

**Muscular Dystrophy Canada**

# Case for Support for Respiratory Care Project

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## Contents

Executive Summary .....	4
Background .....	5
Methodology .....	5
Why Respiratory Care is important for people living with neuromuscular diseases .....	6
Addressing the Issues – Work to date .....	8
Prevention, early intervention and support services.....	10
Summary of Issues .....	14
Recommendations for Muscular Dystrophy Canada .....	16
Potential Partners .....	16
Impact on Muscular Dystrophy Canada.....	16
Impact on people affected by neuromuscular diseases .....	17
References .....	18
Appendix 1	
List of Key Informants & Reviewers .....	22
Appendix 2	
Recommendations for Muscular Dystrophy Canada .....	23
Appendix 3	
Environmental Scan.....	seperate document

## *About the authors.....*

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## A Case Story.....

Late in 2011, the Atlantic branch of the Muscular Dystrophy Canada received notice that a child was being airlifted to the IWK Health Centre in Halifax with respiratory distress (pneumonia and a collapsed lung). Muscular Dystrophy Canada staff contacted the child's father who stated that he was not totally confident with the initial professional care given and their knowledge of the possible complications with neuromuscular disorders and oxygen. The child was showing signs of delirium and confusion which concerned his family. Muscular Dystrophy Canada staff provided information to the health professionals and consultation referral for advanced expertise resulting in reassurance that the child was receiving competent care. Muscular Dystrophy Canada continued to stay in contact with the family and was able to facilitate the provision of travel costs reimbursement. The family was very grateful and stated how they learned how important monitoring oxygen is and maintaining proper airway clearance. They also expressed their surprise that most health professionals that they were in contact with in the hospital were not aware of the effects oxygen can have on a person with neuromuscular disorders. The child was discharged with a prescription for a cough assist machine as well as a bi-pap machine and receiving ongoing support through a local respiratory care equipment supplier. The child is now doing very well.

This case story illustrates the importance of education and awareness among families and health professionals on early intervention and the issues related to caring for individuals with respiratory challenges. Happily, the story had a positive conclusion. It is the hope of Muscular Dystrophy Canada that these situations can be avoided altogether. Education, advocacy and support, underlined by strong evidence, are critical.

## Executive Summary

Research shows that proper respiratory care can extend the life of people with neuromuscular disorders and has a significant impact on their quality of life. Respiratory failure is the number one cause of death among Duchenne Muscular Dystrophy patients, yet today there are preventative measures and interventions that can be beneficial in maintaining and prolonging comfort of life. The costs associated with invasive ventilation are high as many people are unable to live in the community due to mechanical ventilation and are therefore forced to reside in ICU, often unnecessarily. Options of non-invasive ventilation are not well known and understood by both patients and health care providers. Prevention of secondary complex medical conditions is critical in order to prevent long term hospital admissions, loss of independence and or life threatening conditions (McKim et al).

This paper outlines the above issues and concludes with providing recommendations for the important leadership role Muscular Dystrophy Canada can play in addressing priority actions to improve the lives of people with neuromuscular diseases.

## Background

In 2011, the Board of Directors for Muscular Dystrophy Canada asked staff to propose a number of priority focus areas for the organization at its September 2011 planning meeting. One of the issues presented and endorsed by the Board was the promotion of best practices in respiratory care among individuals living with neuromuscular disorders. The Board supported the importance of this initiative and agreed to assess the issues and determine the role for Muscular Dystrophy Canada to play in the coming years. To that end, the Board at its March 2012 meeting requested the development of a case for support which;

- provides background information on the scope of the problem and identifies gaps and opportunities in education, service and advocacy, and
- makes recommendations on Muscular Dystrophy Canada's role in addressing this issue

This report responds to that request.

## Methodology

This report synthesizes information gathered through an environmental scan of key reports, articles and organizations that have addressed this issue (see Appendix 1). In addition, a number of key informants from clinical, community and government settings, as well as people living with neuromuscular diseases have been interviewed to share their opinions on gaps and priorities in respiratory care.

This case for support is not meant to be an exhaustive detailed description of all the issues but a snapshot of challenges and opportunities intended for internal use for Muscular Dystrophy Canada's Board of Directors to determine the best role for the organization.

