VENTILATORS & MUSCULAR DYSTROPHY

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FOREWORD

The preparation of this booklet was initiated four years ago, shortly after Tom Schock, Nancy's son, developed respiratory failure. Both Nancy and Tom questioned me about alternate types of ventilators, the ability to continue to live independently, vocational possibilities, and ultimately what life and death would be like for Tom.

Because I was unable to answer many of these inquiries alone, Nancy and I decided to collaborate in the acquisition of more information. We read all we could find on the use of ventilators for people with Duchenne muscular dystrophy (DMD), conducted a survey of 240 Muscular Dystrophy Association clinic directors, and attended national and international conferences dealing with related issues. We talked with other interested professionals and lay people and finally decided we were ready to share our accumulated knowledge with the people most involved.

Some of our initial questions remain unanswered, but we hope this booklet will provide basic information and guidelines for further investigation on specific topics. We realize there is no model solution for all persons with DMD and are keenly aware that individuals and their families possess vast stores of untapped resources for problem solving.

Agatha P. Colbert, M.D.

ACKNOWLEDGMENTS

We express our gratitude to the people who have provided us with insight, inspiration, and support in this endeavor. In addition to the individuals with Duchenne muscular dystrophy and their families, we thank the contributors to this text who are named in their sections and the authors listed in the bibliography, many of whom are colleagues and close personal friends.

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We also thank Louise MacLeod for her invaluable typing and word processing assistance.

INTRODUCTION

Duchenne muscular dystrophy (DMD) is an inherited disease of muscle that affects mainly male children. This disorder, which begins with weakness in the hip muscles and advances to involve the entire musculature of the legs, trunk, and arms, is usually diagnosed between the ages of 2 and 6 years. As the disease progresses, the muscles used for breathing also deteriorate. In the past, life expectancy had been in the 20-year range, the typical cause of death being respiratory infection or cardiac failure. New perspectives and technologies are changing these expectations.

A consequence of DMD and similar progressive neuromuscular diseases is respiratory failure. Adequate information has not been readily available to either the treating clinicians or involved individuals and their families regarding this fact or how to proceed when life is threatened by this condition.

Some physicians prescribe ventilators to extend the lives of their patients with DMD; others feel it is inappropriate to do so. There is no established method for decision making regarding use or nonuse of these devices, no generally accepted medical protocol for management once the decision is made, and differing opinions about type of ventilators to use.

Lack of consensus on how to proceed creates a dilemma for individuals and family members faced with this eventuality. There are many questions to consider: What additional personal care needs will there be? Who will provide the care? How will the ventilator affect the lifestyle of all involved — physically, psychologically, emotionally? Will the ventilator cause additional suffering and/or disability? If the person chooses to use a ventilator, can that decision be reversed at a later time?

In response to the recognized need for more data, the authors have developed this booklet, which includes information on the basic pathophysiology of respiratory failure, methods of standard respiratory care, types of ventilators used, possible living arrangements, personal and medical care, decision making, and quality of life issues. Our objective is to present facts, rather than offer judgment regarding the issues.

PATHOPHYSIOLOGY

Breathing occurs as a result of the contraction and relaxation of the respiratory muscles, which allow periodic inflation and deflation of the lungs. As the dystrophy progresses, these muscles — the diaphragm (between the chest cavity and the abdomen), the intercostals (between the ribs), and the auxiliary muscles — become too weak to function adequately. The individual is unable to breathe deeply or cough effectively; carbon dioxide builds up in the system and the amount of oxygen taken into the body is less than is needed. When this situation occurs, the individual is experiencing respiratory distress, compromise, insufficiency, or failure, depending on the degree of impairment. Curvature of the spine, which because of the deterioration of the muscles of the back frequently accompanies DMD, can also affect respiration by limiting lung capacity.

Signs of inadequate respiration are irritability, frequent headaches, sleeplessness at night, yawning, drowsiness, and falling asleep during the day. At the same time, the person may have loss of appetite — he cannot or does not want to eat — and as a consequence experiences severe weight loss with possible malnutrition.

Individuals with limited breathing power are susceptible to respiratory infections, which can lead to pneumonia. If the pneumonia cannot be controlled or if the respiratory system fails completely, death will follow.

For additional information regarding the pathophysiology of respiratory compromise in general and details about a particular situation, the person and others involved can turn to the attending physician and other medical professionals. Respiratory compromise is predictable long in advance. Therefore, the opportunity exists for early and continuing discussion on medical issues, personal care needs, adjustment to the disability, choices available, and other concerns.

Communication between the layperson and the physician can be greatly enhanced if they both realize that their knowledge, experience, perspective, and emotional involvement are different; that the disease and its manifestations present a complex situation; and that the terms used for explanation may be unfamiliar. It is essential for the inquirer to organize his thoughts and questions well and to persist until the questions are answered to his satisfaction.

It is important to be aware that during the early stages of respiratory change, the boy or young man is continuing to live according to the pattern he has established. He is attending to daily living needs, socializing, learning, growing, setting and achieving goals, and adjusting to changes in mobility as necessary.

CHOICES/OPTIONS

When the person understands the pathophysiology of respiratory compromise and is aware that this condition is a consequence of the disease, he is prepared in part to consider alternatives for the future. To know how to proceed, he and family members need to explore many issues including the following: courses of action, ventilators and factors affecting choices, living arrangements, and personal and medical care providers and other support persons.

