proceeding it should be noted that patients also possess power within the relationship. The patient's power rests in his ability to define the medical problem for which he is seeking help, to define his values and goals, and to make a final decision based on his life plan.

This is a much more complex affair in the context of family-physician relationships where power differentials also exist amongst family members. Techniques to appropriately "empower" children consistent with their stage of development have been discussed above. I concentrate here on the differential that exists between the physician and the family as a unit, acknowledging that more than one individual in a family is contributing to the conversation and that power negotiations may concurrently be occurring amongst adult family members (possibly including the patient). Recognizing that the family has some power in the family-physician relationship should not detract from the understanding that the physician's power is almost invariably greater and it is the physician who has the ability to encourage or limit a shared decision making approach from the outset.

Most patient-physician encounters can be described as interviews rather than conversations.<sup>176</sup> In an interview the physician molds the flow of information and tends to control the direction of the exchange and what topics are to be discussed. Through interruptions, the physician can allow or cut off the patient's inquiries. Nonverbal cues can also limit conversation. Brody uses the example of the physician who has her hand on the door knob ready to walk out of the room while asking the patient if he has any questions.<sup>174</sup> The intended message is to encourage patient participation but the perceived message is that questions and further discussion would be bothersome and unwelcome. Waitzkin,<sup>177</sup> in a study of recorded transcripts of physician-patient encounters, found that doctors spent little time providing information to patients (mean of 1.3 minutes), and when asked, grossly overestimated the amount of time they did spend (mean of 8.9 minutes). In addition, doctors underestimated the amount of information patients desire. Empirical studies also show that physicians and patients tend to speak in certain repeated patterns. For example Todd,<sup>178</sup> found that doctors and patients framed their

conversations in different ways - the former in medical, scientific terms and the latter in social or biographical terms. She also found that the doctor's interpretations and reasoning took precedence over the patient's. Janet Farrell Smith<sup>175</sup> argues that because physicians have social authority, a recommendation is sometimes perceived by patients as an imperative command rather than a suggestion or one of many possible options. Patients may perceive what the doctor says as "factual and final" even when it was not intended as such.

Shuy<sup>179</sup> suggests that normal conversation involves a balanced participation; that is, all parties contribute more or less equally to the discourse and the raising of new topics. Farrell Smith calls this "communicative equality" where each person involved in the conversation (two or more) is given an equal chance to speak.<sup>175</sup> Participants respect the abilities of others to make and receive speech acts. Speech acts can include expressive acts of emotion, factual statements, prescriptive statements and conjectures. Conversation allows any participant to open new topical lines, explore or challenge recommendations/assumptions and suggest alternatives.

Speaker assertiveness is generally required for initiating questions and challenging the validity of a claim.<sup>175</sup> Physicians may perceive that patients have little desire for information because they do not ask many questions. However, Brody argues that, not having ever experienced shared decision making, they may just "not know what they're missing." He cites empirical evidence which indicates patients can be taught to be assertive in medical encounters fairly easily and subsequently report greater satisfaction with their care.<sup>180</sup> Physicians can assist the participants in a conversation by attending to their own speech acts and nonverbal acts but also by encouraging the speech acts of others. This could involve pausing in their speech, seeking the participants' opinions, and allowing for control of topic to be shared. The developing child can be encouraged to vocalize his concerns within this relationship by being spoken to

directly as a member of the conversation. This will help prepare him for assertive participation later on.

Because of the physician's Aesculapian power, she has the power to frame information in such a way as to manipulate patients' decisions. I have presented empirical evidence to suggest that physicians at times intentionally frame information for the purpose of controlling outcome (see Chapters 2 and 4). Framing, however, can also be unintentional. In any conversation, a speaker may frame information according to his own biases without necessarily having a full appreciation of competing points of view. This is why exploration of values is an important part of the conversational approach. It is important to ask of oneself and others the value basis of a particular view, and to be cautious of "factual" statements that may actually be value judgements (e.g., one cannot enjoy a good quality of life living in an institution). Reflecting on one's views and considering opposing viewpoints will assist physicians to reduce the influence they have over their patients and reduce unintended framing effects. Reflection is aided through open discussion of views with patients and other professionals.

Influencing families is not in and of itself undesirable or unethical. Attempts to persuade patients to adopt choices felt to be in their best interests is part of beneficent care and is compatible with autonomous choice.<sup>181</sup> However, to be consistent with a conversation model that includes transparency, influence needs to be overtly identified as such whenever possible. Normative statements of the form "You ought to" are better rephrased as "I think you ought to", "You might want to consider" or "If you value X, you might want to consider Y," followed by a rationale or an invitation (not necessarily verbal) to question the statement.

For conversation to work in practice, communicative factors that can either reinforce or reduce the power differential inherent in the physician-patient/family relationship must be

acknowledged and addressed. Here I have attempted to point out some of these factors and suggest some solutions. These suggestions are meant to be suggestions only, intended to reinforce the need for each of us to examine our approaches to patient encounters and recognize our power to sway choices. The ethical professional dedicated to shared decision making will have her own personalized approach to addressing the power imbalance and providing excellent care consistent with the patient and family's values.

### *Summary*

An individual with DMD and his family experience chronic illness as part of their collective story and individual life plans. Because management of the illness is an ongoing process that affects all intimate family members, decision making occurs within a physician-patient-family relationship. I have argued that the child with DMD is appropriately involved with decision making throughout his development, with his authority and responsibility gradually increasing as he matures. The goal of medical decision making for these families is the proper application of medical knowledge to the patient's problems in order to secure an outcome that best aids him in living out his life plan and is also consistent with the family's individual and collective goals. The physician and the family have several years in which to discuss the LTV option, care options and palliation options. Choosing the best option for the patient and family requires an exploration of their goals and values. These may not be readily apparent nor are they likely to be static. The young patient's values will be developing over time. I have argued that Katz's "conversation" model lends itself well to the process of shared decision making for these families. Conversation emphasizes the exploration of the values and assumptions of all

participants, including the physician, so that an acceptable decision can be reached. I have augmented Katz's model by suggesting potential barriers to shared decision making that may be found in the discourse and are the result of the imbalance of power between physicians and families. Acknowledging and attempting to redress these barriers is fundamental to achieving shared decision making through "true" conversations that share power.

## **Chapter 6: Conclusions and Recommendations**

Health care professionals face an ongoing struggle to balance the competing duties to promote the patient's good while respecting autonomous choice. This thesis has considered these competing duties in the context of long term ventilation decisions for patients with Duchenne muscular dystrophy. I have approached the decision making process considering the unique contextual factors of this decision: the chronic nature of the illness, the relatively large amount of time available for decision making, and the need to include family members and the developing child as decision makers.

Throughout this thesis I have argued that the competing duties of beneficence and autonomy are addressed through a shared decision making approach. Shared decision making is defended because it is consistent with the goal of applying medical knowledge to the patient's problems in order to secure an outcome that is consistent with the patient and family's life plan(s). Within a shared decision making approach, beneficence is modified by autonomy because the physician's notion of the good is no longer considered definitive. Instead, patients/families make decisions in consultation with their physicians, searching for a shared notion of the good consistent with the families' values and goals, not merely the medical or biological effects of treatment options. I have advocated a model that approaches decision making through a series of conversations between the physician, patient and family over time. The model includes the following elements: information sharing, values clarification, gradual and appropriate involvement of the developing child, acknowledgement and consideration of the

interests of all family members, and attention to forms of undue or unintentional influence that may be embedded in the discourse.

The empirical results of the research indicate that the majority of physicians are disclosing information about LTV to their patients with DMD and are engaging in discussions over time. However, concern arises in regard to the 25.0% of physicians who reported not disclosing to all patients. An initial disclosure has been defended as the minimum moral requirement of informed consent/decision making. I have argued that disclosure is necessary to fulfill the physician's duty of respect for autonomous choice. I have further argued that, while intentional nondisclosure may sometimes be justified based on arguments from futility, therapeutic privilege or appropriate standards of disclosure, none of these arguments justify nondisclosure in the current context examined in this thesis.

The empirical results suggest that quality of life judgements play a significant role in the physicians' approaches to LTV decision making. Quality of life was cited as a reason for a number of behaviours: both disclosing and not disclosing either night-time or and fulltime ventilation options, encouraging or discouraging LTV, framing information to influence outcome and refusing to participate in initiating LTV if the patient were to end up institutionalized. These results are not unexpected as LTV decisions are by their nature value laden and involve a weighing of benefits and burdens that go beyond physiological effect. Medical benefits (relief of symptoms, extension of life) are considered alongside considerations of potential quality of life. Since the physiological effects of ventilation are uncontested, the decision largely rests on considerations of the burdens that the patient and family will have to endure and whether or not the burdens outweigh the potential benefits. Physicians may have valid concerns about poor patient quality of life and may have serious misgivings about participating in care they feel is

ultimately harmful to the patients and families they care for. I have not advocated that physicians attempt to desist from formulating quality of life judgements. Rather I have attempted to point out empirical evidence from the literature and the present study that demonstrates that health care professionals often make mistaken judgements about quality of life and disability that can influence practice. Unexamined assumptions may influence how physicians present information and in turn may unduly influence patients. I have advocated a conversation model that calls for self reflection and open discussion of underlying assumptions. I see no distinction between night-time and fulltime decisions and therefore have not drawn one in my approach to decision making. While physicians may have personal beliefs regarding potential quality of life with a tracheotomy, fulltime ventilation, or life in an institution, these beliefs should not alter their approach to decision making. The goal remains to choose care and treatment options consistent with the family's values. Through conversation, physicians can help patients and families to define these values and goals as well as make transparent their own thinking, including their assumptions and values regarding LTV options. Once goals are identified and assumptions made transparent, physicians and families can challenge or confirm these assumptions and deliberate together on the best option consistent with the family's values and life plans.

In summary, I have argued that physicians have a positive duty to disclose information about ventilation to families affected by Duchenne muscular dystrophy. Once disclosure is made, a shared decision making approach serves to support autonomous choice and determine the course of action that is best for the patient and his family. I have advocated a conversation model that attends to the potential vulnerability of patients/families by addressing power inequities within the family-physician relationship and incorporates family-centred approach to decision making.

## **Recommendations**

Based on the shared decision making approach defended above and incorporating some of the practices of the physicians in the study, I have formulated the following recommendations for clinicians working with individuals with DMD and their families.

**1. Inform families about the option of ventilation early on.** This can be done at an early follow-up appointment post diagnosis when reviewing the progression of the disease and respiratory sequelae. Mentioning ventilation early on gives families ample time to consider the options and ask questions.

**2. Time ongoing discussion of ventilation according to the family's information needs.**

Information for the child will have to be geared to his cognitive level. Much of the LTV discussion may not begin until the child reaches adolescence and the issues become more pertinent. Families may not always feel comfortable initiating discussions. The clinician may want to introduce the topic at appropriate times. Some key events that may trigger discussion are: the child's first lung function tests, the child's first surgical procedure where he requires ventilation (e.g. tendo- achilles lengthening), scoliosis surgery where there is a definite risk that the child may not be able to be weaned from acute ventilation, and at first sign of respiratory deterioration (age 16-20).

**3. Use a variety of resources to discuss the possible benefits and burdens of ventilation.**

Information can be provided using a variety of methods: written materials discussion, videotapes

and organised discussions with peers. Resources include peers and other families who have faced the decision before and individuals who are currently using a ventilator. Provide opportunities for patients to see and touch the equipment, express their views and ask questions freely. All team members with knowledge to share should participate in the process. Provide information regarding care options, such as home care, group homes, or independent living options. In addition team members should be encouraged to attend conferences on home ventilation such as those organized by Citizens for Independence in Living and Breathing to keep abreast of the latest developments surrounding LTV and meet with ventilator users.

Make clear to the patient and family the possible options for discontinuing ventilation once it has begun. Provide information on the expected sequelae if ventilation is not chosen including palliative care options.

**4. Acknowledge and incorporate the interests of all family members in the decision making process.** A decision to commence LTV or progress from night-time to fulltime ventilation does not necessarily require family members to shoulder the entire burden of care. Across Canada alternate care arrangements exist and vary from province to province. Often home nursing or attendant service is available. Group home arrangements exist in most provinces and older individuals have opportunities to live independently in the community with direct funded attendant service. Physicians or designated team members should educate themselves as to the opportunities in their area.