## Why Respiratory Care is important for people living with neuromuscular diseases

It is estimated that tens of thousands of Canadian citizens' lives are affected by neuromuscular disorders (Muscular Dystrophy Canada, 2011, p. 6). These disorders have varying symptoms and rates of progression and can impact the lives of children and adults of all ages. Muscular Dystrophy Canada currently recognizes over 150 neuromuscular disorders (Muscular Dystrophy Canada, 2011). According to Muscular Dystrophy Canada (2007):

Muscular Dystrophy (MD) is the name of a group of muscle disorders that are characterized by progressive weakness and wasting of the voluntary muscles that control body movement. As muscle tissue weakens and wastes away, fatty and connective tissue replace it. The muscular dystrophies are among the most recognizable forms of neuromuscular disorders. (p. 13)

Duchenne Muscular Dystrophy (DMD), one of the more common forms of muscular dystrophy affects approximately 1 in 3000 males (Muscular Dystrophy Canada, 2007; Finder et al., 2004). This form of muscular dystrophy and other common neuromuscular disorders will be discussed throughout this report when referencing various best practices and articles.

Respiratory issues associated with neuromuscular disorders can have a significant impact on the quality of life of those living with the disorder. While breathing interventions for muscular dystrophy will be discussed in this report, it should be noted that respiratory complications can affect individuals with numerous other conditions, including amyotrophic lateral sclerosis (ALS), spinal cord injury, polio, and chronic obstructive pulmonary disease, among others (The Chronic Vent Strategy Task Group, 2006; BC Association of Individualized Technology and Supports for People with Disabilities, Provincial Respiratory Outreach Program, 2006).

Respiratory difficulties are a major cause of morbidity and mortality among those with neuromuscular disorders as weakened muscles make breathing and coughing increasingly difficult (Muscular Dystrophy Association [MDA], 2009; The Rehabilitation Centre, Ottawa, n.d.). Various interventions and equipment used to improve respiratory function have been developed for individuals living with these disorders to prevent complications and improve quality of life (MDA, 2009). **However, lack of research and education, awareness of best practices on this topic, conflicting opinions and variations in access to respiratory care across Canada have created barriers to quality care for those affected by neuromuscular disorders** (Katz et al). In addition, individuals whose respiratory symptoms deteriorate as their disease progresses may require mechanical ventilation. For example, Guillain-Barre´ syndrome and myasthenia gravis are two forms of neuromuscular disorders associated with respiratory complications, with 25-50% and 15-27% needing intubation or mechanical ventilation respectively, during their lifetime (Mehta, 2006). Generally speaking, all people with DMD will be on a ventilator at one point in their care progression, although with proper education and preventive non-invasive management, invasive ventilation should only be required for a very small number. For invasive ventilation, a tracheostomy tube is used to assist the patient with breathing, whereas non-invasive ventilation uses a mask instead of a tracheostomy (The Chronic Vent Strategy Task Group, 2006).

**When not properly prevented, respiratory failure and associated ventilation is associated with major costs to the health care system and to patients and their families.** Numerous reports and task forces have examined the issue of chronically ventilated patients in the intensive care unit (ICU) and its impact on the health care system. These issues and the importance of early intervention to prevent respiratory complications for those with muscular dystrophy will be addressed throughout this report.

## Addressing the Issues – Work to date

An environmental scan was conducted to understand what work has been published to date on respiratory care for those living with neuromuscular disorders. Through searches, various reports, articles, and organizational websites were found and subsequently analyzed to better understand what information and best practices are currently available and what gaps and opportunities currently exist in the literature. The below discussion of research and best practices is not meant to be fully comprehensive, but rather present examples of who has examined this topic area. Based on the scan, a number of best practices for respiratory care were identified.

The Canadian Thoracic Society recently published guidelines regarding home mechanical ventilation for adults with a number of different disorders, including neuromuscular disorders (McKim et al., 2011). In addition, the American Thoracic Society (ATS) and a more recent panel of respiratory experts have released consensus statements concerning respiratory care and management for patients with DMD (Finder et al., 2004; Birnkrant et al., 2010). The ATS also published a joint statement with the European Respiratory Society (ERS) outlining tests used to assess respiratory muscle function (American Thoracic Society/European Respiratory Society, 2002). In 2008, a position paper was produced by the Ontario Spinal Cord Injury (SCI) Solutions Alliance entitled, “Living Fully in Ontario Communities: People with spinal cord injuries and disease who use respiratory supports”. This paper provides recommendations in an effort to optimize the health of those with spinal cord injuries and other diseases, with a focus on supporting individuals as they live in a community setting (Ontario Spinal Cord Injury Solutions Alliance, 2008). Lastly, a Cochrane review entitled, “Nocturnal mechanical ventilation for chronic hypoventilation in patients with neuromuscular and chest wall disorders” was found through the environmental scan (Annane, Orlikowski, Chevret, Chevrolet, & Raphael, 2007).

