INVESTIGATION OF PHYSIOLOGICAL RESPONSES DURING PULMONARY AND EXERCISE STRESS TESTS AND VALIDITY OF THE WHEEL PERCEIVED EXERTION SCALE AMONG ADOLESCENTS AND ADULTS WITH SPINA BIFIDA

by

Yu-Ting Cheng

Bachelor of Science, Physical Therapy, Chung Shan Medical University, 2006

Submitted to the Graduate Faculty of

School of Health and Rehabilitation Sciences

in partial fulfillment of the requirements for the degree of

Master of Science in Rehabilitation Science and Technology

University of Pittsburgh

2014

UNIVERSITY OF PITTSBURGH

SCHOOL OF HEALTH AND REHABILITATION SCIENCES

This thesis was presented

by

Yu-Ting Cheng

It was defended on

July 25, 2014

and approved by

Brad E. Dicianno, M.D., Associate Professor, Department of Physical Medicine and Rehabilitation

Dan Ding, PhD, Associate Professor, Department of Rehabilitation Science and Technology

Ashli M. Molinero, D.Sc, Assistant Professor, Department of Rehabilitation Science and Technology

Thesis Director: Theresa M. Crytzer, PT, DPT, ATP, Assistant Professor, Department of Rehabilitation Science and Technology

INVESTIGATION OF PHYSIOLOGICAL RESPONSES DURING PULMONARY AND EXERCISE STRESS TESTS AND VALIDITY OF THE WHEEL PERCEIVED EXERTION SCALE AMONG ADOLESCENTS AND ADULTS WITH SPINA BIFIDA

Yu-Ting Cheng, MS

University of Pittsburgh, 2014

People with spina bifida have hypoactive lifestyles that are the consequence of lower extremity functional limitations. However, several secondary conditions, such as scoliosis, pneumonia, and obesity may affect activity performance. Even socio-demographic factors, such as lack of accessible transportation to fitness facilities or having low income may affect activity performance as well. Few studies have investigated pulmonary function in people with spina bifida. In the present study, pulmonary function tests (PFTs) and graded arm ergometry exercise stress tests were conducted among adolescents and adults with spina bifida (n = 29). The primary aim of this study was to investigate the relationship between pulmonary function and exercise capacity among people with spina bifida. Another aim of this study was to develop and to validate a newly developed perceived exertion scale (the WHEEL Scale) for regulating exercise intensity for people with spina bifida. Socio-demographic information, body composition measurements, and medical record information were collected in this study and were used as predictors for PFTs and peak oxygen consumption (peak VO_2). The results showed that more than 65% of participants met the criteria of pulmonary restriction and 90% of the people who were not able to achieve a maximal exercise test had pulmonary restriction. Significant models for predicting PFTs and peak VO_2 were found and established in this study. The concurrent validity and construct validity of the newly developed WHEEL Scale were established by using relative heart rate (HR) and relative peak VO_2 from the graded arm ergometry exercise stress

test. Concurrent validity of the WHEEL Scale was established by the finding that the WHEEL scale significantly correlated with relative VO_2 and relative HR. Construct validity of the WHEEL Scale was established by the finding that the WHEEL scale significantly correlated with the Borg Scale. Future studies in a large cohort of individuals with spina bifida are needed to confirm the results and establish the inter-rater and intra-rater reliability of the WHEEL Scale.

TABLE OF CONTENTS

LIST OF TABLES

LIST OF FIGURES

PREFACE

My two-year journey in the United States has been filled with unforgettable and treasurable memories that will last a lifetime. I have been lucky enough over the last two years to meet and work with some amazing people who helped me in the rehabilitation research field and in the assistive technology services system. First of all, I would like to deliver my sincerest gratitude to my advisor, Dr. Theresa Marie Crytzer, who is always generous, patient, and supportive. It is truly an honor and a valuable experience to work and collaborate with her on research and in the clinic. Dr. Crytzer is not only a mentor for me, but plays an important role in my master student life.

Thanks to the Ashley Rose Foundation and the Spina Bifida Association for the funding resources. If this thesis has contributions to the rehabilitation research field, the credit should go to all the participants and the research crew in this study. Thanks to Dr. Sciurba, Mary J. Bryner, and Robert C. Wilson from UPMC Emphysema COPD Research Center, and thanks to Steve J. Anthony and Shannon A. Speight from UPMC Endocrinology and Metabolic Research Center, without their efforts, support, and help, this study would not have been possible.