**5. Discuss and clarify family member values.** A thorough exploration and determination of the family's and child's emerging values can take place over time. The family may have very clear

views about quality of life on a ventilator or these values may require exploration. The teenage child must be encouraged to confront these difficult questions which may be at times be uncomfortable.

**6. Clarify one's own values.** Each clinician must engage in his/her own process of reflection and values clarification to determine why s/he has chosen a specific recommendation.

Recommendations should be based on the stated preferences of the family and the clinician's assessment of whether or not these preferences are in keeping with other stated values of the family. Dialogue should continue until a mutually acceptable decision is reached.

---

## References

1. "In the Matter of Karen Quinlan, An Alleged Incompetent," Supreme Court of New Jersey, March 31, 1976. 70 NJ 10.
2. Gilgoff I.S. (1983) End stage Duchenne patients: Choosing between respirator and natural death, *In* Charash L.I., Ed, *Psychosocial Aspects of Muscular Dystrophy and Allied Diseases*. Charles C. Thomas, Springfield ,IL, 301-307
3. Bach J.R. (1992) Pulmonary rehabilitation considerations for Duchenne muscular dystrophy: The prolongation of life by respiratory muscle aids. *Critical Reviews in Physical and Rehabilitation Medicine* 3(3):239-269.
4. Bach, J.R. (1992) Ventilator Use by Muscular Dystrophy Association Patients. *Archives of Physical Medicine and Rehabilitation* 73,179-183.
5. Colbert A.P.& Schock N.C. (1989) Respirator use in progressive neuromuscular diseases. *Archives of Physical Medicine and Rehabilitation* 70,180-5.
6. Bach, J.R. et al. (1991) Life satisfaction of individuals with Duchenne muscular dystrophy using long term mechanical ventilatory support. *American Journal of Physical Medicine and Rehabilitation* 70, 129-135.
7. Brooke, M.H. et al. (1983) Clinical investigation in duchenne dystrophy: 2. Determination of the "power" of therapeutic trials based on natural history. *Muscle and Nerve* 6,91-103.
8. Hotes, L.S. et al. (1994) Long-term care, rehabilitation, and legal considerations in the management of neuromuscular disease with respiratory dysfunction. *Clinics in Chest Medicine* 15(4), 783-795.
9. Curran J.F. & Colbert A.P. (1989) Ventilator management in Duchenne muscular dystrophy and post poliomyelitis syndrome: Twelve years experience. *Archives of Physical Medicine and Rehabilitation* 70, 180-185.
10. Personal communication with Tom Wagner, Citizens for Independence in Living and Breathing. 1997
11. Schock N.C. & Colbert A.P. (1987) *Ventilators and Muscular Dystrophy*. Gazette International Networking Institute, St.Louis.
12. Arras J.D. & Steinbock B (1995) *Ethical Issues in Modern Medicine: Fourth Edition*. Mayfield Publishing, Mountain View, California, 2.
13. Klemke E.D. et al. (1990) *Philosophy: The Basic Issues, Third Edition*. St. Martins, New York, 16.
14. Beauchamp T.L. & Childress J.F. (1994) *Principles of Biomedical Ethics, 4th edition*. Oxford University Press, New York, 4.
15. Brody H (1992) *In* Crabtree B.F.& Miller W.L. *Doing Qualitative Research*. Sage publications, Newbury Park.

- 
16. Daniels N (1996) *Wide Reflective Equilibrium*, In Sumner L.W. & Boyle J *Philosophical Perspectives on Bioethics*, University of Toronto Press, Toronto.
  17. DeGrazia, D (1992) Moving forward in ethical theory: theories, cases and specified principlism. *The Journal of Medicine and Philosophy* 17, 511-539
  18. Musschenga A.W. (1997) The relation between quality of life, health and happiness. *The Journal of Medicine and Philosophy* 22, 11-28.
  19. Faden, R & Leplege, A, (1992) Assessing quality of life: Moral implications for clinical practice. *Medical Care* 30(5) supplement, MS166-MS175.
  20. Hayry M(1991) Measuring the quality of life: Why, how and what? *Theoretical Medicine* 12, 97-116.
  21. Coy J.A. (1989) Autonomy based informed consent: ethical implications for patient non-compliance. *Physical Therapy* 69(10), 826-833.
  22. Etchells E et al. (1996) Bioethics for clinicians: 1. Consent. *Canadian Medical Association Journal* 155, 177-180
  23. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 145
  24. Macklin R (1987) *Mortal Choices: Bioethics in Today's World*. Pantheon Books, New York, 36
  25. Somerville M.A. (1980) *Consent to medical care: Protection of life series*. Ottawa, Law Reform Commission of Canada, 11.
  26. Lidz C.W., Applebaum P.S.& Meisel A (1988) Two models of implementing informed consent. *Archives of Internal Medicine* 148 (June),1385-89.
  27. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 132.
  28. Reibel v. Hughes (1980) 14 CCLT I (SCC).
  29. Consent to Treatment Act, 1992.
  30. Health Care and Care Facility Act, SBC 1993, c 48.
  31. Consent to Treatment and Health Care Directives Act, Stats PEI 1996, c 10.
  - 32.. Informed Consent: Ethical Considerations for Physicians and Surgeons. (1988) *Annals of the Royal College of Physicians and Surgeons of Canada* 21(1)
  33. Code of Ethics of the Canadian Medical Association (1996) *Canadian Medical Association Journal* 155, 1176A-1176B
  34. The President's Commission for the Study of Ethical Problems in Medicine and Biomedical and Behavioral Research (1983) *Deciding to Forego Life Sustaining Treatment : A Report on the Ethical Medical and Legal Issues in Treatment Decisions*. U.S. Government Printing Office, Washington DC.
  35. Dyer A.R. (1988) Informed consent, autonomy and paternalism: Respecting the patient in research and practice in *Ethics and Psychiatry: Toward Professional Definition*. American Psychiatric Press, Washington, 83-91.

- 
36. Applebaum P.S., Lidz C.W. and Meisel A (1987) *Informed Consent: Legal Theory and Clinical Practice*. Oxford University Press, New York, 12.
37. Dickens B.M. (1985) The doctrine of "informed consent": Informed choice in medical care in Abella R.S. & Rothman M.L., eds., *Justice Beyond Orwell*. Yvon Blais, Montreal, 243-263.
38. Katz J (1984) *The Silent World of Doctor and Patient*. New York, The Free Press, xvi.
39. Faden R.R. and Beauchamp T.L. (1986) *A History and Theory of Informed Consent*. Oxford University Press, New York, 216.
40. Roy D.J., Williams J.R. & Dickens B.M. (1994) *Bioethics in Canada*. Prentice Hall Canada, Scarborough, Ontario, 116-117
41. The National Commission for the Protection of Human Subjects of Biomedical and Behavioral Research (1978) *The Belmont Report: Ethical principles and guidelines for the protection of human subject research*. DHEW Publications no.(OS) 78-0012
42. Thomasma D.C. (1989) The basis of medicine and religion: Respect for persons. In Lammers S.E. & Verhey A, eds. *On Moral Medicine: Theological Perspectives in Bioethics*. William B. Eerdmans, Grand Rapids, 288-292.
43. Purtillo R.B. (1995) Ethical issues in teamwork: The context of rehabilitation In Baylis F et al., eds. *Health Care Ethics in Canada*. Harcourt Brace, Toronto, 191-200.
44. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 56
45. Arras J.D. & Steinbock B (1995) *Ethical Issues in Modern Medicine: Fourth Edition*. Mayfield Publishing, Mountain View, California, 14-17.
46. Beauchamp, T.L. (1982) Philosophical Ethics: An Introduction To Moral Philosophy. McGraw Hill, New York, 107-124.
47. Arras J.D. & Steinbock B (1995) *Ethical Issues in Modern Medicine*, 9.
48. Beauchamp T.L. & McCullough L.B. (1984) Two models of moral responsibility in medicine. *Reprinted in* Mappes T.A. and Zembaty J.S., eds. (1991) *Biomedical Ethics, third ed*. McGraw-Hill, New York, 59-66.
49. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 121.
50. See Macklin R (1987) *Mortal Choices: Bioethics in Today's World*. Pantheon Books, New York, 39-40.
51. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 121-123
52. Faden and Beauchamp (1986) *A History and Theory of Informed Consent*, 235-241.
53. Miller B (1985) In Reich W.T. Ed., *Encyclopedia of Bioethics*. Simon and Schuster MacMillan, New York, 217-218.
54. Faden and Beauchamp (1986) *A History and Theory of Informed Consent*, 238.
55. Faden and Beauchamp (1986) *A History and Theory of Informed Consent*, 239 -240

- 
56. Arras and Steinbock (1995) *Ethical Issues in Modern Medicine*, 15
  57. Purtillo R.B. (1985) In Reich WT Ed., *Encyclopedia of Bioethics*. Simon and Schuster MacMillan, New York, 2096
  58. Pellegrino E.D. and Thomasma D.C. (1988) *For the Patients Good: The Restoration of Beneficence in Health Care*. Oxford University Press, New York, 15.
  59. Pellegrino and Thomasma (1988) *For the Patients Good: The Restoration of Beneficence in Health Care*, 32.
  60. See Faden and Beauchamp's discussion of the Belmont report. Faden and Beauchamp (1986) *A History and Theory of Informed Consent*, 216.
  61. Carse A.L. (1991) The 'voice of care': Implications for bioethical education. *Journal of Medicine and Philosophy* 16, 5-28.
  62. Jecker N.S. (1995) The role of intimate others in medical decision making. In *Health Care Ethics In Canada*, 309-318.
  63. Miller B (1985) In *Encyclopedia of Bioethics*, 218-219.
  64. Wear S (1993) *Informed Consent: Patient autonomy and Physician Beneficence within Clinical Medicine*. Kluwer Academic Publishing, Dordrecht.
  65. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 260.
  66. Frankena, W (1973) *Ethics, 2nd edition*. Prentice-Hall, Englewood Cliffs NJ, 47.
  67. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 272 -273 (quoting the Hippocratic epidemics).
  68. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 272.
  69. Pellegrino and Thomasma (1988) *For the Patients Good: The Restoration of Beneficence in Health Care*.
  70. Loewy E (1989) Doctor patient relationship in *Textbook of Medical Ethics*. Plenum publishing, New York, 60.
  71. Faden and Beauchamp (1986) *A History and Theory of Informed Consent*, 14.
  72. Pellegrino and Thomasma (1988) *For the Patients Good: The Restoration of Beneficence in Health Care*, 29.
  73. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 272-273.
  74. Churchill L (1985) Beneficence. In Reich W.T. ed., *Encyclopedia of Bioethics*. Simon and Schuster MacMillan, New York, 244.
  75. Beauchamp T.L. (1995) Paternalism. In Reich W.T. ed., *Encyclopedia of Bioethics*, Simon and Schuster MacMillan, New York, 1915.
  76. Parsons T (1951) Illness and the role of the physician: A sociological perspective. *Reprinted in Stoeckle J.D. (1987) Encounters Between Patients and Doctors, An Anthology*. The MIT Press, Cambridge, 147-156.