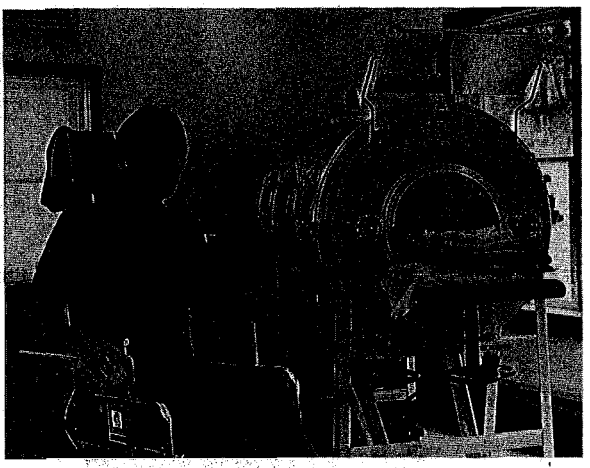
Courses of Action

It is possible to allow life and death to take a natural course without intervention, or to extend life by means of a mechanical device. Opinions vary regarding the wisdom of one choice over the other based on ethical and practical considerations. Ethical issues concern life and quality of life. Can it be in the person's best interest to allow death to occur when it is possible to sustain life? Can a person with DMD who is dependent on a mechanical ventilator maintain a life of quality with meaningful human interaction, a satisfactory degree of personal fulfillment, and acceptable physical comfort? And, practically, can appropriate personal and other essential care be provided? Can costs be absorbed?

Ventilators

Two types of ventilatory support systems are available: negative pressure and positive pressure. Use of a negative pressure device generally requires enclosing the individual's body either totally, as shown in Figures 1A,B or partially, as shown in Figures 2A,B. These devices operate by creating pressure changes between the chest

Figure 1A



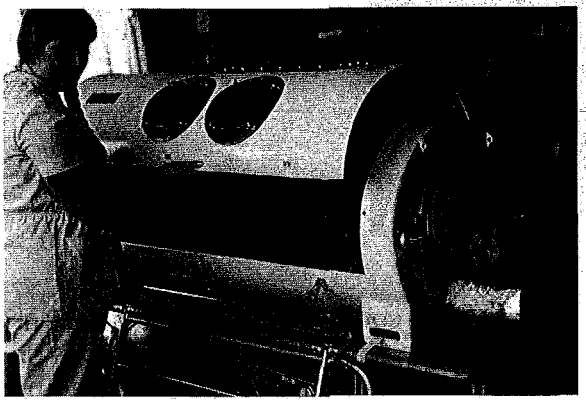


Figure 1B

wall and the encasing shell. As the pressure in this space becomes negative with respect to atmospheric pressure, air is drawn into the lungs through the mouth and nose. The pressures are then equalized and exhalation occurs, allowing carbon dioxide to be blown out.

Positive pressure ventilators act by blowing air directly into the lungs via the mouth, nose, or tracheostomy, as shown in Figures 3A,B,C,D. The pneumobelt, also a positive pressure device, acts by compressing and releasing the abdomen, thus raising and lowering the diaphragm as shown in Figure 4.

Each type of ventilator offers advantages and disadvantages, and the choice of a ventilator will be influenced by the specific medical condition of the individual, his degree of respiratory impairment, his

Figure 2A

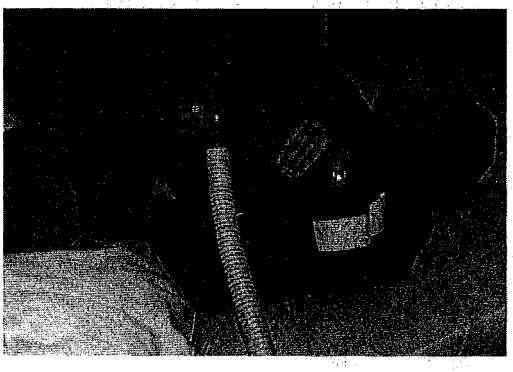


Figure 2B

body configuration, the severity of contractures, the presence or absence of scoliosis, his living situation, his personal preference, the preference of his care providers, and the opinion of his attending physician. (Table 1 provides a list of ventilators used by persons with DMD and associated advantages and disadvantages of each.)

Factors to be considered when selecting the appropriate device include efficiency in providing adequate aeration, reliability in performance, comfort when in the system (including noise level), maintenance requirements, cost, and availability. The following are questions to evaluate: Can the use of this ventilator be incorporated into the individual's current lifestyle? How will a ventilator affect the personal care needs of someone with DMD? Who will provide, repair, and pay for the ventilator, the auxiliary equipment, and training in its use?

With the onset of respiratory compromise, individuals with DMD may require ventilatory assistance on a part-time basis only (often at night). As the disease progresses, more time on the ventilator becomes necessary and the person may then use a combination of

TABLE 1.

NEGATIVE PRESSURE VENTILATION	Advantages	Disadvantages
Iron lung	100% efficient Comfortable Reliable Durable Minimal maintenance Ease of postural drainage	Size Not portable Obstructs view and hearing of user
Porta-lung	Same as iron lung	Cylindrical shape Smaller diameter than iron lung May not accommodate abducted legs
Cuirass	Reliable Portable	60% efficient Musculoskeletal discomfort Custom fit
Plastic Wrap	One size fits all Portable	40% efficient Moves body Discomfort Noisy
POSITIVE PRESSURE VENTILATION	**Advantages**	**Disadvantages**
Pneumobelt	Portable Inconspicuous Ease of daytime use	50% efficient Semi-upright position needed Efficiency dependent on body contours
Nasal Mask or Seal	Ease of use Noninvasive Mouth free	Appearance
Mouth Mask or Lipseal	Good daytime breathing supplement	Pressure on teeth Nighttime use difficult
Tracheostomy	100% efficient Direct easy access for secretion removal	Invasive Potential complications

systems. For example, he may sleep in the Porta-lung at night and use an intermittent positive pressure ventilator (IPPV) for varying periods during the day. Small portable systems are available that can be transported on the back of a motorized wheelchair and thus allow the individual more mobility during the day.