I am so grateful to my thesis committee members, Dr. Dicianno, Dr. Ding, and Dr. Molinero, for whose valuable suggestions and constructive comments on this thesis that make it more meaningful and have more clinical significance. Also, I would like to take this opportunity to appreciate all professors, faculty, staffs, and students at Human Engineering Research

Laboratories, UPMC Center for Assistive Technology, and HGAC Center for Assistive Rehabilitative Technology.

There is an old saying “one would rely on friends when out of home.” Thanks to Buddha for looking after me by giving me so many lovely and wonderful friends in Pittsburgh. Thank you Wendy Li-Yun Chang, Meng-Hsuan Chou, YingJu Yu, Rachel Tsai-Ling Huang, Dwight Liu, Eric Wu, Ying-Chen Chen, and Owen Tsai for your accompany in the last two years. I was fortunate to have you all here and to create so many treasurable moments together! I would also like to thank all my friends, colleagues, and mentors from the Research Center on International Classification of Functioning, Disability and Health and Assistive Technology (RICFAT) in my beloved hometown, Taiwan. This small and beautiful island nourishes me and gives me full energy and courage to take adventure abroad, she makes me understand that the farther you away from home, the more you feel close to it. Also, special thanks to Professor Shwn-Jen Lee, Professor Hsin-Hsien Yeh, Dr. Li-Shan Chang, Jhih-Ren Wang, and Shao-Yi Yen, without your continuous support and encouragement, I would not be able to complete this journey.

Finally, I would like to express gratefulness to my family. I owe a great deal to my family. I am so fortunate to have such a supportive family in my life. My beloved parents, Chih-Shiang Cheng and Hsiu-Chih Lee, your love always keep me warm and directs me on the right path. My beloved two brothers and two sisters-in-law, whose love always accompanies me and allows me to go further. Also, special thanks to the Kung’s family, the Hsaio’s family, and the Lai’s family, you guys are so amazing.

Thanks to all the people I met in Pittsburgh, in Taiwan, and I love you all so much.

1.0 INTRODUCTION

Spina Bifida, a type of spinal dysraphism, is a neural tube defect (NTD) that results from failure or incomplete closure of the neural tube in the process of embryogenesis. An estimated incidence of spina bifida in the United States is around 0.3-0.4 per 1,000 live births with 1,500 cases annually (Center for Disease control and Prevention, 2014; Parker et al., 2010,). The classification system of spina bifida varies depending on whether the purpose is for clinical management or for research purposes (Özek, Cinalli, & Maixner, 2008). In 2000, Dr. Tortori-Donati and colleagues proposed a classification system of spinal dysraphism based on the relation between clinio-neuroradiological, pathological aspects and embryological development, which yielded more than 20 types of spinal dyraphism. However, the most commonly seen and discussed in textbooks, literature, research and websites are the following three types of spina bifida: spina bifida occulta, meningocele, and myelomeningocele (Figure 1) (Botto, Moore, Khoury, & Erickson, 1999; Spina Bifida Association, 2014). Among these three types of spina bifida, meningocele and myelomeningocele are usually referred to as spina bifida aperta or spina bifida cystica (Özek et al., 2008; Spina Bifida Association, 2014).

The cause of neural tube defects, such as anencephaly or spina bifida, remains unclear. Genetic and environmental influences are thought to play a role (Özek et al., 2008); other risk factors that have been shown to increase the risk of neural tube defects include folate deficiency,

maternal obesity, diabetes, and anti-convulsion medication (Melvin et al., 2000; Özek et al., 2008; Shaw, Velie, & Schaffer, 1996).