- 
77. Gallagher, E.B. (1976) Lines of reconstruction and extension in the parsonian sociology of illness. *Social Science and Medicine* 10,207-218.
78. Feinberg, J (1971) Legal paternalism. *Canadian Journal of Philosophy* 1,105-124.
79. Pellegrino and Thomasma (1988) *For the Patients Good: The Restoration of Beneficence in Health Care*, 7-8.
80. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 277-278.
81. Smith, D.H. and Veatch, R.M. eds.(1987) *Guidelines on the Termination of Life Sustaining Treatment and the Care of the Dying: A Report by The Hastings Centre*. Indiana University Press, Bloomington.
82. Task Force on Ethics of The Society of Critical Care Medicine (1990) Consensus on the ethics of foregoing life-sustaining treatments in the critically ill. *Critical Care Medicine* 18(12), 1435-1439.
83. Miller, J.R. et al. (1990) Ventilator dependency: decision making, daily functioning and quality of life for patients with duchenne muscular dystrophy. *Developmental Medicine and Child Neurology* 32, 1078-1086.
84. Pellegrino and Thomasma (1988) *For the Patients Good: The Restoration of Beneficence in Health Care*, 7-8.
85. Emanuel E.J. and Emanuel L.L. (1995) Four models of the physician-patient relationship. In Baylis F et al., eds. *Health Care Ethics in Canada*. Harcourt Brace, Toronto, 163-178.
86. Kluge E.H.K (1992) *Biomedical Ethics in a Canadian Context*. Prentice-Hall Canada, Scarborough.
87. Pellegrino and Thomasma (1988) *For the Patients Good: The Restoration of Beneficence in Health Care*, 5.
88. Dejong G (1979) Independent living: From social movement to analytic paradigm. *Archives of Physical Medicine and Rehabilitatio*, 60, 435-446.
89. Veatch R.M. (1972) Models for ethical medicine in a revolutionary age. *Hastings Centre Report* vol. 2 (June), 5-7.
90. *National Strategy for the Integration of Persons with Disabilities: Discussions with the Disability Community on a Future Initiative in Support of Persons With Disabilities* (September 1995) Ottawa: Human Resources Development Canada
91. Baylis F et al., eds. (1995) *Health Care Ethics in Canada*. Harcourt Brace, Toronto, 159.
92. Nelson L and Nelson R(1992) Ethics and the provision of futile, harmful or burdensome treatment to children. *Critical Care Medicine* 20(3), 427-433.
93. Hayry M (1991) Measuring the quality of life: Why, how and what? *Theoretical Medicine* 12, 97-116.
94. I use power here and later in the normative analysis to refer to one's ability to control another.
95. Williams G.H. (1993) The movement for independent living: An evaluation and critique. *Social Science and Medicine* 17(15), 1003-9.
96. Personal communication with Dr. Karen Yoshida, University of Toronto.

- 
97. Meisel A & Kuczewski M (1996) Legal and ethical myths about informed consent. *Archives of Internal Medicine* 156, 2521-2526.
98. Lynn J & Degrazia D (1991) An outcomes model of medical decision making. *Theoretical Medicine* 12, 325-343
99. Brody H (1992) *The Healer's Power*. Yale University Press, New Haven and London.
100. Loewy, E.H. (1989) *Textbook Of Medical Ethics*, 61.
101. Eisenberg JM (1979) Sociological influences on decision making by clinicians. *Annals of Internal Medicine* 90, 957-64.
102. Beausoleil, J.L. et al. (1994) The influence of education and experience on the ethical attitudes in neonatal intensive care. *Medical Decision Making* 14, 403-408.
103. Perry S.W., Schwartz H.I. & Amchin J (1986) Determining resuscitation status: A survey of medical professionals. *General Hospital Psychiatry* 8, 198-202.
104. Sullivan K.E. et al. (1996) What do physicians tell patient with end stage COPD about intubation and mechanical ventilation? *Chest* 109 (January), 258-264.
105. Miller A and Lo B (1985) How do doctors influence discuss do-not-resuscitate orders. *Western Journal of Medicine* 143(2), 256-258.
106. Lidz, C.W. et al. (1983) Barriers to informed consent. *Annals of Internal Medicine* 99, 539-543.
107. Walter J.J. (1985) Quality of Life in Clinical Decisions. In Reich W.T. ed., *Encyclopedia of Bioethics* Simon and Schuster MacMillan, New York, 1352-1356.
108. Drane, J.F. and Coulehan, J.L. (1995) The best interest standard: Surrogate decision making and quality of life. *The Journal of Clinical Ethics* Spring, 20-29.
109. Musschenga A.W. (1997) The relation between quality of life, health and happiness. *The Journal of Medicine and Philosophy* 22, 11-28
110. Musschenga also contends that objective measures are appropriate in some cases of assessing excellence. An important point, but not directly related to this thesis.
111. Draper, P (1992) Quality of life as quality of being: An alternative to the subject-object dichotomy. *Journal of Advanced Nursing* 17, 965-970.
112. Fullbrook, P (1992) Assessing quality of life: the basis for withdrawal of life support treatment? *Journal of Advanced Nursing*. 17, 1440-1446.
113. Gerhart, K.A. et al. (1994) Quality of life following spinal cord injury: Knowledge and attitudes of emergency care providers. *Annals of Emergency Care Medicine* 23(4), 807-812
114. Schneiderman, L.J. (1993) Do physicians' own preferences for life sustaining treatment influence their perceptions of patients' preferences? *The Journal of Clinical Ethics* Spring, 28-33.
115. Quoted in: Fine M and Asch A (1988) Disability beyond stigma: Social interaction, discrimination and activism. *The Journal of Social Issues* 44 (1), 3-21.

- 
116. Hahn H (1988) The politics of physical differences: Disability and discrimination. *The Journal of Social Issues* 44, 39-47.
117. Guillem F, Bombardier C & Beaton D (1993) Cross-Cultural adaptation of health related quality of life measures: Literature review and proposed guidelines. *Journal of Clinical Epidemiology* 46(12), 1417-32.
118. Makas E (1985) *The MIDS (Modified Issues in Disability Scale)*. The George Washington University, Washington, DC.
119. Makas E (1991) In the MIDSt: Modified Issues in Disability Scale testing *In Kiger G & Hey S.C., eds. The Social Organization of Disability Experience*. The Society for Disability Studies, Salem, 109-114.
120. Dillman, D.A. (1978) *Mail and Telephone Surveys: The Total Design Method*. John Wiley and Sons, New York.
121. SPSS for Windows v 6.0.1. Copyright SPSS Inc. 6/26/95
122. Kuzel, A.J. (1992) Sampling in Qualitative Inquiry. *In Crabtree, B.F. & Miller W.L., eds., Doing Qualitative Research*. Sage Publications, Newbury Park, CA, 31-44.
123. The Ethnograph, Version 3.0 for IBM PC, XT, AT and compatibles. Copyright 1988
124. Strauss, A and Corbin J (1990) *Basics of Qualitative Research: Grounded Theory Procedures and Techniques*. Sage Publications, Newbury Park, CA, 57-175.
125. Charmaz K (1990) Discovering Chronic Illness: Using Grounded Theory. *Social Science and Medicine* 30 (11), 1161-1172.
126. Hilton T et al. (1993) End of life care in Duchenne muscular dystrophy. *Paediatric Neurology* 9, 165-77.
127. Makas E. (1993) Yet another year: Further testing on the MIDS. *In Pfeiffer D, Hey SC and Kiger G eds. The Disability Perspective: Variations on a Theme*. The Society for Disability Studies and Willamette University, Salem, Oregon, 143-150.
128. Unpublished data quoted in Makas E. (1993) Yet another year: Further testing on the MIDS
129. Unpublished data quoted in Makas E. (1993) Yet another year: Further testing on the MIDS
130. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 147.
131. Faden and Beauchamp (1986) *A History and Theory of Informed Consent*, 306-7.
132. Faden and Beauchamp (1986) *A History and Theory of Informed Consent*, 30.
133. Etchells E et al. (1996) Bioethics for clinicians: 2. Disclosure, *Canadian Medical Association Journal* 155, 387-391.
134. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 150.
135. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 149.
136. Applebaum (1987) *Informed Consent: Legal Theory and Clinical Practice*, 42.

- 
137. Applebaum (1987) *Informed Consent: Legal Theory and Clinical Practice*, 150-151.
138. Applebaum (1987) *Informed Consent: Legal Theory and Clinical Practice*, 78.
139. Applebaum (1987) *Informed Consent: Legal Theory and Clinical Practice*, 72-78.
140. Wear S (1993) *Informed consent: Patient autonomy and Physician Beneficence within Clinical Medicine*. Kluwer Academic publishing, Dordrecht, 17.
141. Faden and Beauchamp (1986) *A History and Theory of Informed Consent*, 36-38
142. Brody B.A. and Halevy A (1995) Is futility a futile concept? *The Journal of Medicine and Philosophy* 20, 123-144.
143. Griener G (1995) The physician's authority to withhold futile treatment. *The Journal of Medicine and Philosophy* 20, 207-224.
144. Murphy D.J. (1988) Do-not -resuscitate orders: Time for a reappraisal in long term care institutions. *JAMA* 260, 2098-2101
145. Schneiderman L.J., Jecker N.S. & Jonsen A.R. (1990) Medical futility: Its meaning and ethical implications. *Annals of Internal Medicine* 112, 949-954.
146. Schneiderman L.J., Jecker N.S. & Jonsen A.R. (1996) Medical futility: Responses to critiques. *Annals of Internal Medicine* 125, 669-674.
147. Taylor C (1995) Medical futility and nursing. *Image: Journal of Nursing Scholarship* 27(4), 301-306.
148. Solomon M.Z. (1993) How physicians talk about futility: Making words mean too many things. *Journal of Law, Medicine and Ethics* 21, 231-237.
149. Youngner S.J. (1988) Who defines futility? *JAMA* 260, 2094-2095.
150. American Thoracic Society (1991) Withholding and withdrawing life sustaining therapies. *American review of respiratory disease* 144, 726-731.
151. Draper P (1992) Quality of life as quality of being: an alternative to the subject-object dichotomy. *Journal of Advanced Nursing* 17, 965-970.
152. Paternalism or consumerism may be appropriate in some narrowly defined circumstances. For example paternalism may be justified for patients whose autonomy is substantially limited; consumerism may be appropriate for the highly educated, well informed patient whose values and goals are well defined. See chapter 2 for further discussion.
153. Katz J (1984) *The Silent World of Doctor and Patient*. The Free Press, New York.
154. Christie R.J. & Hoffmaster C.B. (1986) *Ethical Issues in Family Medicine*. Oxford University Press, New York, 17-31
155. Beauchamp and Childress (1994) *Principles of Biomedical Ethics*, 145.

- 
156. Gallagher E.B. (1976) Lines of reconstruction and extension in the parsonian sociology of illness. *Social Science and Medicine*, Vol 10, 207-218.
157. Locker D and Kaufert J (1988) The breath of life: Medical technology and the careers of people with post-respiratory poliomyelitis. *Sociology of Health and Illness* 10 (1), 23-40.
158. Strauss A.L. and Glaser B (1975) Managing and shaping the trajectory. In Strauss AL and Glaser B., *Chronic Illness and the Quality of Life*. Mosby, St. Louis, 64-74.
159. Yoshida K.K. (1993) Reshaping of self: a pendular reconstruction of self and identity among adults with traumatic spinal cord injury. *Sociology of Health and Illness* 15(2), 217-45.
160. King N.M.P and Cross A.W. (1989) Children as decision makers: Guidelines for pediatricians. *The Journal of Pediatrics* 115(1), 10-16.
161. Brock D.W. (1989) Children's Competence for Health Care Decision Making. In Kopelman L.M. and Moskop J.C. eds., *Children and Health Care: Moral and Social Issues*. Kluwer Academic Publishing, Dordrecht, 193.
162. Leiken S (1989) A proposal concerning decisions to forgo life-sustaining treatment for young people. *The Journal of Pediatrics* 115(1), 17-21.
163. Harrison C, Kenny N.P., Sidarous M & Rowell M (1997) Bioethics for clinicians 9: Involving children in medical decisions. *Canadian Medical Association Journal* 156 (March 15), 825-8.
164. Grant V (1991) Consent in paediatrics: a complex teaching assignment. *Journal of Medical Ethics* 17, 199-204.
165. Nelson H.L. and Nelson J.L. (1995) *The Patient In The Family: An Ethics Of Medicine And Families* Routledge, New York, 102.
166. Arras J.D. and Dubler N.N. (1994) Bringing the hospital home: Ethical and social implications of high-tech home care. *Special supplement, Hastings Center Report* 24(5), S19-S28.
167. Hardwig J.H. (1990) What about the family? *Hastings Centre Report* March /April, 5-10
168. Katz J (1984) *The Silent World of Doctor and Patient*, .xviii.
169. Katz J (1984) *The Silent World of Doctor and Patient*, .95.
170. Katz J (1984) *The Silent World of Doctor and Patient*, 124.
171. Brody H (1989) Transparency: Informed consent in primary care. *Hastings Center Report*. Sept/Oct, 5-9.
172. Katz J (1984) *The Silent World of Doctor and Patient*, .126.
173. I use "power" here to refer to one's ability to control the actions of another. "Authority" refers to socially legitimated power.
174. Brody H (1992) *The Healer's Power*. Yale University Press, New Haven and London, 105.
175. Farrell Smith J (1996) Communicative Ethics in Medicine: The Physician-Patient Relationship. In Wolf SM ed. *Feminism and Bioethics: Beyond Reproduction*. Oxford University Press, New York, 184-215.