Figure 3A

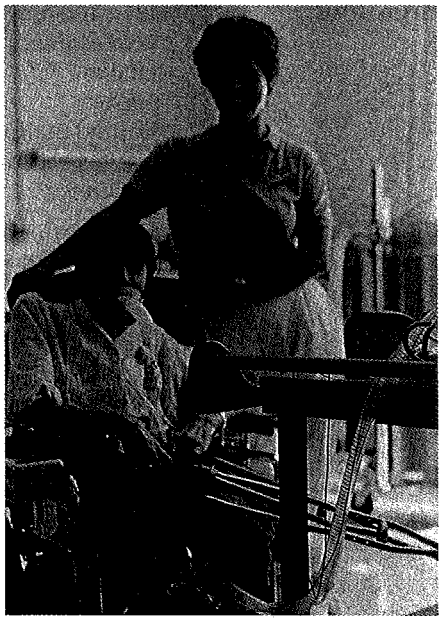


Figure 3B

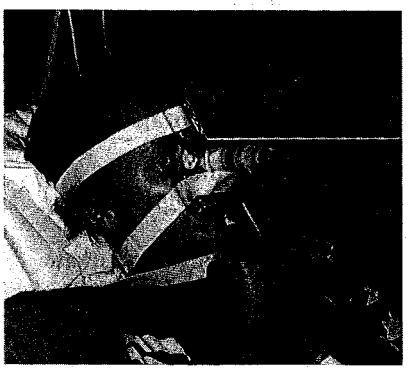
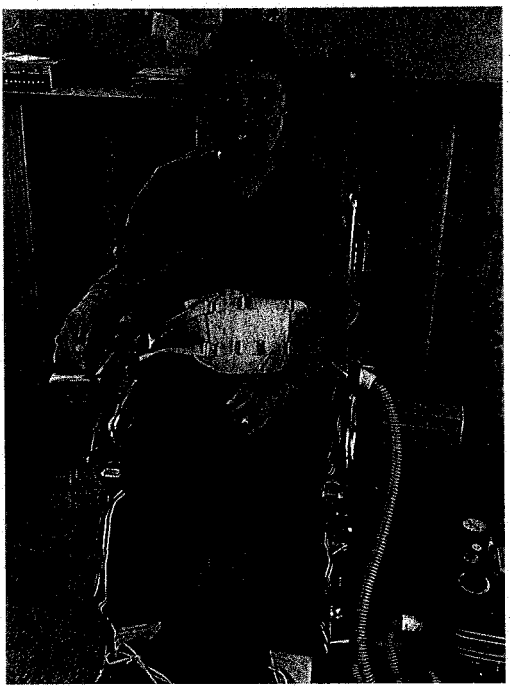


Figure 3C

Figure 3D



The selection of a ventilator is a highly individualized process that requires discussion between the person involved, the physician, and other medical professionals. A minimum of a week to 10-day hospital stay may be necessary to experiment with the various types of ventilators and determine which one will best meet an individual's needs.

Figure 4

Living Arrangements

Living arrangements for young people severely disabled by DMD may be with family members, in an institution, or independently with one or more personal care attendants. Responsibility for managing and providing for personal care and other needs varies according to the setting. No one arrangement is suitable for all. The best solution is one that provides maximally for the person's physical, psychological, and other needs.

Generally, the ongoing care of a person with DMD who lives with family members is managed by those family members. Family members also provide housing and maintain the living space; accept responsibility for feeding, bathing, and dressing; and may assist with educational, recreational, social, and other activities. They may also be involved with coordinating medical care, purchasing and maintaining equipment, seeking information regarding resources, and obtaining services from agencies. If a person living at home is able to manage his care and activities, the family acts as backup. The family may receive assistance, when available, from respite care providers, home health aides, visiting nurses, or personal care attendants.

An institution such as a hospital, nursing home, or hospital school has the responsibility for total care of the patient/resident and his environment. However, the individual does have the opportunity to pursue personal interests and engage in productive or recreational activities depending on his abilities and choice, in accord with institutional regulations.

When living independently, using the services of one or more personal care attendants, the person assumes responsibility for his own life. This includes arranging for housing, furnishings, food, clothing, and all the other necessities an individual living in a separate residence requires. In addition, he is responsible for managing his finances, arranging for medical care, acquiring and maintaining special equipment, and hiring and training personal care attendants who will perform necessary personal care and home management tasks. Young adults with DMD who are living independently may subsist on Supplemental Security Income and/or Social Security Disability Income, live in apartments (often subsidized), and receive money from the state and/or federal government to pay personal care attendants. The choice of a lifestyle depends on the individual and his situation.

MEDICAL AND PERSONAL CARE PROVIDERS AND OTHER SUPPORT PERSONS

Medical and health professionals involved in the care and support of a person with DMD reaching the point of respiratory distress cover a range of specialists whose services may be required on a regular or as-needed basis. The authors decided that it would be appropriate for these people to describe their own roles and appreciate their willingness to do so. Statements appear on the following pages.

PHYSICIANS

Primary Care Physician

The role of the general internist or family physician in the care of persons with neuromuscular disorders who arrive at the stage of respiratory failure is important because it is that doctor who is usually the most readily available. He or she is generally most quickly accessible by telephone and most likely to make a house call. He or she is also most likely to know the family. Although the family physician does not have the specialized training of a neurologist or a pulmonary physiologist, he or she is generally able to handle the problems of respiratory infections, skin ulcers, urinary tract infections, etc. He or she can listen to chests, look into throats, palpate abdomens, obtain cultures of throat and urine, and perform a variety of other diagnostic measures. Nothing about DMD protects one from the garden variety of human illness with which the family physician probably has more experience.

Robert C. Stewart, M.D.

Director of Muscular Dystrophy Association (M.D.A.) Clinic

The director of a Muscular Dystrophy Association clinic may be a neurologist, physiatrist, orthopedist, pediatrician, internist, or a specialist in another field of medical practice. In addition to accepting the role appropriate to his or her specialty, the clinic director coordinates medical and other support services that relate to the dystrophy and the resulting disabilities.

He or she closely monitors the course of the neuromuscular disease and assists in preparing the individual and his family for the onset of respiratory failure. This can involve a series of discussions regarding progression of the disease, life expectancy, and options of prolonging life by means of ventilatory aids.