**The lack of high quality randomized controlled trials and specialized research regarding respiratory care for neuromuscular disorders was also discussed** by key informants during the interviews, in the statement published by McKim et al. (2011) and in the Cochrane review described above (Annane et al, 2007). Therefore, while there are a number of different consensus statements and guidelines published, as described above, **more targeted research is needed in the future**. In addition to the need for more research, **there is currently a lack of awareness among health care practitioners regarding current best practices and guidelines in respiratory care. Therefore, developing strategies to disseminate evidence to those who will benefit from it will be important as more research is produced.**

Through the environmental scan, one theme that emerged **is the issue of transitioning chronically-ventilated patients out of the Intensive Care Unit (ICU) into a community-based setting**. Many ventilated-assisted individuals (VAIs) do not have the level of personal or third-party insurance to cover the initial costs of the ventilator and associated respiratory equipment. In addition, if the equipment malfunctions or fails, they may not have the resources to repair or obtain replacement in a timely fashion, thus increasing the likelihood of a return to hospital. **Therefore, it is strongly suggested that a publicly funded system to support VAIs in the community be available and include timely access to equipment, maintenance services and a structured, ongoing educational program**. Such a program is also expected to entirely prevent the need for hospitalization and when necessary, facilitate the transition to home, thus reducing hospital days and improving access to critical care beds. It has been shown that some patients who are chronically-ventilated in ICU settings are stable, but there is a lack of alternate settings in the community and supports for these individuals (Chronic Vent Strategy Task Group, 2006). In Ontario, task forces and committees have been established to develop strategies and recommendations to help transition and support patients out of the ICU. In a report published by the

Chronic Vent Strategy Task Group (2006) it was estimated that in Ontario, between 1,000 and 2,000 additional ICU admissions could be accepted into current facilities if medically-stable chronically-ventilated patients were discharged in a more timely fashion to a more appropriate setting. This number will increase significantly over time if there is no change to how these patients are managed. (p. ii)

The College of Respiratory Therapists of Ontario set out to address the issues of transitioning patients out of the ICU discussed above. They conducted an 18 month project and were able to successfully transition 30 patients from the ICU into a community-based setting. This was achieved through the use of Respiratory Therapist services for patients requiring mechanical ventilation for a number of different diseases. **This project was also estimated to result in a significant cost savings to the health care system** (College of Respiratory Therapists of Ontario, 2011). **These findings suggest the need for a community based model of care, similar to B.C.'s Provincial Respiratory Outreach Program** <http://www.bcits.org/proplink.htm> . However, equally important, is an understanding of effective best practices that prevent patients from being in ICU in the first place or effectively weaning them from non-invasive ventilation to NIV and ensuring their ability to return to the community.

**In summary, while there are best practices and guidelines available for respiratory care, there is a lack of awareness of these practices amongst health providers. In addition, further research and analysis is required to enhance community supports for those living with neuromuscular disorders who require respiratory care.**

## **Prevention, early intervention and support services**

While those with neuromuscular disorders cannot prevent respiratory problems indefinitely, there are some ways that those living with muscular dystrophy can try and limit the number of respiratory

complications they face. For example, having childhood vaccinations, getting the flu shot, avoiding colds and treating infections, ensuring effective cough, monitoring spinal curvature for scoliosis, treating sleep apnea, and recognizing under ventilation have been identified as preventative measures (MDA, 2009). The early signs of respiratory complications and failure are sometimes misinterpreted by those with neuromuscular disorders; however, the earlier the symptoms are identified the more beneficial it will be. These symptoms may include: tiredness, sleep problems, headaches, weight loss, weakened cough or voice to name a few (MDA, 2009).

There are a number of respiratory supports available to individuals that range from preventive lung volume recruitment (McKim et al) using a hand-held resuscitation bag to stack breaths and increasing lung capacity and cough effectiveness through to the Cough Assist device. Unfortunately for some a tracheotomy, an invasive surgical procedure may be required. Adequate airway clearance and effective cough strategies remove mucus from the lungs and are important in preventing infections for those with muscular dystrophy (McKim et al., 2011). Cough assistance can take the form of manual support which includes a caregiver applying pressure to the abdominal area, or mechanically, through a device called the Cough Assist that removes secretions (MDA, 2009). **The use of Cough Assist machines was discussed as an important preventative strategy for those with neuromuscular disorders by a number of the key informants, but, continued research on proving its cost effectiveness is important in the future for advocacy purposes.** While the use of Cough Assist machines was identified as capable of preventing acute infections and reducing health care costs, **access to this equipment is also an issue.** In Ontario, for example, adults are eligible for partial or full funding of ventilators and related respiratory equipment through the Ministry of Health and Long-Term Care's (MOHLTC) Assistive Devices Program (ADP), while the Cough Assist tm machines are not funded under this same program (Assistive Devices Program, Ministry of Health and Long-Term Care, 2007). **Therefore, the current approach to**