- 
176. West C (1983) 'Ask me no questions': An analysis of queries and replies in physician-patient dialogues *In* Todd A.D. and Fisher S (1993) *The Social Organization of Doctor-Patient Communication, Second Edition*. Ablex Publishing Corporation, New Jersey, 183-209.
177. Waitzkin H (1985) Information giving in medical care. *Journal of Health and Social Behaviour* Vol. 26(June), 81-101.
178. Todd A.D. (1993) A diagnosis of doctor-patient discourse in the prescription of contraception. *In* Todd A.D. and Fisher S, *The Social Organization of Doctor-Patient Communication, Second edition*. Ablex Publishing Corporation, New Jersey, 183-209.
179. Shuy R.W. (1993) Three types of interference to an effective exchange of information in the medical interview. *In* Todd A.D. and Fisher S, *The Social Organization of Doctor-Patient Communication, Second edition*. Ablex Publishing Corporation, New Jersey, 17-30.
180. Greenfield S, Kaplan S., & Ware J.E. (1985) Expanding patient involvement in care: Effects on patient outcomes. *Annals of Internal Medicine* 102, 520-528.
181. Faden and Beauchamp argue that the types of influence that can be compatible with autonomous choice lie on a continuum with coercion on one end, persuasion at the other and manipulation lying somewhere in the middle depending on the form of manipulation. See Faden and Beauchamp (1986) *A History and Theory of Informed Consent*, Chapter 10 Coercion, Manipulation and Persuasion, 337-381.

## **Appendix 1a: English Version of Questionnaire**

### **Ventilation and Duchenne Muscular Dystrophy Questionnaire**

#### **Instructions for completing the questionnaire**

The questionnaire is divided into five sections. Unless otherwise specified, all of the questions relate to patients with Duchenne muscular dystrophy only. When you are answering questions please keep this in mind.

Some of the questions ask for information about practice only, other questions will solicit your opinion on a given issue. It is important that you answer these questions according to your personal beliefs. There are no right or wrong answers. We simply want to know your opinion and what you do in your practice. Remember the answers are completely confidential. There is a section at the end of the survey for your comments.

Throughout the questionnaire you will see information written in bold capitals and contained within square brackets:[**INSTRUCTION**]. The bracketed information is either instruction on answering the question, which question to answer next, or a definition of a key term within the question. Please read the bracketed information carefully, it serves as your guide to answering the questions and proceeding through the survey. It is estimated the survey will take approximately 15 minutes to complete.

**Thank you very much for your participation!**

## **Ventilation and Duchenne Muscular Dystrophy Questionnaire**

### **SECTION A: CLINICAL PRACTICE**

To begin with, we would like to ask some general questions related to the practice in your clinic.

1. Is the patient population at your clinic primarily [CHECK ONE]

- adult  
 paediatric  
 mixed

2. Currently, how many patients with Duchenne muscular dystrophy (DMD) are followed at your clinic? [Check one]

- |                                  |                                  |
|----------------------------------|----------------------------------|
| <input type="checkbox"/> 1 - 10  | <input type="checkbox"/> 41 - 50 |
| <input type="checkbox"/> 11 - 20 | <input type="checkbox"/> 51 - 60 |
| <input type="checkbox"/> 21 - 30 | <input type="checkbox"/> 61 - 70 |
| <input type="checkbox"/> 31 - 40 | <input type="checkbox"/> > 70    |

3. Generally, how frequently do DMD patients attend your clinic for routine clinical review? Patients are seen every \_\_\_\_ months.

4. Generally, how frequently are patients seen when they have known or expected imminent respiratory compromise? Patients are seen every \_\_\_\_ months

[ FOR QUESTION 5, VENTILATORY SUPPORT INCLUDES THE USE OF ANY EQUIPMENT THAT FACILITATES RESPIRATION. THIS INCLUDES NON INVASIVE AND INVASIVE METHODS OF DELIVERY AND ANY AND ALL FORMS OF NEGATIVE OR POSITIVE PRESSURE VENTILATION. VENTILATORY SUPPORT CAN BE REQUIRED FULL TIME OR PART TIME.]

5. Currently how many DMD patients followed at your clinic are receiving ventilatory support?

Number of patients: \_\_\_\_\_ [IF ANSWER IS ZERO, PLEASE GO TO SECTION B]  
 [IF ANSWER IS ONE OR MORE, PLEASE GO TO QUESTION # 6]

6. How many patients with DMD are receiving ventilation 24 hours/day?

Number of patients: \_\_\_\_\_ [IF ANSWER IS ZERO PLEASE GO TO QUESTION # 8]  
 [IF ANSWER IS ONE OR MORE, PLEASE GO TO QUESTION # 7]

7. Which types of ventilatory support are being used by your patients receiving ventilation 24 hours/day? [PLEASE INDICATE WITH A NUMBER]

- a) Iron lung
- b) Chest shell
- c) Pulmo wrap
- d) Bi-level ventilation ("BiPap") via nasal mask
- e) Bi-level ventilation via mouth piece
- f) Bi-level ventilation via lip seal
- g) Volume cycled ventilation via nasal mask
- h) Volume cycled ventilation via mouth piece
- i) Volume cycled ventilation via lip seal
- j) Positive pressure via tracheotomy
- k) Other (please specify) \_\_\_\_\_

8. How many patients with DMD are receiving ventilation for part of the day (less than 24 hours per day)?

Number of patients: \_\_\_\_\_

[IF ANSWER IS ZERO PLEASE GO TO SECTION B]

[IF ANSWER IS ONE OR MORE, PLEASE GO TO QUESTION # 9]

9. Which types of ventilatory support are being used by your patients receiving ventilation for part of the day? [PLEASE INDICATE WITH A NUMBER]

- a) Iron lung
- b) Chest shell
- c) Pulmo wrap
- d) Bi-level ventilation ("BiPap") via nasal mask
- e) Bi-level ventilation via mouth piece
- f) Bi-level ventilation via lip seal
- g) Volume cycled ventilation via nasal mask
- h) Volume cycled ventilation via mouth piece
- i) Volume cycled ventilation via lip seal
- j) Positive pressure via tracheotomy
- k) Other (please specify) \_\_\_\_\_

## SECTION B: FACTORS INFLUENCING INDIVIDUAL PRACTICE

You are finished answering questions about the practice in your clinic. Now we would like to ask you questions related to your **personal interactions** with patients and your **opinions** regarding long term ventilation.

10. How frequently do you tell patients and/or families about the option of long term ventilatory support?

[CHECK THE MOST APPROPRIATE RESPONSE.]

- a) always [GO TO QUESTION #12]
- b) almost always [GO TO QUESTION # 12]
- c) sometimes [GO TO QUESTION # 11]
- d) rarely [GO TO QUESTION # 11]
- e) never [GO TO QUESTION # 13]

11. For those clinicians who answered *sometimes* or *rarely* in question # 10, which of the following reasons might you have for not offering ventilation?

[FOR THIS QUESTION, PLEASE CHOOSE THE REASON(S) FROM THE LIST THAT BEST APPLY TO YOU. IF YOU CHOOSE MORE THAN ONE REASON PLEASE RANK YOUR CHOICES, WITH A NUMBER "1" INDICATING THE MOST IMPORTANT REASON, "2" INDICATING THE SECOND MOST IMPORTANT ETC.]

- a) patients already know about it
- b) inadequacy of patient cognitive functioning
- c) inadequacy of home environment
- d) inadequacy of home care support available
- e) appropriate institutional care is too distant from family
- f) burden on the family
- g) burden on the patient - poor quality of life
- h) inappropriate use of limited health care resources
- i) lack of financial resources of family
- j) ventilation is prolonging the patient's suffering
- k) ventilation is prolonging the patient's death
- l) other (please specify) \_\_\_\_\_

[GO TO QUESTION # 14]

12. For those clinicians who answered *always or almost always* in question #10, which of the following reasons apply to you?

**[FOR THIS QUESTION, PLEASE CHOOSE THE REASON(S) FROM THE LIST THAT BEST APPLY TO YOU. IF YOU CHOOSE MORE THAN ONE REASON PLEASE RANK YOUR CHOICES, WITH A NUMBER "1" INDICATING THE MOST IMPORTANT REASON, "2" INDICATING THE SECOND MOST IMPORTANT ETC.]**

- a) you feel you have a legal obligation to inform the patient of all options  
 b) you feel you have an ethical obligation to inform the patient of all options  
 c) you feel patients and families should make the decision without influence from clinicians  
 d) you feel it is possible for these patients to enjoy a good quality of life on ventilation  
 e) other (please specify) \_\_\_\_\_  
 f) other (please specify) \_\_\_\_\_  
 \_\_\_\_\_  
 \_\_\_\_\_

**[GO TO QUESTION # 14]**

13. For those clinicians who answered *never* in question # 10, which of the following reasons might you have for not offering ventilation?

**[FOR THIS QUESTION, PLEASE CHOOSE THE REASON(S) FROM THE LIST THAT BEST APPLY TO YOU. IF YOU CHOOSE MORE THAN ONE REASON PLEASE RANK YOUR CHOICES, WITH A NUMBER "1" INDICATING THE MOST IMPORTANT REASON, "2" INDICATING THE SECOND MOST IMPORTANT ETC.]**

- a) patients already know about it  
 b) inadequacy of patient cognitive functioning  
 c) inadequacy of home environment  
 d) inadequacy of home care support available  
 e) appropriate institutional care is too distant from family  
 f) burden on the family  
 g) burden on the patient - poor quality of life  
 h) inappropriate use of limited health care resources  
 i) lack of financial resources of family  
 j) ventilation is prolonging the patient's suffering  
 k) ventilation is prolonging the patient's death  
 l) other (please specify) \_\_\_\_\_

**[GO TO SECTION C]**

14. When you do discuss ventilation, when do you normally **first** tell patients and families about ventilation? [Check only one response.]

- a) at first visit
  - b) soon after diagnosis
  - c) only when family/patient asks about it
  - d) when patient develops laboratory signs (ABG's, PFT's) of respiratory compromise
  - e) when patient develops clinical symptoms of respiratory compromise
  - f) when the patient requires ventilation during a hospital admission for surgery
  - g) when the patient is in respiratory failure and requires permanent ventilation to maintain life
  - h) dependent on family situation. (please specify) \_\_\_\_\_
  - i) other (please specify) \_\_\_\_\_
- 

15. When do you normally begin more **in-depth** discussion about ventilation options?

[for example this would include providing the family with written information, peer hook-up, discussions regarding the pro's and cons of the option of ventilation vs palliation etc]  
[Check only one response.]

- a) no in-depth discussion
  - b) at first visit
  - c) soon after diagnosis
  - d) only when family/patient asks about it
  - e) when patient develops laboratory signs (ABG's, PFT's) of respiratory compromise
  - f) when patient develops clinical symptoms of respiratory compromise
  - g) when the patient requires ventilation during a hospital admission for surgery
  - h) when the patient is in respiratory failure and requires permanent ventilation to maintain life
  - i) dependent on family situation (please specify) \_\_\_\_\_
- 

j) other (please specify) \_\_\_\_\_

---

16. When you tell patients about the option of ventilation, what percentage of the time would you say you,

- a. recommend ventilation \_\_\_\_\_ % [if you've written '100' here go to question # 18]  
 b. advise against \_\_\_\_\_ % [if you've written '0' here go to question # 18]  
 c. discuss impartially \_\_\_\_\_ % [if you've written '100' here go to question # 18]
- = 100%

17. When ventilation is not recommended, which of the following reasons apply ?

**[FOR THIS QUESTION, PLEASE CHOOSE THE REASON(S) FROM THE LIST THAT BEST APPLY TO YOU. IF YOU CHOOSE MORE THAN ONE REASON PLEASE RANK YOUR CHOICES, WITH A NUMBER "1" INDICATING THE MOST IMPORTANT REASON, "2" INDICATING THE SECOND MOST IMPORTANT ETC.]**

- \_\_\_\_ a) inadequacy of patient cognitive functioning  
 \_\_\_\_ b) inadequacy of home environment  
 \_\_\_\_ c) inadequacy of home care support available  
 \_\_\_\_ d) appropriate institutional care is too distant from family  
 \_\_\_\_ e) burden on the family  
 \_\_\_\_ f) burden on the patient - poor quality of life  
 \_\_\_\_ g) inappropriate use of limited health care resources  
 \_\_\_\_ h) lack of financial resources of family  
 \_\_\_\_ i) ventilation is prolonging the patient's suffering  
 \_\_\_\_ j) ventilation is prolonging the patient's death  
 \_\_\_\_ k) patient/family unwilling to direct their own care  
 \_\_\_\_ l) other (please specify) \_\_\_\_\_

18. When ventilation is recommended or discussed impartially, which of the following reasons apply,

**[PLEASE CHOOSE THE REASON(S) THAT BEST APPLY TO YOU. IF YOU CHOOSE MORE THAN ONE REASON PLEASE RANK YOUR CHOICES, WITH A NUMBER "1" INDICATING THE MOST IMPORTANT REASON, "2" INDICATING THE SECOND MOST IMPORTANT ETC.]**

- \_\_\_\_ a) you feel you have a legal obligation to inform the patient of all options  
 \_\_\_\_ b) you feel you have an ethical obligation to inform the patient of all options  
 \_\_\_\_ c) you feel patients and families should make the decision without influence from clinicians  
 \_\_\_\_ d) you feel it is possible for these patients to enjoy a good quality of life on ventilation  
 \_\_\_\_ e) other (please specify) \_\_\_\_\_  
 \_\_\_\_ f) other (please specify) \_\_\_\_\_  
 \_\_\_\_\_  
 \_\_\_\_\_

**SECTION C: ALLOCATION OF RESOURCES OPINIONS**

As you know, recently there has been a great deal of discussion regarding issues of appropriate allocation of health care resources. Often such discussions include debate about the appropriate use of expensive technologies, including ventilators. We would like to ascertain your opinions regarding some of the issues surrounding resource allocation. Please remember there are no right or wrong answers and that your answers are completely confidential.