Agatha P. Colbert, M.D.
Nancy C. Schock, M.A.

Physiatrist

The physiatrist functions as the medical director of the rehabilitation team, prescribing and supervising services such as physical, occupational, respiratory, and speech therapy, psychosocial services, and vocational counseling. When caring for the person with advanced DMD, she or he is cognizant of the individual's physical disability and monitors the need for appropriate adaptive devices, toileting aids, lifts, wheelchairs, communication-enhancement systems, ventilatory aids, and other durable medical equipment.

The goals of the rehabilitation team are to maximize functional independence and facilitate an optimal quality of life for the disabled person and his family. The physiatrist can offer information with regard to quality of life issues. She or he is available to assist in the decision making process related to extending life with ventilatory aids and to support the individual and his family in their choice to allow the disease to take its natural course or to find the most suitable ventilator.

Agatha P. Colbert, M.D.

Neurologist

The decision about whether to provide ventilator support to persons with neuromuscular disease is a matter that requires careful study of each individual case. The primary issue is whether the person will experience an improvement in the quality of his life, as well as a longer life.

The neurologist can describe for the family as accurately as possible the prognosis for the disease, the most likely mechanism of death, and how this transition can be either delayed or made easier for the patient. He or she must also act as a sensitive and caring physician, aware of the philosophical position of the patient, and the patient's special concerns about himself, his family, and his society. Appropriate management can be undertaken only in the background of understanding on the part of the physician. A simple prescription for a home ventilator is just not adequate.

Jack H. Petajan, M.D., Ph.D.

Cardiologist

Individuals with DMD invariably have skeletal muscle weakness. By contrast, involvement of heart muscle leading to symptoms occurs less frequently. The role of the cardiologist includes identification and treatment of heart disease when present.

Because of ambulatory limitation especially in adolescence, diagnosing cardiac disease is often difficult because patients rarely exhibit typical symptoms. As a result, screening in DMD is necessary to detect the early onset of heart abnormalities and should take place yearly. Assessment with the standard chest x-ray and electrocardiogram may not be helpful because many individuals without heart involvement will have abnormal test results. Because of the limited value of these tests, more sensitive methods for diagnosing heart involvement, such as the echocardiogram and nuclear scan, are now employed to quantitate heart function. The reproducibility and noninvasive nature of these tests make accurate serial comparison possible.

After diagnosis, the cardiologist manages therapy, including drugs to improve heart contracture, diuretics to eliminate excess water from the blood, and salt restriction. For the patient requiring hospitaliza-

tion, cardiac status may be monitored by placing a special catheter (tube) into an arm or leg vein and advancing it to the pulmonary artery. The measurements obtained permit separation of lung disease from cardiac involvement.

David R. Fulton, M.D.

Pulmonologist

The pulmonary (lung) specialist becomes involved in the care of patients with DMD when the disease has progressed to produce severe muscle weakness. Invariably, the breathing muscles that expand the lungs become weakened, making it difficult for the patient to move adequate amounts of air in and out of the lungs or to cough sufficiently. The pulmonary specialist then closely monitors the patients to assist in treating lung complications such as pneumonia or inadequate breathing.

Breathing muscle function is monitored by measuring the vital capacity — the total amount of air someone can expel by taking the deepest possible breath and exhaling fully — and blood gases that measure the amounts of oxygen and carbon dioxide in the blood. When breathing becomes inadequate, the vital capacity is usually below 15% of the expected amount, blood carbon dioxide rises, and oxygen levels drop. Patients notice increasing tiredness, weakening of the voice, nightmares, and occasionally bedwetting, but often no shortness of breath.

When this occurs, the pulmonary specialist will often recommend intermittent use of a ventilator, such as an iron lung, to help the breathing muscles in an attempt to decrease the amount of work the person must do to breathe and to leave more energy for other activities. The pulmonary specialist works in concern with other members of the health team and the patient to optimize the patient's breathing.

Nicholas Hill, M.D.

Orthopedist

The use of operative stabilization for scoliosis in patients with muscular dystrophy is gaining wider acceptance. When the choice has been made to extend life beyond the second decade by means of mechanical ventilation, maintaining the spine and joints in the best possible alignment becomes even more important. The decision to undergo surgical intervention such as soft tissue releases and spinal fusion is linked closely with the decision to prolong life. A straight back and flexible joints permit greater comfort and improved function in individuals with advanced muscular dystrophy. Admittedly, there are risks to these surgeries and such decisions can be extremely difficult for the person and his parents. However, the long-term consequences of contracted joints and a severely scoliotic spine need to be assessed in view of the possibility of living an additional 5, 10, or 15 years.

Clifford L. Craig, M.D.

Other Specialists

The physicians listed above are those most frequently involved in the care of persons with DMD. Other specialists such as a gastroenterologist, a dermatologist, and a psychiatrist may be of help for specific problems that arise periodically.

ALLIED HEALTH PROFESSIONALS

Physical Therapist

The rolé of the physical therapist in the care of persons with DMD and respiratory insufficiency lies in assessing the positioning and equipment needs of the individual, and providing appropriate therapeutic exercise to maintain comfort and function. It is the physical therapist's responsibility to adapt the wheelchair so that it offers optimal body alignment and comfort while allowing the person as much independent functioning as possible.

In addition, the therapist will often be involved in helping the individual attain a comfortable position when using an iron lung, Porta-lung, cuirass, or body wrap. Specific breathing exercises, such as the use of blow bottles and inspiratory muscle training, have not proven to be helpful at this stage of the disease. Continued stretching exercises especially of the neck musculature, shoulders, and hands will help to maintain an adequate range of motion and maximize the person's residual strength.

Judy Freyermuth, R.P.T.

Occupational Therapist

The work of the occupational therapist involved in the care of a person with muscular dystrophy includes providing aids to enhance independent mobility, self-care, environmental control, and written communication.