**funding equipment in Ontario is not preventative in nature, but rather focuses on coverage for equipment needed in the later stages of the disorders.**

In addition to early identification and intervention, **it is important to discuss ventilation options and care plans with a health care practitioner before an acute respiratory complication or episode.** In the literature and through the interviews, it was discussed that many times patients and their families only discuss ventilation and other interventions once a tracheotomy is needed (Sritippayawan, Kun, Keens, & Ward, 2003). This reactive approach does not allow for patients and their families to carefully weigh their care options. For example, in their study of home mechanical ventilation (HMV) for children with neuromuscular disorders, Sritippayawan et al. (2003) found that, “HMV was initiated electively in 21% of patients with neuromuscular disease; 69% of the nonelective HMV group had HMV initiated after respiratory failure caused by pneumonia” (p. 481). The authors therefore concluded that conversations regarding ventilation are not adequately addressed with families before a crisis occurs.

This issue relates back to the lack of consensus on when people with neuromuscular disorders should begin respiratory support, and the lack of consistent practice regarding invasive and non-invasive approaches. This is due in part to patient caseload. Health care professionals who do not see many patients with neuromuscular disorders may be unnecessarily overcautious and prescribe a tracheotomy. However, there is evidence that non-invasive techniques may be preferred by the patient, and result in fewer hospitalizations, a longer lifespan and enhanced quality of life (Bach, 2012). Muscular Dystrophy Canada is currently developing educational handbooks for health professionals and also for patients and families to address this need.

Unfortunately, **the degree and quality of care one receives with respect to respiratory care depends on which province one lives in.** B.C. with its Provincial Respiratory Outreach Program (PROP) was frequently cited as a stellar model program for all of Canada. It was created by consumers and community care professionals and does not follow a medical model but rather addresses the full range of issues an individual may face living in the community. The PROP programs includes financial support, access to equipment, community care giver support, and to trained health professionals with expertise in respiratory care issues

One key theme that was identified throughout the informant interviews was the **decreased availability of support services as individuals with neuromuscular disorders transition from paediatric to adult care.** The challenges associated with transitioning to adult care are not just specific to neuromuscular disorders, but have also been discussed in the literature with regards to a number of different conditions (Rosen, Blum, Britto, Sawyer, & Siegel, 2003).

Thanks to advances in research and clinical care, children and adolescents with neuromuscular disorders are living longer and reaching adulthood; therefore, issues of transitioning are increasingly relevant today. Some children who are cared for by paediatric practitioners may continue to receive services from these professionals as they age; however, these practitioners may not have the expertise or resources that patients may require in adulthood (Panitch, 2006). In an article by Pantich (2006), he states:

Some of the challenges for ventilator-assisted young adults with neuromuscular disease include increased reliance on others for activities of daily living, funding for home-care skilled nurses or attendants, coordination of care among multiple subspecialists, and access to transportation, employment, and independent housing. (p. 891)

Decreased access to services and resources for adults that are often offered in childhood and adolescence was discussed by a number of the informants. A couple of the informants discussed a program offered in British Columbia called the At Home Program as an example. This program only offers funding support and other resources and services to children under the age of 18 (Government of British Columbia, n.d.).

The Muscular Dystrophy Association has created a website for adolescents and young adults with neuromuscular disorders called the “Transitions Resource Centre” to help bridge the gap discussed above. This website is specifically designed for this population of individuals with resources and information about living independently, including information on housing, transportation, and careers. The site also uses social media and connects adults with one another, with the goal of empowering this population (MDA, n.d.).