19. In general, do you consider the inappropriate use of medical resources to be a serious, moderate, slight, or no problem at all in your province? **[Check only one response.]**

- a) serious
- b) moderate
- c) slight
- d) no problem at all

20. In general, do you believe physicians consider the public cost of treatment when making treatment decisions in order to control health care expenditures? **[Check only one response.]**

- a) strongly agree
- b) agree
- c) unsure
- d) disagree
- e) strongly disagree

21. Do you feel that in this time of increasingly limited health care resources, society can not afford to pay for providing individuals with DMD long term ventilatory support? **[Check only one response.]**

- a) strongly agree
- b) agree
- c) unsure
- d) disagree
- e) strongly disagree

22. In general, do you feel that your profession needs to establish guidelines to regulate which care options physicians may offer to specific patient groups? **[Check only one response.]**

- a) strongly agree
- b) agree
- c) unsure
- d) disagree
- e) strongly disagree

## SECTION D. MODIFIED ISSUES IN DISABILITY SCALE

The purpose of this section is to gather information on some disability related issues.

Please indicate, using the scale below, your opinion on each of the statements in this section. Although some of these items may appear to be factual, there are really no right or wrong answers. We are simply looking for your opinion (i.e. whether or not you personally agree or disagree with each statement). Therefore, even though you respond "don't know/no opinion," you should use this response only when you have no idea at all what your answer should be. Here is how you should rate the items:

1- strongly disagree		5- somewhat agree
2- disagree	4- don't know/no opinion	6- agree
3- somewhat disagree		7- strongly agree

If you feel a statement is completely true put a "7" in the blank before that particular statement. If you feel the statement is completely false put a "1" in the blank before the statement.

All of the other numbers indicate partial agreement or partial disagreement with these statements. For example, if you consider a statement to be quite true (but not completely true) you should rate it a "6." (A "2" means that it is quite false.) If you feel that it is somewhat true (but not neutral), you should rate it a "5." (A "3" means it is somewhat false.) Use a "4" only if you have absolutely no opinion or absolutely no idea whether it is factually true or false.

Please rate all the items. Also, please make a separate judgement for each item. Do not look back and forth through the statements or try to remember how you rated similar items before.

- \_\_\_ 23. The majority of physically disabled adolescents should attend special schools which are specifically designed to meet their needs.
- \_\_\_ 24. Certain jobs should be set aside for blind persons so that they don't have to compete directly with sighted persons.
- \_\_\_ 25. Disabled children should not have to compete academically with nondisabled children.
- \_\_\_ 26. Most people who have physical disabilities expect no more love and reassurance than anyone else.
- \_\_\_ 27. If you are walking with a blind person, it is easier for him/her to take your arm than for you to take his/her arm.
- \_\_\_ 28. Physically disabled drivers should pay more for their automobile insurance than nondisabled drivers.

1- strongly disagree		5- somewhat agree
2- disagree	4- don't know/no opinion	6- agree
3- somewhat disagree		7- strongly agree

- \_\_\_ 29. It is more humane to allow a child with a severe disability to die at birth than for him/her to live as a severely disabled person.
- \_\_\_ 30. Efforts to place physically disabled people who have been institutionalized back in the community are really pressing them to do more than they are capable of doing.
- \_\_\_ 31. If a person with epilepsy becomes angry with people over little things, it should be overlooked because of his/her disability.
- \_\_\_ 32. Disabled people are generally easier to get along with than nondisabled people.
- \_\_\_ 33. Parents of disabled teenagers should be as strict as other parents.
- \_\_\_ 34. Sheltered workshops (noncompetitive factory work exclusively for disabled people) cannot adequately solve the employment problems of people who happen to be disabled.
- \_\_\_ 35. People with physical disabilities should be expected to meet the same vocational standards as other people.
- \_\_\_ 45. Teachers should not expect students who have epilepsy to participate fully in physical education activities.
- \_\_\_ 46. Trained workers who use wheelchairs are no more likely to have accidents on the job than equally trained nondisabled workers.
- \_\_\_ 47. Disabled people are no more likely than nondisabled people to be churchgoers.
- \_\_\_ 48. Since a physical disability interferes with certain activities, the disability is foremost in a disabled person's mind practically all the time.
- \_\_\_ 49. Blind people tend to get a more accurate first impression of others than most people do.
- \_\_\_ 50. A man or a woman with a physical disability is much more likely than a nondisabled person to have a child who will also have a disability.

**1- strongly disagree**

**5- somewhat agree**

**2- disagree**

**4- don't know/no opinion**

**6- agree**

**3- somewhat disagree**

**7- strongly agree**

- \_\_\_ 51. For a severely disabled person, the kindness of others is more important than an educational program.
- \_\_\_ 52. Disabled people are more accident prone than nondisabled people.
- \_\_\_ 53. Most disabled people would rather socialize with other disabled people than with nondisabled people.
- \_\_\_ 54. Employers' attitudes are a greater handicap to a disabled person than lack of ability.
- \_\_\_ 55. A physically disable high school student will probably feel inadequate in a regular classroom.
- \_\_\_ 56. Physically disabled drivers have more automobile accidents than nondisabled drivers.
- \_\_\_ 57. Disabled people should be expected to fit into our competitive society.
- \_\_\_ 58. It would be much easier for disabled people if they lived in residential units (e.g., apartment buildings) with other disabled people.
- \_\_\_ 59. It is logical for a woman who uses a wheelchair to consider having a baby.

**SECTION E. SOCIODEMOGRAPHIC AND CLINICAL INFO**

You are now finished with the Modified Issues in Disability Scale. The last few questions are just to tell us a little about you.

60. What is your age:\_\_\_\_\_ (years)

61. What is your gender: [CIRCLE]        m        f

62. What is your marital status:

- \_\_\_a) single, never married
- \_\_\_b) married
- \_\_\_c) separated
- \_\_\_d) divorced
- \_\_\_e) widowed

63. What is the number of children you have in each age group. [IF NONE, WRITE "0"]

- \_\_\_a) under 5 years of age
  - \_\_\_b) 5 to 13
  - \_\_\_c) 14 to 18
  - \_\_\_d) 19 to 24
  - \_\_\_e) 25 and over
- For a total of \_\_\_\_\_ children.

64. What is your clinical specialty: [CHECK ONE]

- \_\_\_ a) paediatrics
- \_\_\_ b) neurology
- \_\_\_ c) respirology
- \_\_\_ d) other (specify)\_\_\_\_\_

65. Number of years you have been affiliated with a neuromuscular clinic:

\_\_\_\_\_ years

66. Is your clinical practice 100% devoted to the area of neuromuscular diseases?

- \_\_\_yes [DO NOT ANSWER QUESTION #67]
- \_\_\_no [GO TO QUESTION #67]

67. If you answered no to the previous question, please describe your other areas of clinical practice and the percentage of time you devote to each:

area of practice _____	% of time _____
area of practice _____	% of time _____
area of practice _____	% of time _____
	_____
	= 100 %

Additional comments on any of the items in the questionnaire:

---



---



---



---



---



---



---



---

**Mailing instructions**

Upon completion of the questionnaire, please check to make sure you have responded to all the questions. Please return the questionnaire using the addressed, stamped envelope enclosed. There is a postcard enclosed that you can mail separately if you wish a summary of the results of the survey to be sent to you.

**\*PLEASE NOTE**

As part of this research, we are also conducting interviews with physicians who follow patients with DMD. The interviews are intended to help us gain a deeper understanding of some of the complex issues found in the questionnaire and to hear more about physicians' views.

We will be arranging interviews at a time and place convenient to our respondents. If you are interested in participating, please check "yes" on the enclosed post card and send it separately.

**THANK YOU VERY MUCH FOR YOUR PARTICIPATION**

## **Appendix 1b: French Version of Questionnaire**

### **Questionnaire sur la ventilation assistée pour les patients atteints de dystrophie musculaire de type Duchenne**

#### **Instructions pour compléter le questionnaire**

Ce questionnaire est divisé en cinq parties. Sauf indication contraire, toutes les questions concernent des patients atteints uniquement de dystrophie musculaire de type Duchenne. N'oubliez pas cette précision en répondant aux questions.

Certaines questions portent uniquement sur la pratique, d'autres visent à obtenir votre opinion sur une question précise. Il est important que vous répondiez à ces questions selon vos convictions personnelles. Il n'y a ni bonne, ni mauvaise réponse. Nous voulons simplement connaître votre opinion et savoir comment vous agissez dans votre pratique. N'oubliez pas que ces réponses sont entièrement confidentielles. A la fin du questionnaire, vous trouverez une section pour indiquer vos commentaires.

Tout au long du questionnaire vous trouverez des informations en lettres majuscules, en caractère gras et entre crochets: **[INSTRUCTION]**. Ces informations fournissent des instructions sur la manière de répondre à la question, vous indiquent la prochaine question à laquelle vous devez répondre ou encore précisent la définition d'un terme-clé utilisé dans la question. Veuillez lire attentivement ces instructions, destinées à guider vos réponses aux questions et à progresser dans le questionnaire. Nous estimons qu'il faut environ 15 minutes pour compléter ce questionnaire.

**Merci beaucoup de votre participation!**

**Questionnaire sur la ventilation assistée pour les patients atteints de dystrophie musculaire de type Duchenne**

**PARTIE A: PRATIQUE CLINIQUE**

Pour commencer, nous aimerions vous poser quelques questions d'ordre général sur la pratique dans votre clinique.

1. Votre clinique reçoit-elle surtout une clientèle [COCHEZ UNE RÉPONSE]

- \_\_\_ adulte  
 \_\_\_ pédiatrique  
 \_\_\_ mixte

2. Actuellement, combien de patients atteints de dystrophie musculaire de type Duchenne (DD) sont suivis dans votre clinique? [COCHEZ UNE RÉPONSE]

- |             |             |
|-------------|-------------|
| ___ 1 - 10  | ___ 41 - 50 |
| ___ 11 - 20 | ___ 51 - 60 |
| ___ 21 - 30 | ___ 61 - 70 |
| ___ 31 - 40 | ___ > 70    |

3. En règle générale, à quelle fréquence les patients atteints de DD se présentent-ils à votre clinique pour un examen de routine? Ces patients sont examinés tous les \_\_\_ mois.

4. En règle générale, à quelle fréquence examinez-vous les patients qui présentent un risque respiratoire imminent connu ou prévisible? Ces patients sont examinés tous les \_\_\_ mois.

**[POUR LA QUESTION 5, LA VENTILATION ASSISTÉE INCLUT LE RECOURS À TOUT ÉQUIPEMENT QUI FACILITE LA RESPIRATION. ELLE ENGLOBE TOUTES LES MÉTHODES INVASIVES ET NON-INVASIVES D'ASSISTANCE ET TOUTES LES AUTRES FORMES DE VENTILATION PAR PRESSION NÉGATIVE OU POSITIVE. LA VENTILATION ASSISTÉE PEUT S'AVÉRER NÉCESSAIRE DE MANIÈRE CONTINUE OU À TEMPS PARTIEL.]**

5. Actuellement, parmi les patients atteints de DD suivis dans votre clinique, combien bénéficient de ventilation assistée?