Independent mobility may be achieved with a motorized wheelchair. The occupational therapist often works with the physical therapist or physiatrist to determine whether a hand, mouth, chin, or other type of control is appropriate for the individual. She or he is concerned with designing work spaces that are barrier free and at the necessary height for activities in which the individual may wish to engage.

Computers and related equipment have provided numerous opportunities for persons who are severely disabled. Keyboards and switches may be adapted to interface with the computer, which can then be accessed by finger touch, hand, arm, or head movement, or chin, tongue, lip, or breath control.

The occupational therapist can assist in designing or modifying an environmental control system. Devices include the simple strategically placed buzzer/bell help call, as well as the more complex remote controls that can activate light switches, open and close doors, and operate televisions, radios, tape recorders, or phones.

The therapist may also assist the individual and his family with the instruction of caretakers who attend to his personal needs. Ultimately the therapist's goal is to enhance the person's functional independence and control over his own life.

Ellen O'Gorman, O.T.R.

Speech-Language Pathologist

In much the same way that physical therapists are involved with people who have DMD, the speech-language pathologist may be able to work directly or indirectly in teaching preservation, conservation, and compensation of function.

There may be some individuals for whom vocal communication may not be functionally or physiologically possible at the later stages of the disease. However, these individuals may be able to use some form of augmentative communication system that can be individually programmed to fit the needs and capabilities of the person and his environment. Such systems are also able to produce a hard copy (printout) and synthesized speech (voice output). Additionally, much of this equipment is now compatible with computers. This feature could be a motivating factor emotionally, socially, and educationally.

Lenore Daniels-Miller, ScD., CCC-SpL.

Communication Specialist

The ability to communicate is fundamental to growth, development, and human interaction. If the individual with DMD is unable to use pencil or pen manually to sign or to gesture in a meaningful way, some mechanism for effective exchange of information may be necessary. The role of the communication specialist is the evaluation and remediation of that individual's status so that he can effectively communicate basic survival needs and more advanced information to others within his environment. The communication specialist is knowledgeable about devices and systems that help individuals to communicate under the most challenging conditions.

Susan H. Shealey, O.T.R.

EDS' NOTE:

Persons with DMD who are being ventilated through a tracheostomy experience a varying amount of interference with oral communication. Although technical advances are improving this situation, a communications specialist can provide effective assistance.

Respiratory Therapist

In decision making, the respiratory therapist can offer facts regarding various devices used for ventilatory support. Information is first presented with demonstrations and ample time for answering questions. Whenever possible, a supervised trial of the devices by the person is useful.

Once a course is decided on, the initial setup and adjustment period require frequent change, as well as trial and error. The prescribing physician depends on subjective responses from the person and on physiological testing, such as blood gas analysis, pulmonary function, ear oximetry, and end tidal carbon dioxide measurements. Much of this testing is performed by the respiratory therapist, as is the setting and changing of the controls on the various devices.

After a regimen is established, the respiratory therapist can monitor the effectiveness of therapy, service and maintain the devices, provide the physician with information, and, of course, respond to the patient's concerns.

The responsibility for the final decision regarding ventilator use belongs to the person with DMD and no one else should usurp that role. It is the responsibility of the respiratory therapist to be supportive, understanding, and professional in providing information and helping to carry out decisions.

Kevin E. Strong, R.R.T.

Nurse

A nurse shares responsibility with other members of the health care team in assessing and evaluating an individual's current and future needs and cooperatively designing and implementing a plan for appropriate care.

Having knowledge of general health maintenance, characteristics of muscular dystrophy, and implications of respiratory compromise, the nurse can inform the involved person and others about the expected course of the disease, accompanying disability, and available options for management.

As personal care provider, she or he is concerned with details such as hygiene, nutrition, hydration, infection, pain prevention and control, positioning, and quality of the environment. As nurturer, she

or he is concerned with the person's mental health, activity, and opportunity for social interaction.

The nurse will also facilitate problem solving in areas such as equipment repair, follow-up home care, and obtaining community services.

Donna Euerle, R.N.
Penelope F. Simpson, R.N.

Dietitian

The dietitian becomes involved in the care of people with DMD when the disease has progressed to cause muscle weakness in the throat and/or intestines. Individuals who experience swallowing difficulties lose weight because they are not taking in enough calories. The dietitian evaluates the person's condition and recommends special high-calorie, high-protein drinks that will provide necessary calories, vitamins, and minerals. Milk-based drinks are available for individuals who can tolerate milk products without increased mucus and congestion. For persons who must avoid milk, high-calorie, high-protein clear liquid drinks are available.

Just as weak throat muscles make swallowing difficult, weak intestinal muscles make bowel movements difficult. The intestinal muscles of the person with DMD do not readily contract to move waste products out of the body. As a result, waste material (stool) tends to build up and becomes very hard or impacted. The dietitian evaluates the patient's usual intake and offers suggestions regarding the fiber and fluid content of the diet. Additional fiber and fluids will help keep the stools soft for easy elimination.

After a nutrition support program has been established, the dietitian can monitor progress based on weight gain, fat thickness measurements, bowel patterns, and routine blood work.

Gretchen Simendinger, R.D.

Psychologist

Confronting respiratory insufficiency can tax and even overwhelm an individual's emotional equilibrium. The mental health professional can help the person experiencing emotional distress identify his feelings. This initial stage in therapy is often very helpful in its own right because frequently one can simply feel overwhelmed but not understand how or why. For instance, one person may need to work through feelings stimulated by the additional loss of functioning. Another may experience fear and anxiety with this adjustment. The major focus of any therapeutic intervention is to assist the individual in understanding, expressing, and bearing his feelings. It is only then that the distress can be put into perspective.

David Sloan-Rossiter, Ph.D.

Social Worker

For individuals with late-stage muscular dystrophy, the decision to use ventilatory support presents a complex challenge. Social work services can assist the person and family in a variety of ways.