It is apparent that issues of transitioning are pertinent to adolescents and young adults with neuromuscular disorders today. **While there are supports and services for children with neuromuscular disorders, it is important to examine the gaps in services and resources for those transitioning to young adulthood and beyond.**

## Summary of Issues

In summary, the environmental scan and the key informant interviews highlighted some consistent themes related to issues of respiratory care. They include:

- Respiratory care among people with neuromuscular disorders results in significant financial cost to the health care system
- Significant provincial variances in financial support, access to equipment, and community care (B.C.s' Provincial Respiratory Outreach Program was repeatedly noted as a stellar model)
- Early intervention including funding of preventive equipment (e.g., LVR, Cough Assist) versus later stage equipment (ventilators) and early discussion on respiratory care options and plans before a respiratory complication or episode are important
- Transition from intensive care to an alternate level of care or community setting (lack of public funding programs, timely access to equipment, maintenance services and structured education programs)
- Transition from pediatric to adult care (i.e., decreased availability of support)
- Lack of awareness among health professionals about existing best practices in respiratory care
- No consensus on when people with neuromuscular disorders should start respiratory support and in what form (including when to transition from night time to day time as well) including initiating invasive approaches too early which can lead to a false sense of respiratory security
- Inadequate awareness among health providers of the extent of benefits of non-invasive approaches and the corresponding implications, as well as a failure to appreciate the burdens of invasive ventilation
- Lack of education for people with neuromuscular disorders on respiratory care and working with their practitioners
- Lack of high quality, targeted research that may provide evidence for advocacy efforts (e.g., research on effectiveness of Cough Assist mechanical devices to increase access to equipment choices)
- Lack of dissemination of existing research results to inform decision making

## Recommendations for Muscular Dystrophy Canada

### Advocacy

1. To advocate for a provincial respiratory outreach program (i.e., BC PROP) in other provinces and to ensure access to Cough Assist machines

### Education

2. To further education efforts to health professionals and people with neuromuscular disorders

### Research

3. To identify and support necessary targeted research and ensure the results are disseminated appropriately

## Potential Partners

All of the noted recommendations should be carried out in partnership with other like-minded organizations. Joining forces maximizes resources, efforts and leverages communication channels to priority audiences. Possible partners include clinicians, not-for-profit organizations, health care professional associations, government, hospitals and community care agencies, researchers and consumer support and advocacy groups.

## Impact on Muscular Dystrophy Canada

The recommendations in this report are fully aligned with Muscular Dystrophy's mission of enhancing the lives of those affected by neuromuscular disorders by continually working to provide ongoing support and resources while relentlessly searching for a cure through well-funded research. Addressing this issue will also assist in expanding the organization's national role in providing important education, advocacy programs and services. It will serve to enhance Muscular Dystrophy Canada's role in knowledge transfer and build stronger relationships and collaboration with partners. Properly executed,

this project can easily become an important signature program for the organization.

## Impact on people affected by neuromuscular disorders

The potential benefits and outcomes of this initiative include:

- Empowered and educated patient population
- Knowledge translation from research to practice in the clinical setting
- Enhanced access to breathing equipment
- Respiratory therapists and other health care providers will have enhanced expertise in neuromuscular disorders in the community and will be more available to clients
- Alleviating financial pressure on the health care systems

## Conclusion

Through the very cornerstones of Muscular Dystrophy Canada's mandate: education, advocacy and research, advances can be made to improve the health and quality of life of people with neuromuscular diseases. The purpose of this paper was to provide an overview of the issues and gaps related to respiratory care and to identify a possible realistic leadership role for Muscular Dystrophy Canada. In conclusion, in carrying out the research for this paper, informants and expert reviewers expressed their appreciation and encouragement of Muscular Dystrophy Canada to tackle this important issue.

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## Appendix 1

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### List of Key Informants & Reviewers

Peter Anthanasopoulos	Louise Rose
Canadian Paraplegic Association – Ontario	Researcher
Toronto, ON	St. Mike’s Hospital
	Toronto, ON
Sherry Katz	
Respirologist	Hugh McMillan
Children’s Hospital of Eastern Ontario	Pediatric Neurologist
Ottawa, ON	Children’s Hospital of Eastern Ontario
	Ottawa, ON
Hilda Perry	
Physiotherapist	Luke Melchoir
BC Children’s Hospital	Consumer
Vancouver BC	Victoria, BC
Simon Cox	Doug McKim
Executive Director	Medical Director
Technology for Independent Living	Respiratory Rehabilitation Services
Vancouver, BC	Ottawa, ON
Karen Rimmer	Dan Hughes
Respirologist	Pediatric Respirologist
Alberta Health Service	Halifax, NS
Calgary, AB	

## Appendix 2

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### Recommendations for Muscular Dystrophy Canada

#### Advocacy

1. To advocate for a provincial respiratory outreach program (i.e., BC PROP) in other provinces and to ensure access to Cough Assist machines

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3. To identify and support necessary targeted research and ensure the results are disseminated appropriately

## Appendix 3

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### Environmental Scan

**See Separate Excel Document**