Nombre de patients: \_\_\_\_\_ [SI LA RÉPONSE EST 0, PASSEZ À LA PARTIE B]  
 [SI LA RÉPONSE EST 1 OU PLUS, PASSEZ À LA QUESTION # 6]

6. Combien de patients atteints de DD bénéficient d'une ventilation assistée, 24 heures sur 24?

Nombre de patients: \_\_\_\_\_ [SI LA RÉPONSE EST 0, PASSEZ À LA QUESTION # 8]  
 [SI LA RÉPONSE EST 1 OU PLUS, PASSEZ À LA QUESTION # 7]

7. Quel type de ventilation assistée est fournie aux patients qui en bénéficient 24 heures sur 24?

**[VEUILLEZ RÉPONDRE PAR UN NOMBRE]**

- a) Poumon d'acier
  - b) Coque thoracique
  - c) "pulmo wrap"
  - d) Ventilation à double niveau ("BiPap") par masque nasal
  - e) Ventilation à double niveau ("BiPap") par appareil buccal
  - f) Ventilation à double niveau ("BiPap") par obturation labiale
  - g) Ventilation du volume par masque nasal
  - h) Ventilation du volume par appareil buccal
  - i) Ventilation du volume par obturation labiale
  - j) Pression positive par trachéotomie
  - k) Autres (veuillez préciser)
- 
- 

8. Combien de patients atteints de DD bénéficient de ventilation à temps partiel (moins de 24 heures par jour)? Nombre de patients: \_\_\_\_ [SI LA RÉPONSE EST 0, PASSEZ À LA PARTIE B]

[SI LA RÉPONSE EST 1 OU PLUS, PASSEZ À LA QUESTION # 9]

9. Quel type de ventilation assistée est fourni aux patients qui en bénéficient à temps partiel?

**[VEUILLEZ RÉPONDRE PAR UN NOMBRE]**

- a) Poumon d'acier
  - b) Coque thoracique
  - c) "pulmo wrap"
  - d) Ventilation à double niveau ("BiPap") par masque nasal
  - e) Ventilation à double niveau ("BiPap") par appareil buccal
  - f) Ventilation à double niveau ("BiPap") par obturation labiale
  - g) Ventilation du volume par masque nasal
  - h) Ventilation du volume par appareil buccal
  - i) Ventilation du volume par obturation labiale
  - j) Pression positive par trachéotomie
  - k) Autres (veuillez préciser)
-

## PARTIE B: FACTEURS QUI INFLUENCENT VOTRE PRATIQUE PERSONNELLE

Nous aimerions maintenant vous poser des questions sur vos **interactions personnelles** avec ces patients et connaître votre **opinion** sur la ventilation à long terme.

10. À quelle fréquence mentionnez-vous aux patients et/ou à leurs familles l'option de la ventilation assistée à long terme? [COCHEZ LA RÉPONSE LA PLUS APPROPRIÉE.]

- a) toujours [PASSEZ À LA QUESTION # 12]
- b) presque toujours [PASSEZ À LA QUESTION # 12]
- c) parfois [PASSEZ À LA QUESTION # 11]
- d) rarement [PASSEZ À LA QUESTION # 11]
- e) jamais [PASSEZ À LA QUESTION # 13]

11. Pour les cliniciens qui ont répondu *parfois ou rarement* à la question # 10, parmi les raisons suivantes, laquelle invoqueriez-vous pour ne pas proposer la ventilation?

[POUR CETTE QUESTION, VEUILLEZ CHOISIR DANS LA LISTE LA OU LES RAISON(S) QUI CORRESPOND(ENT) LE MIEUX À VOTRE CAS. SI VOUS CHOISISSEZ PLUS D'UNE RAISON, VEUILLEZ CLASSER VOS RÉPONSES EN INDIQUANT LE CHIFFRE "1" POUR LA RAISON LA PLUS IMPORTANTE, LE CHIFFRE "2" POUR LA DEUXIEME RAISON ETC.]

- a) les patients sont déjà au courant
- b) fonctionnement cognitif du patient est insuffisant
- c) environnement inadéquat au domicile du patient
- d) disponibilité des soins à domicile limitée
- e) des soins institutionnels appropriés sont trop éloignés de la famille
- f) fardeau pour la famille
- g) fardeau pour le patient - qualité de vie médiocre
- h) utilisation inappropriée de ressources en soins de santé limitées
- i) manque de ressources financières dans la famille
- j) la ventilation prolonge les souffrances du patient
- k) la ventilation prolonge l'agonie du patient
- l) autres (veuillez préciser) \_\_\_\_\_

[PASSEZ À LA QUESTION #14]

12. Pour les cliniciens qui ont répondu *toujours ou presque toujours* à la question # 10, parmi les raisons suivantes, laquelle correspond à votre cas?

**[POUR CETTE QUESTION, VEUILLEZ CHOISIR DANS LA LISTE LA OU LES RAISON(S) QUI CORRESPOND(ENT) LE MIEUX À VOTRE CAS. SI VOUS CHOISISSEZ PLUS D'UNE RAISON, VEUILLEZ CLASSER VOS RÉPONSES EN INDIQUANT LE CHIFFRE "1" POUR LA RAISON LA PLUS IMPORTANTE, LE CHIFFRE "2" POUR LA DEUXIEME RAISON ETC.]**

- a) vous estimez avoir l'obligation légale d'informer le patient de toutes les options
- b) vous estimez avoir l'obligation morale d'informer le patient de toutes les options
- c) vous estimez que les patients et leurs familles devraient prendre la décision sans l'influence des cliniciens
- d) vous pensez que la ventilation peut offrir une bonne qualité de vie à ces patients
- e) autres (veuillez préciser) \_\_\_\_\_
- f) autres (veuillez préciser) \_\_\_\_\_

**[PASSEZ À LA QUESTION # 14]**

13. Pour les cliniciens qui ont répondu *jamais* à la question # 10, parmi les raisons suivantes, laquelle vous amène à ne pas proposer la ventilation?

**[POUR CETTE QUESTION, VEUILLEZ CHOISIR DANS LA LISTE LA OU LES RAISON(S) QUI CORRESPOND(ENT) LE MIEUX À VOTRE CAS. SI VOUS CHOISISSEZ PLUS D'UNE RAISON, VEUILLEZ CLASSER VOS RÉPONSES EN INDIQUANT LE CHIFFRE "1" POUR LA RAISON LA PLUS IMPORTANTE, LE CHIFFRE "2" POUR LA DEUXIEME RAISON ETC.]**

- a) les patients sont déjà au courant
- b) fonctionnement cognitif du patient est insuffisant
- c) environnement inadéquat au domicile du patient
- d) disponibilité des soins à domicile limitée
- e) des soins institutionnels appropriés sont trop éloignés de la famille
- f) fardeau pour la famille
- g) fardeau pour le patient - qualité de vie médiocre
- h) utilisation inappropriée de ressources en soins de santé limitées
- i) manque de ressources financières dans la famille
- j) la ventilation prolonge les souffrances du patient
- k) la ventilation prolonge l'agonie du patient
- l) autres \_\_\_\_\_

**[PASSEZ À LA PARTIE C]**

14. Lorsque vous parlez de la ventilation, normalement quand la mentionnez-vous pour la première fois aux patients et à leurs familles? [Cochez une seule réponse.]

- a) à la première visite
- b) peu après le diagnostic
- c) seulement si la famille/le patient pose la question
- d) lorsque le patient présente en laboratoire des signes de risque respiratoire (les ABG, les fonctions pulmonaires)
- e) lorsque le patient présente des symptômes cliniques de risque respiratoire
- f) lorsque le patient a besoin de ventilation lors de son admission à l'hôpital pour une intervention chirurgicale
- g) lorsque le patient souffre d'insuffisance respiratoire et requiert une ventilation continue pour rester en vie
- h) selon la situation familiale (veuillez préciser)

---

j) autres (veuillez préciser)

---

15. Normalement, à quel moment entamez-vous une discussion **plus approfondie** sur les options de ventilation?

**[par exemple, en proposant à la famille des informations écrites sur le sujet, les coordonnées de groupes de soutien, par des discussions sur les avantages et les inconvénients de la ventilation par rapport à l'option palliative, etc]**

**[Cochez une seule réponse.]**

- a) je n'en parle pas en détail
- b) à la première visite
- c) peu après le diagnostic
- d) seulement si la famille/le patient pose la question
- e) lorsque le patient présente en laboratoire des signes de risque respiratoire (les ABG, les fonctions pulmonaires)
- f) lorsque le patient présente des symptômes cliniques de risque respiratoire
- g) lorsque le patient a besoin de ventilation lors de son admission à l'hôpital pour une intervention chirurgicale
- h) lorsque le patient souffre d'insuffisance respiratoire et requiert une ventilation continue pour rester en vie
- i) selon la situation familiale (veuillez préciser)

---

j) autres. Veuillez préciser (veuillez préciser)

---

16. Lorsque vous mentionnez l'option de la ventilation à vos patients, dans quel pourcentage des cas,

- a. la recommandez-vous \_\_\_\_\_% [si vous indiquez 100, passer à la question # 18]  
 b. la déconseillez-vous \_\_\_\_\_% [si vous indiquez 0, passer à la question # 18]  
 c. en discutez-vous impartialement \_\_\_\_\_% [si vous indiquez 100, passer à la question # 18]  
 = 100%

17. Lorsque la ventilation n'est pas recommandée, laquelle des raisons suivantes s'applique?  
 [VEUILLEZ CHOISIR DANS LA LISTE LA OU LES RAISON(S) QUI CORRESPOND(ENT) LE MIEUX À VOTRE CAS. SI VOUS CHOISISSEZ PLUS D'UNE RAISON, VEUILLEZ CLASSER VOS RÉPONSES EN INDIQUANT LE CHIFFRE "1" POUR LA RAISON LA PLUS IMPORTANTE, LE CHIFFRE "2" POUR LA DEUXIEME RAISON ETC.]

- \_\_\_\_ a) fonctionnement cognitif du patient est insuffisant  
 \_\_\_\_ b) environnement inadéquat au domicile du patient  
 \_\_\_\_ c) disponibilité des soins à domicile limitée  
 \_\_\_\_ d) des soins institutionnels appropriés sont trop éloignés de la famille  
 \_\_\_\_ e) fardeau pour la famille  
 \_\_\_\_ f) fardeau pour le patient - qualité de vie médiocre  
 \_\_\_\_ g) utilisation inappropriée de ressources en soins de santé limitées  
 \_\_\_\_ h) manque de ressources financières dans la famille  
 \_\_\_\_ i) la ventilation prolonge les souffrances du patient  
 \_\_\_\_ j) la ventilation prolonge l'agonie du patient  
 \_\_\_\_ k) la famille\le patient ne souhaite prendre le contrôle de leurs soins  
 \_\_\_\_ l) autres (veuillez préciser) \_\_\_\_\_

18. Lorsque la ventilation est recommandée ou discutée impartialement, laquelle des raisons suivantes s'applique?

[VEUILLEZ CHOISIR DANS LA LISTE LA OU LES RAISON(S) QUI CORRESPOND(ENT) LE MIEUX À VOTRE CAS. SI VOUS CHOISISSEZ PLUS D'UNE RAISON, VEUILLEZ CLASSER VOS RÉPONSES EN INDIQUANT LE CHIFFRE "1" POUR LA RAISON LA PLUS IMPORTANTE, LE CHIFFRE "2" POUR LA DEUXIEME RAISON ETC.]