The social worker, in conjunction with other members of the health care team, can provide education during the decision making process that will facilitate understanding of the choices involved as well as related outcomes. Education can address technical and financial aspects of positive and negative pressure devices, community resources, and subsequent stress that the person and family may experience in daily living.

Through counseling sessions the social worker can help the individual and family address both the short-term and long-term impact. Counseling provides a vehicle by which options can be discussed objectively, aiding the person and family in making decisions that are right for them. In addition, the social worker can provide assistance in assessing quality of life and how the ventilator influences that quality of life. In ongoing counseling sessions the social worker can address special issues and challenges that arise, provide support, and facilitate problem solving.

Jessica Robins Miller, M.S.W.

Recreation Therapist

Because of choice and/or circumstance, some persons with disabilities may not have vocational or educational outlets, but everyone has the opportunity for recreation. Persons with DMD often experience social isolation because of the difficulty in getting out into the community. A recreation therapist may be helpful in identifying leisure activities that hold a special interest for the individual, facilitating socialization opportunities with people whose company he or she enjoys, and providing experiences that are relaxing and refreshing. This therapist may also be instrumental in organizing and facilitating common interest groups to attend lectures, museums, sports events, and other recreative activities.

James B. Ross III, Recreation Therapist

COMMUNITY RESOURCES

Family

Grandparents, aunts, uncles, and cousins can relate to the family member with DMD in many different ways. As interested listeners, they can encourage him and others close to him to retell what they have recently learned from the physician or other specialist about changes caused by progression of the disease and procedures and devices useful for the person's continued functioning. This process, which allows for learning, adapting, and reacting to the new information, can be helpful to all.

The extended family members who become involved can assist with personal care or problem solving, or they can help the person who is disabled to develop his intellect, to become adept socially, and to find creative ways of using his talents.

It is essential for any close relationship that the participants be knowledgeable about the nature of the disease and its effects. Keeping family members informed is the responsibility of the person with dystrophy, parents, or other primary care providers.

Francis McCarthy, Grandmother
Mary Ann McCarthy, Mother

EDS' NOTE:

Inherent in day-to-day living for the family of a person with muscular dystrophy are dealings with a progressive disability, attempts to meet increasing care demands, and searches for answers to complex questions about the future. Each immediate and extended family member approaches and adapts to the person's changing needs according to his or her own unique style. Individual coping strategies must be respected, although at times they may not be understood by all persons involved.

As the child with DMD becomes an adult his physical, emotional, social, educational, and psychological requirements are altered. Parents who formerly found care providing moderately inconvenient may now experience the situation as overpowering. The family member who has assumed a primary role in caring for the disabled individual is particularly prone to overextending herself or himself if left with the complete responsibility of that individual's care. There are few families who are able to meet these additional demands without its impacting significantly on their daily lives. The majority of families welcome some degree of support.

It is important that each family recognize when outside help is needed, who within the extended family is willing and able to offer assistance, and which community agencies might accommodate these needs.

Peer Counselor I

Respiratory insufficiency is often the first sign that disability of the individual with muscular dystrophy has progressed to a life-threatening stage. His response to his future closing in, his increased debilitation, and the stress experienced by his family is likely to be a confusing welter of painful emotions — fear, rage, guilt, and despair.

The task of mental health providers who are responsible for his care is to offer support and comfort and to help him explore the most fundamental questions. Does life have meaning that can reconcile one to suffering and death? What gives life value? Can life be worth living even within the limits imposed by a disability?

In sharing these circumstances of living with a disability, the peer counselor may have a number of advantages in accomplishing these

tasks. First, a person with DMD may be inclined to trust more readily in the peer counselor's ability to empathize and to remain non-judgmental. Second, he may be less likely to engage in self-defeating "games" designed to vindicate self-pity. Third, he may regard the peer counselor as a positive role model for living with a disability. In fact, this last may be the peer counselor's greatest contribution — his or her ability to extend the challenge and hope that the person can make life worthwhile.

Paul Kahn, M.Ed.

Peer Counselor II

The role of the peer counselor is to help a person who, like himself, has DMD and requires respiratory support to maintain life. Through discussion of common experiences and sharing of ideas, the peer counselor and person he is assisting can decide on objectives to work on to achieve the ultimate goal of creating a reasonably comfortable, stable, and fulfilling life.

Basic needs objectives are to obtain and coordinate medical care, to manage personal care providers, to cope with respiratory insufficiency and lack of mobility, to deal with societal attitudes, and to provide an accessible residence with congenial cohabitants, near family and friends.

Personal development objectives are to participate effectively in life's activities at home or in an accessible educational, recreational, or employment setting, and to have the opportunity for pursuing interesting endeavors, and for developing meaningful relationships.

Tom Schock, B.A., M.S.

Support Groups

Support groups are composed of persons with a common experience who are attempting to deal with similar issues in their lives. The groups may be formal, led by a professional who guides participants in exploring their situations, their feelings, and their responses, or informal with participants discussing (without a professional counselor) issues, tasks, attitudes, feelings, resources, coping skills, and other subjects that are important to them.

Coming together on a regular basis to share and interact can help group members develop mutually supportive relationships with emotional and practical benefits. Exchanging information and support can also facilitate adjustment to the disability, help group members recognize personal strengths, diminish a sense of isolation, and serve to clarify options regarding the use of respiratory assistance.

E. Virginia Harting
Sandra Reilly, L.S.W.

Ventilator Equipment Vendor

The role of the equipment vendor is to clearly define the vendor's responsibilities regarding equipment, service, billing, and continuing education to hospital personnel, client, and family members. The policies and procedures involving third-party reimbursement are explained fully to the client and family.

The vendor evaluates the home environment for suitability and safety before discharge and makes recommendations for possible changes in specific areas (e.g., public service, telephone, access/exit, insurance, etc.). He or she also arranges for delivery of equipment to the hospital before the client's discharge. Thorough instruction is provided to the client, family members, and hospital personnel in proper use, maintenance, safety, cleaning, and troubleshooting of home care equipment. The vendor ensures that all equipment is in the home and functioning correctly before the client's arrival.