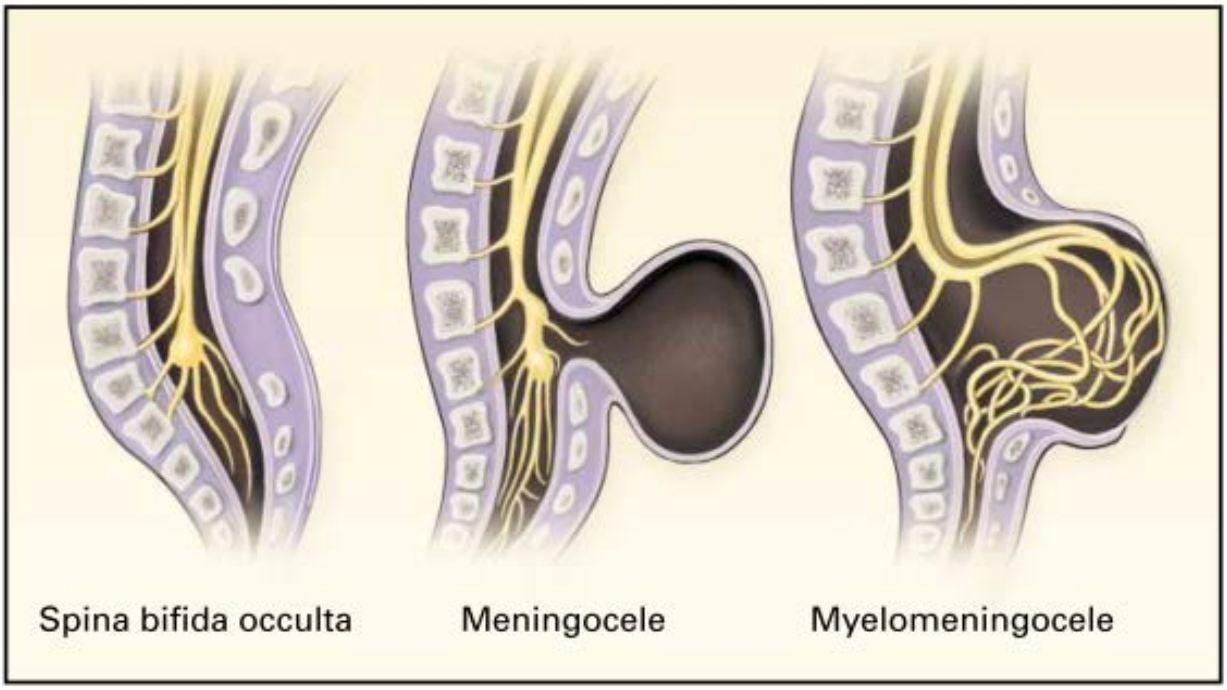

Figure 1. Sagittal view of the spinal cord in three types of spina bifida

1.1 MEDICAL ASPECTS AND FUNCTIONAL CAPACITY OF SPINA BIFIDA

Myelomeningocele is the most common and the most severe form of spina bifida. It is a condition that involves the nerves of the spinal cord protruding through the defect in the vertebrae to the outer part of the body. The majority of lesions are in the lumbosacral region (Falvo, 2013). Because the central area protrusion has no subcutaneous covering, the vascular tissue and spinal nerves are exposed, and cerebrospinal fluid or tissue transudate may leak, which increases the risk of infection while the fetus is in the uterus (Özek et al., 2008). The manifestation of spina bifida depends on which part of the spinal cord is affected, as well as the

severity of the lesion. The severity can range from mild with a few manifestations to severe. Severe lesions could include muscle paralysis, loss of sensation, loss of bowel and bladder control, hydrocephalus, Chiari II malformation, tethered cord syndrome, cognitive impairment, and sleep apnea. The treatment or management of spina bifida depends on the extent of neurologic problems, the functional level of lesion, and the existence of any complication (Dicianno et al., 2008; Falvo, 2013; Özek et al., 2008). Advancements in medical science, surgical technique, rehabilitation management, and folic acid fortification have improved the survival rate and life expectancy of people with spina bifida (Boulet et al., 2008; Dicianno et al., 2008). Although spina bifida is not a progressive condition, other complications and health issues may be associated with it during the lifespan. Secondary complications include renal dysfunction, cardiac disease, pulmonary dysfunction, urinary tract infection, scoliosis, joint deformity, decubitus ulcer, visual impairment, obesity, latex allergy, and cognitive issues (Dicianno et al., 2008; Falvo, 2013; Friedman et al., 2009; Özek et al., 2008; Spina Bifida Association, 2014). Thus, there is a growing need for research to improve health status, increase independence, and enhance quality of life for individuals with spina bifida.

The World Health Organization (WHO) revised the classification system to provide a new insight into a person's health and well-being in the form of the "International Classification of Functional, Disability, and Health (ICF)" (World Health Organization, Geneva, 2010). In contrast to the medical model that focuses on curing impairment or disease, the ICF model (Figure 2) addresses the dynamic nature of a person's health condition, meaning that the individual's health status is an interrelationship between body structure and function, activity, participation, environmental factors and personal factors. The ICF model was used in previous studies as a conceptual framework to examine the health condition and secondary conditions

among people with spina bifida. The results showed that general health condition and social participation in this population were low. But the secondary conditions, such as obesity, scoliosis, and reduced participation would be preventable by identifying the mechanisms associated with individuals' manifestations (Liptak et al., 2010; Simeonsson et al., 2002).