- \_\_\_\_ a) vous estimez avoir l'obligation légale d'informer le patient de toutes les options  
 \_\_\_\_ b) vous estimez avoir l'obligation morale d'informer le patient de toutes les options  
 \_\_\_\_ c) vous estimez que les patients et leurs familles devraient prendre la décision sans l'influence des cliniciens  
 \_\_\_\_ d) vous pensez que la ventilation peut offrir une bonne qualité de vie à ces patients  
 \_\_\_\_ e) autres.(veuillez préciser) \_\_\_\_\_  
 \_\_\_\_ f) autres (veuillez préciser) \_\_\_\_\_

### **PARTIE C: OPINIONS SUR LA RÉPARTITION DES RESSOURCES**

Comme vous le savez, depuis quelque temps, la répartition appropriée des ressources en soins de santé fait l'objet d'un débat majeur. Ce débat comporte souvent des discussions sur l'utilisation appropriée des technologies coûteuses, notamment les ventilateurs. Nous aimerions connaître vos opinions sur certaines questions liées à la répartition des ressources. Encore une fois, il n'y a ni bonne, ni mauvaise réponse et vos réponses sont entièrement confidentielles.

19. De façon générale, l'utilisation inappropriée des ressources médicales constitue, selon vous, un problème grave, modéré, mineur ou aucun problème du tout dans votre province? [Cochez une seule réponse.]

- a) grave
- b) modéré
- c) mineur
- d) aucun problème du tout

20. De façon générale, croyez-vous que dans leur choix de traitement, les médecins tiennent compte du coût public du traitement afin de contrôler les dépenses en soins de santé? [Cochez une seule réponse.]

- a) tout à fait d'accord
- b) d'accord
- c) incertain
- d) en désaccord
- e) fermement en désaccord

21. À une époque de restriction dans les ressources en soins de santé, croyez-vous que la société ne peut plus se permettre d'offrir une ventilation assistée à long terme aux patients atteints de DD? [Cochez une seule réponse.]

- a) tout à fait d'accord
- b) d'accord
- c) incertain
- d) en désaccord
- e) fermement en désaccord

22. De façon générale, pensez-vous que votre profession doit établir des lignes directrices pour réglementer les options de soins que les médecins peuvent offrir à certains groupes de patients particuliers? [Cochez une seule réponse.]

- a) tout à fait d'accord
- b) d'accord
- c) incertain
- d) en désaccord
- e) fermement en désaccord

## PARTIE D: PROBLEMES MODIFIÉS PAR LE DEGRÉ D'INCAPACITÉ

Dans cette partie, nous voulons recueillir des informations sur certaines questions reliées aux incapacités.

**Veillez donner votre opinion sur chacune des affirmations contenues dans cette partie, à l'aide de l'échelle ci-dessous. Même si certaines questions peuvent vous sembler de nature factuelle, il n'y a vraiment ni bonne ni mauvaise réponse. Nous cherchons simplement à connaître votre opinion (c'est à dire si vous êtes personnellement d'accord ou en désaccord avec ces affirmations). Par conséquent même si vous avez la possibilité de répondre "ne sait pas/sans opinion", vous devriez utiliser cette réponse uniquement lorsque vous n'avez absolument aucune idée de la réponse. Voici comment classer les affirmations:**

1 - fermement en désaccord		5 - plutôt d'accord
2 - en désaccord	4 - ne sait pas/sans opinion	6 - d'accord
3 - plutôt en désaccord		7 - fortement d'accord

Si vous considérez une affirmation entièrement vraie, mettez un "7" dans l'espace qui précède cette affirmation. Si vous considérez une affirmation entièrement fausse, mettez un "1" dans l'espace qui précède cette affirmation.

Tous les autres chiffres indiquent un accord partiel ou un désaccord partiel avec ces affirmations. Par exemple, si vous considérez qu'une affirmation est largement vraie (mais pas entièrement vraie), vous lui mettez un "6." (un "2" correspond à une affirmation largement fausse). Si vous trouvez l'affirmation plutôt vraie (mais pas neutre), vous lui mettez un "5." (un "3" correspond à une affirmation plutôt fausse). Utilisez le "4" seulement lorsque vous n'avez absolument aucune opinion ou aucune idée si les faits sont vrais ou faux.

Nous vous demandons de noter toutes les affirmations et d'envisager chacune individuellement. N'essayez pas de comparer les affirmations et ne tentez pas de vous souvenir comment vous avez noté des affirmations similaires.

- \_\_\_23. La majorité des adolescents handicapés physiques devraient fréquenter des écoles spéciales, aménagées spécialement pour répondre à leurs besoins.
- \_\_\_24. Il faudrait réserver certains emplois aux personnes aveugles afin qu'elles n'aient pas à se retrouver directement en concurrence avec les voyants.
- \_\_\_25. Les enfants handicapés ne devraient pas être en concurrence sur le plan scolaire avec des enfants non-handicapés.
- \_\_\_26. La plupart des personnes handicapées physiques ne s'attendent pas à recevoir d'amour ou de réconfort que les autres.

<b>1 - fermement en désaccord</b>		<b>5 - plutôt d'accord</b>
<b>2 - en désaccord</b>	<b>4 - ne sait pas/sans opinion</b>	<b>6 - d'accord</b>
<b>3 - plutôt en désaccord</b>		<b>7 - fortement d'accord</b>

- \_\_\_27. Si vous marchez avec une personne aveugle, il lui est plus facile de prendre votre bras que de vous donner le sien.
- \_\_\_28. Les personnes handicapées physiques qui conduisent devraient payer des assurances automobiles plus élevées que les autres conducteurs.
- \_\_\_29. Il est plus humain de laisser mourir à la naissance un enfant atteint d'une déficience grave que de le laisser vivre comme une personne gravement handicapée.
- \_\_\_30. Les efforts de réintégration dans la communauté des personnes handicapées physiques provenant d'institutions, les obligent en fait à faire plus qu'elles ne sont capables de faire.
- \_\_\_31. Si une personne atteinte d'épilepsie se met en colère contre d'autres personnes à propos de détails, il faut ignorer sa réaction en raison de son incapacité.
- \_\_\_32. En règle générale, les personnes handicapées sont plus faciles à vivre que les autres.
- \_\_\_33. Les parents d'adolescents handicapés devraient être aussi stricts que les autres parents.
- \_\_\_34. Les ateliers protégés (travail dans des établissements non concurrentiels réservés exclusivement aux personnes handicapées) ne peuvent résoudre adéquatement les problèmes d'emploi des personnes handicapées.
- \_\_\_35. On devrait exiger des personnes handicapées physiques qu'elles atteignent les mêmes normes professionnelles que les autres travailleurs.
- \_\_\_36. Les personnes handicapées physiques sont souvent des gens faciles à vivre et qui se fâchent rarement.
- \_\_\_37. Il faut éviter de poser aux personnes handicapées des questions sur leur incapacités.

<b>1 - fermement en désaccord</b>		<b>5 - plutôt d'accord</b>
<b>2 - en désaccord</b>	<b>4 - ne sait pas/sans opinion</b>	<b>6 - d'accord</b>
<b>3 - plutôt en désaccord</b>		<b>7 - fortement d'accord</b>

- \_\_\_38. Les personnes handicapées n'ont pas suffisamment d'influence en politique.
- \_\_\_39. Le revenu tiré des impôts versés par une personne handicapée qui occupe un emploi est supérieur au montant dépensé pour remettre cette personne au travail.
- \_\_\_40. Les personnes en fauteuils roulants ont souvent des "petits accidents" (c.a.d. qu'elles n'arrivent pas à temps aux toilettes).
- \_\_\_41. Les programmes éducatifs pour étudiants handicapés physiques sont très coûteux par rapport à ce qu'en retirent les enfants handicapés physiques.
- \_\_\_42. Vous devez faire très attention à ce que vous dites lorsque vous parlez avec des personnes handicapées physiques.
- \_\_\_43. En règle générale, les personnes handicapées ne sont pas plus anxieuses ou tendues que les autres.
- \_\_\_44. Des logements adaptés pour les personnes handicapées ne sont pas plus chers, ni plus difficiles à construire.
- \_\_\_45. Les enseignants ne devraient pas s'attendre à ce que des étudiants atteints d'épilepsie participent pleinement aux activités d'éducation physique.
- \_\_\_46. Des travailleurs en fauteuils roulants qui ont reçu une formation n'auront pas plus d'accidents de travail que des travailleurs non handicapés ayant reçu la même formation.
- \_\_\_47. Les personnes handicapées n'ont pas plus tendance que les autres à fréquenter l'église.
- \_\_\_48. Puisque une incapacité physique interfère avec certaines activités, cette incapacité se retrouve surtout dans l'esprit de la personne handicapée pratiquement tout le temps.

<b>1 - fermement en désaccord</b>		<b>5 - plutôt d'accord</b>
<b>2 - en désaccord</b>	<b>4 - ne sait pas/sans opinion</b>	<b>6 - d'accord</b>
<b>3 - plutôt en désaccord</b>		<b>7 - fortement d'accord</b>

- \_\_\_49. Les personnes aveugles ont tendance à se faire une première impression plus exacte des gens que les autres.
- \_\_\_50. Une personne avec une incapacité risque davantage qu'une personne non-handicapée d'avoir un enfant atteint lui aussi d'une incapacité.
- \_\_\_51. Pour une personne handicapée grave, la gentillesse des autres est plus importante que n'importe quel programme éducatif.
- \_\_\_52. Les personnes handicapées sont plus sujettes à des accidents que les autres.
- \_\_\_53. La plupart des personnes handicapées préfèrent la compagnie d'autres personnes handicapées à celle des non-handicapés.
- \_\_\_54. Pour la personne handicapée, l'attitude des employeurs représente un plus grand handicap que sa propre incapacité.
- \_\_\_55. À l'école secondaire, un étudiant handicapé se trouvera probablement mal à l'aise dans une classe régulière.
- \_\_\_56. Les personnes handicapées physiques qui conduisent ont davantage d'accidents que les autres conducteurs.
- \_\_\_57. Les personnes handicapées devraient s'intégrer dans notre société compétitive.
- \_\_\_58. La vie serait beaucoup plus facile pour les personnes handicapées si elles vivaient dans des unités résidentielles (par ex. des appartements) avec d'autres personnes handicapées.
- \_\_\_59. Il est logique qu'une femme en fauteuil roulant envisage d'avoir un bébé.

### E. INFORMATIONS SOCIO-DÉMOGRAPHIQUES ET CLINIQUES

Vous avez terminé la partie sur les problèmes modifiés par le degré d'incapacité. Voici quelques dernières questions qui nous permettront de savoir qui vous êtes.

60. Age: \_\_\_ ans

61. Sexe: [ENCERCLEZ]    M    F

62. Statut matrimonial: [COCHEZ UNE SEULE RÉPONSE]

- a) célibataire, jamais marié(e)
- b) marié(e)
- c) séparé(e)
- d) divorcé(e)
- e) veuf(veuve)

63. Combien avez-vous d'enfants dans chaque groupe d'âge? [SI VOUS N'EN AVEZ PAS, INDIQUER 0]

- a) moins de 5 ans
- b) de 5 à 13 ans
- c) de 14 à 18 ans
- d) de 19 à 24 ans
- e) 25 ans et plus

Pour un total de \_\_\_ enfants

64. Quelle est votre spécialité clinique? [COCHEZ UNE SEULE RÉPONSE]

- a) pédiatrie
- b) neurologie
- c) pneumologie
- d) autres (préciser) \_\_\_\_\_

65. Depuis combien d'années êtes-vous affilié(e) à une clinique neuromusculaire?

\_\_\_\_\_ années

66. Consacrez-vous 100% de votre pratique clinique aux maladies neuromusculaires?

oui [NE PAS RÉPONDRE À LA QUESTION # 67]

non [PASSEZ À LA QUESTION # 67]

67. Si vous avez répondu non à la question précédente, veuillez décrire les autres domaines de votre pratique clinique et le pourcentage de temps consacré à chacun:

domaine de pratique\_\_\_\_\_ % de temps\_\_\_\_\_

domaine de pratique\_\_\_\_\_ % de temps\_\_\_\_\_

domaine de pratique\_\_\_\_\_ % de temps\_\_\_\_\_

= 100%

Commentaires additionnels sur certains points du questionnaire

---



---



---



---



---

**Instructions pour nous retourner le questionnaire**

Veuillez vous assurer d'avoir bien répondu à toutes les questions. Veuillez ensuite nous retourner le questionnaire dans l'enveloppe pré-adressée et pré-affranchie.

La carte postale ci-jointe peut nous être retournée séparément si vous souhaitez obtenir un exemplaire des résultats de cette étude.

**\*À NOTER**

Dans le cadre de cette recherche, nous menons aussi des entrevues avec des médecins qui suivent des patients atteints de DD. Ces entrevues ont pour but de nous apporter une compréhension plus approfondie sur certaines questions complexes relevées dans le questionnaire et de nous éclairer davantage sur les opinions des médecins.