All equipment is labeled with appropriate emergency telephone numbers and the vendor is available to respond to emergencies in equipment malfunctions on a 24-hour basis.

Verbal and written reports are made readily available to physicians and hospital coordinators regarding equipment performance. The vendor evaluates the equipment for adequacy in meeting the client's needs and makes recommendations for changes to the physician.

The equipment vendor provides credentialed respiratory therapists for periodic checks and follow-ups and assists with travel requirements of the client.

Joseph P. Cook, B.A., R.P.T.

Independent Living Center

In recent years it has become clear to a growing number of persons with severe physical disabilities that they are able to manage their own lives and live quite securely outside the safety of abnormal living situations. Included in this number are persons with DMD who, because of respiratory insufficiency, require support in one or more forms to maintain optimal health and functioning.

The support and adaptation required may vary considerably from person to person. For example, one person may benefit from nightly use of a ventilator, whereas another may require frequent aspiration, and yet another may simply need to observe good preventive health practices.

In principle, the supportive role that independent living center staff play remains constant across the spectrum of individual needs. Regardless of the particular physical needs of an individual that must be taken into consideration, the goal of the independent living center staff is to assist each individual to acquire the proper blend of knowledge, skill, and community-based resources that he may go ahead with the business of life and all that it involves.

Eric L. Griffin

Respite Care

Meeting the needs of a person with muscular dystrophy may be tiring for the people caring for him, especially if they are members of his family.

Among other tasks, ventilatory needs include removal of respiratory tract secretions requiring assisted cough and/or chest physical therapy once or several times daily. When secretions build up, the involved person is frequently frightened about not being able to breathe. People caring for him also worry when he has these problems. All concerned may feel the need for respite.

Respite is care given to a person in either a facility or a home by a specially trained care provider. For individual families, respite services may be available for a 1- to 2-week period or for a certain number of weekends during the course of a year.

The availability of institutional and/or home-based respite care varies geographically. It is most often provided through the Department of Social Services or the Department of Mental Health. Specific

information on the regulations for each state may be found by contacting the National Developmental Disabilities Councils, 1234 Massachusetts Avenue, Suite 203, Washington, DC 20005 (202/347-1234).

Gertrude H. Murphy, M.D.

Special Educator

The role of the special educator working with a student with special needs is to understand the nature of the condition affecting the student and the impact this condition has on his ability to learn. Understanding special needs is a key part of the development of an individualized education plan designed to minimize the effects of these constraints and maximize the student's learning strengths. The special educator is responsible for carrying out this individual education plan while being sensitive to the student's condition on any given day. The special educator is responsible for all educational aspects of the student's program in consultation with other members of the student's evaluation team, which would include a medical representative.

Mary Gaynor, Special Educator

Attorney

Attorneys play an important role when a person with muscular dystrophy is anticipating respiratory insufficiency and is considering use or nonuse of a ventilator. The person may wish to sign a durable power of attorney that will allow someone else to make known his decisions about ventilator use if he is unable to do so himself. If the person is in a custodial facility, such as a hospital or nursing home, he may find that the institution does not go along with his decisions. This may require argument before a Probate and Family Court. Representation by the person's lawyer is necessary so that his wishes can be carried out in a proper and legal fashion. For these reasons, a person with DMD who is anticipating respiratory failure, along with his family members, may want to consult with an attorney before an emergency arises.

Kenneth I. Kolpan, Attorney

EDS' NOTE:

As society has become increasingly aware that many government programs and services have discriminated against people with disabilities, laws have been enacted to correct this situation. Recent laws ensure that all citizens have equal opportunity to acquire an appropriate public education, equal opportunity for employment, and access to public transportation, recreation, housing, and more. Section 504 of the 1973 Rehabilitation Act states in part, "No otherwise qualified handicapped individual in the United States shall solely by reason of his handicap, be excluded from the participation in, be denied the benefits of, or be subject to discrimination under any program or activity receiving federal financial assistance."

When these opportunities, entitlements, benefits, etc., are not accessible or available to an eligible person, he can turn to an attorney for assistance in gaining what is his right by law.

CONCLUSION

As anyone living with this situation is aware, increasing muscle weakness with accompanying disability is a fact of life for a person with DMD. From an early age the child's condition requires that he and people close to him become knowledgeable regarding all aspects of the disease, including respiratory failure and methods for management.

Although ventilators have been available for people with respiratory problems, physicians and lay people have not had easy access to relevant information regarding their use in muscular dystrophy.

The authors have described the process of respiratory failure and the possible courses of action when this occurs and have identified resources that are available with the hope that this information will be useful.

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GLOSSARY

Acute Pulmonary Decompensation — sudden inability of the lungs and/or pulmonary muscles to maintain adequate respiration.

Airway Secretions — fluids produced in the mouth, nose, pharynx, trachea, or lungs to provide natural lubrication.

Alimentation — the act of giving or receiving nourishment.

Anoxia — absence or lack of oxygen.

Apnea — cessation of breathing.

Arterial Blood Gases (ABG) — a measure of the oxygen and carbon dioxide levels in the blood stream.

Aspiration Pneumonia — inflammation of the lungs as a result of inhaling food particles or other foreign matter.

Assisted Cough — application of pressure to the abdomen to strengthen the cough.

Atelectasis — collapse of the whole or part of the lung.

Bronchial Hygiene — the proper health care of the airways leading to the lungs.

Carbon Dioxide Narcosis — a condition characterized by stupor or unconsciousness as a result of excessively high levels of carbon dioxide in the blood.

Carbon Dioxide Retention — a condition resulting from an individual's inability to ventilate adequately.

Cardiopulmonary Function — the special action of the heart and lungs that maintains adequate circulation and respiration.

Congestion — excessive or abnormal accumulation of secretions in the lungs or airways.

Cor Pulmonale — heart disease caused by increased pressure in the lungs.