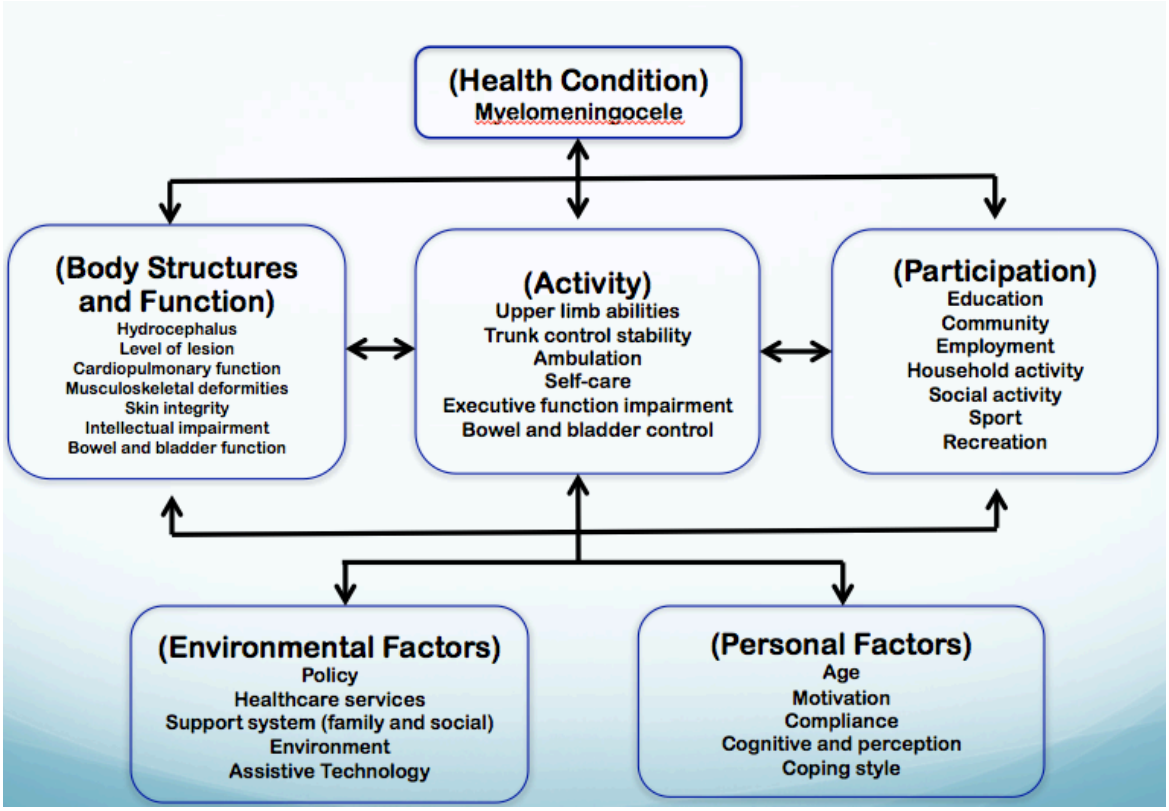

Figure 2. An illustration of the interaction concepts in the ICF model: using myelomeningocele as an example

Dicianno and Wilson (2010) conducted a retrospective study that analyzed hospitalization admissions among people with spina bifida and found that over one third of admissions were from potentially preventable conditions with 35.7% resulting in death from these conditions. These preventable conditions that resulted in hospitalization and death include: cardiovascular and pulmonary complications, such as congestive heart failure (CHF), acute cerebrovascular disease, pneumonia, and chronic obstructive pulmonary disease (COPD). Another research study also found that metabolic syndrome, a constellation of known cardiovascular disease risk factors,

was prevalent (32%) among participants with spina bifida, and especially higher prevalence (45.8%) was found in participants who were obese (Nelson et al., 2007).