Nous organiserons des entrevues aux heures et lieux qui conviennent aux participants. Si vous souhaitez participer à ces entrevues, veuillez cocher la case "oui" sur la carte postale ci-jointe et nous la retourner séparément.

**MERCI BEAUCOUP DE VOTRE PARTICIPATION**

February 26, 1997

Dear Doctor

We are contacting all physicians across Canada who follow patients with Duchenne muscular dystrophy in order to study aspects of current practice. We are specifically interested in end stage management and the option of long term ventilation. As you know, the issue of whether or not to offer ventilation to these individuals is often considered controversial. There is evidence from the United States that physicians vary widely in their practices and beliefs concerning the prescription of long term ventilation to their patients with Duchenne muscular dystrophy.

We are conducting a study to investigate the current practice in Canada and the attitudes and beliefs of the physicians involved. As you are probably aware, information about practices across the country is unknown. We believe that the clinicians who work with patients with neuromuscular diseases will be very interested to know what is happening in other parts of the country. The information collected could also be a starting point to formulate practice guidelines or to encourage dialogue on these sometimes complex issues.

We would appreciate your input on this important topic and have enclosed a questionnaire that we ask you to complete. Your responses are completely confidential. Neither you nor the clinic you are affiliated with will be identifiable in any publication of the results.

Your personal participation in the survey is important. Your input will help to formulate a complete picture of the current practice in Canada. It is estimated the questionnaire will take 15 minutes to complete. We appreciate that your time is valuable, and are therefore offering an honorarium of twenty dollars as a token of our appreciation. A cheque will be forwarded to you once we have received your completed questionnaire.

Enclosed you will find:

- 1) A copy of the questionnaire
- 2) Instructions on completing and mailing the questionnaire
- 3) A stamped envelope for returning the questionnaire
- 4) A stamped postcard for you to return if you would like a copy of the results

We thank you in advance for your time and cooperation. Please return the questionnaire at your earliest possible convenience. If you have any questions you may contact Barbara Gibson (reverse the charges) at (416) 961-5240.

Sincerely,

Barbara Gibson, MSc (Candidate)  
Institute of Medical Science

Karen Yoshida PhD, Graduate Supervisor  
Institute of Medical Science and  
Department of Physical Therapy

**\*This research is endorsed by the Muscular Dystrophy Association of Canada**

le 26 février 1997

Cher ,

Nous contactons tous les médecins canadiens qui suivent des patients atteints de dystrophie musculaire de type Duchennes afin d'étudier leur pratique actuelle. Nous nous intéressons plus particulièrement à la gestion des cas en phase terminale et à l'option de ventilation à long terme. Comme vous le savez, la décision d'offrir l'option de la ventilation à cette catégorie de patients est souvent controversée. Des études provenant des États-unis attestent d'une grande diversité dans les pratiques et les convictions des médecins quant au recours à la ventilation à long terme pour les patients atteints de dystrophie musculaire de type Duchennes.

Nous effectuons cette étude afin d'étudier la pratique actuelle au Canada ainsi que les attitudes et les convictions des médecins impliqués. Comme vous le savez probablement, il n'existe pour l'instant aucune information disponible sur les pratiques appliquées à travers le pays. Nous pensons que les cliniciens qui travaillent avec des patients atteints de maladies neuromusculaires seront très intéressés d'apprendre ce qui se passe dans d'autres régions. Les informations recueillies pourraient également servir de point de départ à la formulation de lignes directrices de pratique ou à l'amorce d'un dialogue sur ces questions parfois complexes.

Nous aimerions bénéficier de votre contribution sur ce sujet important. Vous trouverez ci-joint un questionnaire que nous vous demandons de compléter. Une fois complété, ce questionnaire est entièrement confidentiel. Ni vous, ni la clinique à laquelle vous êtes affilié ne seront identifiables lors de la publication des résultats.

Votre participation personnelle à cette enquête est importante. Votre contribution nous aidera à dresser un tableau complet de la pratique actuelle au Canada. Nous estimons qu'il faut environ 15 minutes pour compléter ce questionnaire. Nous savons combien votre temps est précieux et nous offrons à tous ceux qui participeront à cette enquête une rémunération de vingt dollars pour leur participation. Un chèque vous sera envoyé dès que nous aurons reçu votre questionnaire complété.

Vous trouverez ci-joint:

- 1) un exemplaire du questionnaire
- 2) des instructions sur la manière de le compléter et de nous le retourner
- 3) une enveloppe pré-affranchie pour nous retourner le questionnaire
- 4) une carte postale pré-affranchie à nous retourner si vous souhaitez obtenir un exemplaire des résultats de l'étude.

Nous vous remercions d'avance de votre temps et de votre coopération et vous prions de nous retourner le questionnaire dans les plus bref délais. Si vous avez des questions, vous pouvez contacter Barbara Gibson (à frais virés) au (416)961-5240.

Veillez croire, cher Docteur, à l'assurance de nos sentiments distingués.

Barbara Gibson  
MSc (Candidate)  
Institute of Medical Science

Karen Yoshida PhD  
Graduate Supervisor  
Institute of Medical Science and  
Department of Physical Therapy

**[\*Cette recherche est supportéé par l'Association de Dystrophie Musculaires du Canada]**

### **Appendix 3: Interview Guide**

The purpose of this interview is to discuss your practice and opinions regarding long term ventilation in greater detail than was possible on the questionnaire. There exists a wide variety of opinions on the appropriateness of long term ventilation for patients with DMD. I am interested in hearing about what you do in your clinic and your general opinions about this practice.

1. How many patients with DMD are you currently following?

2. Do you have any patients receiving ventilation?

Probe: What types?, How many hours/day?

3. Could you tell me your opinion about long term ventilation for patients with DMD?

Probe: Why do you feel that way?

What factors do you think are important to consider?

4. Do you always recommend ventilation? probe: what factors do you consider?

5. Did you always feel the same about it?

probe: what or who influenced your opinion

6. Who initiates discussions where you work?

probes: Does everyone feel the same about it?

Whose opinion gets presented to the family?

Who do you think should make the decision?

roles of patient, family, doctor, others?

How do the families hear about it?

From who? When does discussion begin? How often?

Written materials? Formal vs informal? Are other patients involved?

7. Tell me about a recent case.

8. Do you think families need help making the decision? probe: What kind of help?

9. Do families ever seem to make the wrong decision? probe: Is there anything you do in that situation?

10. Is there anything else you would like to share with me that's relevant to this or anything else you think I should know?

## Appendix 4: Consent Form

### Consent Form For Interview Respondents

I have been invited to participate in a research project entitled " Long Term Ventilation for Patients with Duchenne Muscular Dystrophy: An Inquiry into Physicians' Beliefs and Practices." The research is conducted by Barbara Gibson, a graduate student at the Institute of Medical Science, University of Toronto under the supervision of Dr. Karen Yoshida.

As part of this research project, I have been asked to participate in an interview. I understand that during the interview I will be asked questions about my clinical practice and my opinions regarding ventilation for individuals with Duchenne muscular dystrophy.

I agree to participate in one interview that will take approximately one hour. I am aware that the interview will be tape recorded and that written notes will be taken. I understand that the interview will be conducted by Barbara Gibson.

I understand that if necessary, after the interview I may be contacted by telephone and asked to clarify some of the information from the interview.

I understand that the study will not benefit me directly.

I understand that my name will not appear in the published results of this study. Any other identifying information (eg location) will also not appear.

I understand the information that has been given to me about this study and if I have any questions I may contact Barbara Gibson (416-961-5240).

I understand that participation in this study is voluntary and that I am free to refuse to answer any of the interview questions or withdraw from the study at any time.

---

Participant's signature

---

Date

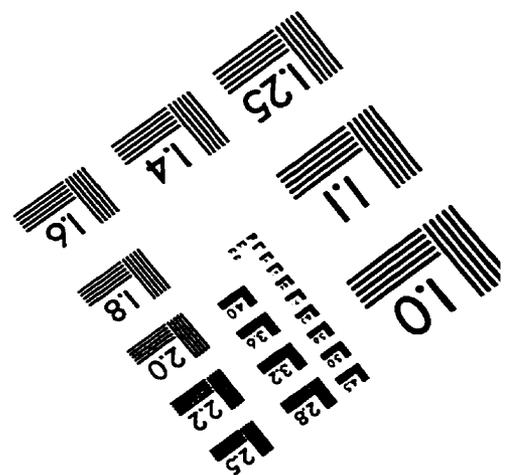
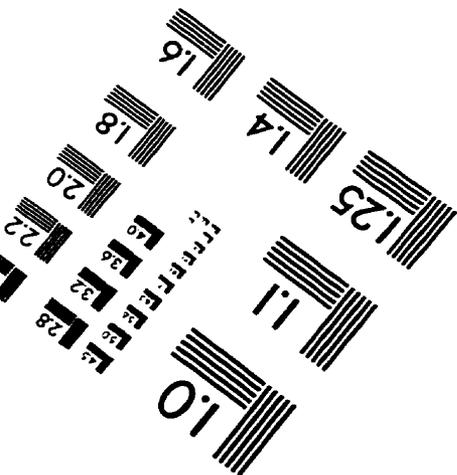
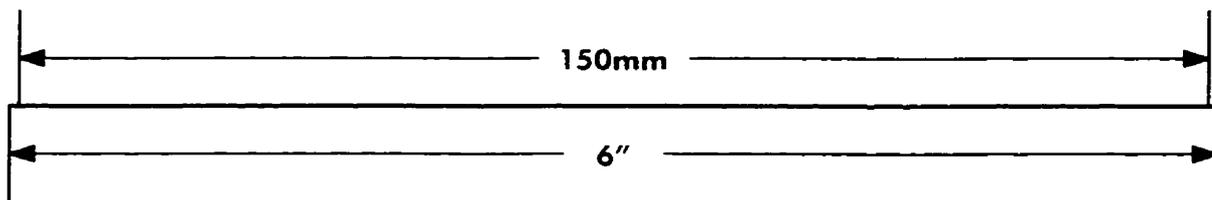
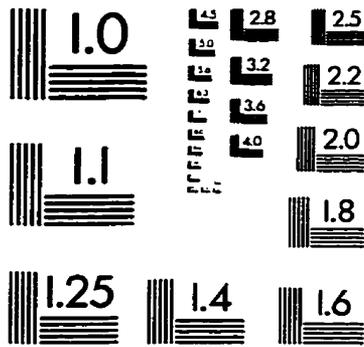
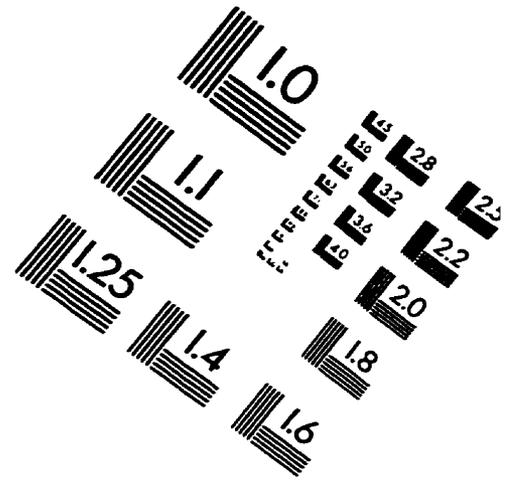
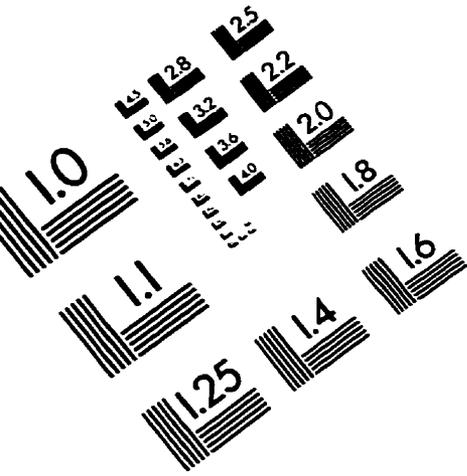
---

Interviewer's signature

---

Date

# IMAGE EVALUATION TEST TARGET (QA-3)



**APPLIED IMAGE, Inc**  
1653 East Main Street  
Rochester, NY 14609 USA  
Phone: 716/482-0300  
Fax: 716/288-5989

© 1993, Applied Image, Inc., All Rights Reserved