Cuirass — a negative pressure ventilator consisting of a plastic shell that covers the chest, attached to a motor. (See Figure 2A.)

Cyanosis — a bluish discoloration of the skin caused by insufficient oxygen in the blood.

Diaphragm — the most important muscle for breathing, located between the chest cavity and the abdominal cavity.

Dysarthria — imperfect articulation of speech as a result of poor muscle control.

Dysphagia — difficulty in swallowing.

Dyspnea — difficult or labored breathing.

Ear Oximeter — a photoelectric device attached to the ear that measures oxygen levels in the blood.

Echocardiogram — a motion recording of the position of the internal structures of the heart.

Electrocardiogram (EKG) — a graphic tracing of the electrical activity of the heart.

Endotracheal — within the trachea (windpipe).

Endotracheal Tube — a plastic tube inserted into the windpipe via the nose or mouth.

Etiology — the study of the cause of a disease.

Extubation — removal of a previously inserted tube such as an endotracheal tube.

Frog Breathing — See glossopharyngeal breathing (GPB).

Gastrostomy Tube — a tube surgically inserted through the wall of the stomach to allow for feeding.

Glossopharyngeal Breathing (GPB) — a method of forcing air into the lungs using the tongue (glosso) and the throat (pharynx).

Hypercapnia — an excess of carbon dioxide in the blood.

Hypotonia — a condition of diminished muscular tone associated with weakness and floppiness.

Hypoxemia — a condition of diminished oxygen in the blood.

Intercostal Muscles — muscles situated between the ribs that assist the diaphragm in the act of breathing.

Intermittent Positive Pressure Ventilation (IPPV) — assisted breathing from a ventilator via a face mask, mouthpiece, or nose tube.

Invasive — diagnostic or therapeutic procedures on the body obtained by surgical means.

Iron Lung — a cylindrical negative pressure ventilator enclosing the body, often called a "tank." (See Figure 1A and 1B.)

Operative Stabilization — a surgical procedure by which the spine is fixed in an aligned position.

Pathophysiology — the study of how normal physiological processes are altered by disease — in this case, the effect of Duchenne muscular dystrophy on the respiratory system.

Plasma Bicarbonate — a blood product that indicates the relative acidity of the blood.

Plastic Body Wrap — a negative pressure ventilator consisting of a non-porous garment attached to a motor with a shelled arch over the chest. (See Figure 2B.)

Pneumobelt — an inflatable corset-like device placed around the abdomen intermittently pushing the diaphragm up to assist in breathing. (See Figure 4.)

Porta-Lung — a smaller fiberglass version of the iron lung or "tank" ventilator. (See Figure 1B.)

Postural Drainage — therapeutic drainage of lungs through positioning and rhythmic beating with the hands over the affected lung area.

Recumbent — lying down.

Respiratory Acidosis — excessive retention of carbon dioxide, also called hypercapnia.

Scoliosis — a sideward curvature of the spine.

Suctioning — aspiration or removal of fluids by mechanical means.

Tachycardia — a rapid heart rate.

Tank Ventilator — an iron lung or Porta-lung.

Tracheostomy — a surgical opening in the trachea or windpipe through which a tube is inserted to facilitate breathing.

Ventilator — a breathing machine using either positive or negative pressure that increases air movement to and from the lungs.

Vital Capacity — the volume of gas that can be expelled from the lungs after breathing in as deeply as possible.

RESOURCES AND PUBLICATIONS

Accent on Living, Inc.
P.O. Box 700
Bloomington, IL 61702
309/378-2961

Accent on Living is a nonprofit organization serving people who are disabled by collecting and disseminating specialized information. Services are available through *Accent on Living,* a quarterly magazine; Accent on Information, a computerized retrieval system; and Accent on Special Publications.

Association for the Care of Children's Health (ACCH)
3615 Wisconsin Avenue, N.W.
Washington, DC 20016
202/444-1801

ACCH is dedicated to meeting the needs of children with chronic illness and handicapping conditions, supporting parent networks, and acting as referral for information and advocacy regarding quality care.

Federation for Children with Special Needs
312 Stuart Street
Boston, MA 02116
617/482-2915

The Federation, a parent-run information and training center, develops programs and projects that address the multiple concerns of families and professionals who work with children with special needs.

Gazette International Networking Institute (GINI)
4502 Maryland Avenue
St. Louis, MO 63108
314/361-0475

An organization that encourages communication and sharing among people who are disabled regarding such issues as independent living, housing, integration into the community, and ventilator use. GINI publishes the *Rehabilitation Gazette,* an international journal of independent living by individuals with disabilities.

International Ventilator Users Network (IVUN)
4502 Maryland Avenue
St. Louis, MO 63108
314/361-0475

IVUN links ventilator users with each other and with health care professionals interested in home mechanical ventilation.

Muscular Dystrophy Association (MDA)
810 Seventh Avenue
New York, NY 10019
212/586-0808
(offices in many cities)

MDA provides many services for people with muscular dystrophy and related diseases. For services not included in their program, the Association acts as liaison between the person needing help and the appropriate agency.

OSERS News in Print
Office of Special Education and Rehabilitative Services
Room 3018, Switzer Building
330 C Street, S.W.
Washington, DC 20202
202/732-1250

Free. Includes information on federal programs, current issues, and publications.

Sick Kids (Need) Involved People (SKIP)
216 Newport Avenue
Severna Park, MD 21146
301/647-0164

SKIP addresses issues of home care for children assisted by medical technologies, such as children using ventilators.

The Association for Severely Handicapped (TASH)
7010 Roosevelt Way, N.E.
Seattle, WA 98115
206/523-8446

TASH stresses the importance of integration in living, working, and learning and maintains a register of professional contact people available for assistance with specific problems.

The Exceptional Parent
605 Commonwealth Avenue
Boston, MA 02115
617/536-8961

Bimonthly journal. Practical guidance for everyone concerned with disability, professionals as well as parents.

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