1.2 CARDIOPULMONARY FUNCTION OF SPINA BIFIDA

Metabolic syndrome and obesity are a constellation of known risks for the development of cardiovascular disease (CVD). Individuals with spina bifida who are obese have a 45% higher risk of developing metabolic syndrome than individuals who are not obese (Nelson et al., 2007). A cross-sectional study among people with spina bifida also showed that the obesity rate was higher in adults than in children and adolescents (Dosa et al., 2009). Buffart et al. (2008) reported that people with spina bifida who were non-ambulatory tend to have increased risk factors of CVD. Several research studies were conducted to investigate the relationship between metabolic syndrome, obesity, physical fitness, and exercise capacity within this population. Exercise testing is the gold standard used to investigate aerobic fitness and capacity. Two studies in the Netherlands demonstrated that the spina bifida population had 32% lower aerobic fitness than the general population and 23% lower than a spastic diplegia group (Buffart et al., 2008; van den Berg-Emons et al., 2013). As for functional capacity in this population, one study conducted by Abresch et al. (2007) showed that children and adolescents with spina bifida had a lower score of health-related quality of life (HRQL) in school functioning and social functioning sessions compared to those with spinal cord injuries who had a similar lesion level. As for pulmonary function in this population, there was one study in 1997 that reported that children with spina bifida had restrictive lung disease and had lower exercise capacity compared to

control subjects (Sherman et al., 1997). Research studies that investigated the pulmonary function and exercise capacity among people with spina bifida are limited, and given that cardiopulmonary complications are recognized as life-style related conditions and could be prevented by regular physical activity or exercise (ACSM, 2010), the aim of this study was to conduct pulmonary and exercise tests among adolescents and adults with spina bifida in order to investigate their cardiopulmonary functions. Due to the lower exercise capacity and lower participation in the spina bifida population, the maximal heart rate that is used for regulating exercise intensity may not be appropriate for this population. An exercise intensity tool for clinicians to regulate and prescribe exercise intensity for people with spina bifida is also investigated in this study.

2.0 THE RELATIONSHIP BETWEEN BODY MORPHOLOGY AND PULMONARY FUNCTION AMONG ADOLESCENTS AND ADULTS WITH SPINA BIFIDA

2.1 INTRODUCTION

Spina bifida is not a progressive condition, however, secondary complications and other health issues may be associated with it across the lifespan, such as renal dysfunction, cardiac disease, pulmonary dysfunction, urinary tract infection (UTI), joint deformity, decubitus ulcers, scoliosis, and obesity (Buffart et al., 2008, Dicianno et al., 2008, Dosa et al., 2009). The advancements in medical and rehabilitative sciences, and improvements of health-related quality of life issues are gaining more attention in recent decades. Examples of such advancements and improvements include: spina bifida fetal surgery for improving the prognosis of locomotion (Adzick et al., 2011), rehabilitation approach and assistive devices for increased mobility and independence (de Groot et al., 2007, Lynch et al., 2009), and urologic care management and bowel management (Verhoef et al., 2005). A retrospective study reported that pneumonia (9.4%) and respiratory failure (7.0%) were the second and the third leading causes of death respectively. Furthermore, pneumonia (6.5%) was the leading cause of admission in the age group over 65 years (Dicianno et al., 2010). Pneumonia is an infection of the lungs that can be caused by viruses, bacteria, and fungi. There are risk factors of pneumonia, including cigarette smoking, respiratory infection, difficulty swallowing, chronic lung disease, heart disease, living in a nursing facility, impaired

consciousness, having a weakened immune system, or other underlying medical conditions, among others. (American Lung Association, 2014; Center for Disease Control and Prevention, 2014). Reports have shown that people with disabilities, such as spinal cord injury and cerebral palsy, develop impairment of respiratory capacity due to neurological injuries, poor ability for lung expansion, muscle constriction, or muscle weakness due to degeneration of motor neurons (Staggenborg 2009, Galieras Vazquez et al., 2013, Young et al., 2011). For example, people with traumatic spinal cord injuries who had higher lesion level had a higher risk of respiratory dysfunction resulting from inability to expand the chest cavity and disruptions of nerve impulses that control brain signal to respiratory muscles (Galieras Vazquez et al., 2013). Limited research investigated the relationship between level of lesion and pulmonary function among people with spina bifida. One study found that 58% youth with spina bifida had restrictive lung disease, which is thought to be a predisposing factor for pneumonia (Sherman et al., 1997). However, they had a small sample size (n=12) with youth of spina bifida (13.1 ± 2.7 years) and provide no information regarding the relationship between participants' medical history, body composition, and pulmonary function. Another study retrospectively examined the relationship between the degree of scoliosis and pulmonary function among children with spina bifida (n=32, mean age: 14 years). The results showed that most of the participants displayed a restrictive pattern of lung function, and scoliosis was negatively correlated ($R^2 = 0.31$, $p < 0.05$) with maximum expiratory flow over the middle 50% of the vital capacity (FEF 25-75%), but a greater degree of scoliosis was not related to reduced FVC (Patel et al., 2011).

In order to provide appropriate approaches and intervention for reducing the secondary complications among people with spina bifida, identifying individuals at risk of restricted pulmonary function and pneumonia would be important. In light of limited studies that focus on

pulmonary function among people with spina bifida, the aim of this study was to investigate the pulmonary function and exercise capacity among adolescents and adults with spina bifida. This session examined the relationship between body composition and pulmonary function by conducting the pulmonary function tests. The next session further examined the relationship between exercise capacity and pulmonary function by conducting a graded exercise stress test.

2.1.1 Aims and hypotheses

Aims:

1. To investigate the pulmonary function of adolescents and adults with spina bifida.
2. To examine the relationship among body composition, physical activity, and pulmonary function of adolescents and adults with spina bifida.

Hypotheses:

1. More than 60% of participants will meet the criteria for restrictive lung disease.
2. A significant difference will found between PFTs results in the restricted lung condition group and non-restricted lung condition group.
3. Body mass index (BMI), level of lesion, and spinal fusion will be the most significant predictors of forced vital capacity (FVC).
4. Body mass index (BMI), level of lesion, and spinal fusion will be the most significant predictors of forced expiratory volume in one second (FEV_1).
5. Body mass index (BMI), level of lesion, and spinal fusion will be the most significant predictors of total lung capacity (TLC).

2.2 METHODS

2.2.1 Recruitment procedures

Adolescents and adults with spina bifida were recruited through flyers and clinician referral from spina bifida clinics in the southwestern Pennsylvania area and the University of Pittsburgh Medical Center (UPMC) Center for Assistive Technology. Participants were screened during a phone interview prior to enrollment based on inclusion/exclusion criteria. Inclusion criteria were: (a) age 13 – 80 years, (b) having spina bifida but not of the occulta type, (c) having scoliosis, and (d) inability to pedal a standard (two-wheel) bicycle. Exclusion criteria were: (a) having a history of coronary artery disease, coronary bypass surgery, or other cardiopulmonary events, (b) upper extremities injury or loss of shoulder, elbow, and/or wrist range of motion that would prevent performing arm ergometry exercise testing, (c) upper extremity or thoracic surgery in the last 6 months that would be a contraindication to perform arm ergometry exercise testing, and (d) any other medical condition for which the participant's primary care physician determined was a contraindication to arm ergometry exercise testing.

A written medical clearance from the participant's primary care physician was required prior to participation. Once participants were screened and medical clearance received, participants were scheduled for a one-time visit to the Emphysema and COPD Research Center and the Endocrine and Metabolic Laboratory at UPMC Montefiore Hospital. A welcome letter with directions on how to get to the hospital and pre-test instructions were mailed to participants. Participants were informed that the testing process required up to four hours. The pre-test instructions were provided through a phone call to participants and included the following

information: (a) DO NOT do any strenuous physical activity a day before the test, (b) DO NOT eat anything four hours before the test, unless you need a light snack with your medications, (c) DO NOT change the medication routine prescribed by your physician, (d) DO NOT drink alcohol or caffeine four hours before the test, (e) DO NOT smoke one hour before the test, and (f) wear comfortable clothing that allows you to move your arms easily and that you can exercise on the test day.

Before signing written informed consent, participants completed the MacArthur Competence tool (Appendix A) (Grisso et al., 1997) to confirm their ability to understand the study. All participants, and parents of adolescents aged less than 18 years, were required to have a score of 8 out of ten on the MacArthur to participate. Participants were provided with a copy of an unsigned consent form to protect their confidentiality. This study was approved by the University of Pittsburgh Institutional Review Board.

2.2.2 Experimental protocol

The experimental protocol included three parts during the one-time visit. The first part included a socio-demographic survey (Appendix B), body composition measurements (Appendix C), and a physical activity assessment (Appendix D). Each participant started with a socio-demographic survey, which includes gender, race, education, employment, income, health condition, current mobility device, transportation, smoking history, living situation, level of lesion, assistant service, and exercise habits. After collecting the participant's demographic information, anthropometric measures were conducted. Body composition data were gathered, including participant's height, weight, arm span, waist circumference, handgrip strength, and triceps

skinfold were obtained by a physical therapist. All participants completed the timed 10-meters wheel test twice with their regular propel speed.

After socio-demographic data were collected, participants were accompanied to the Emphysema and COPD Research Center for pulmonary function tests (Appendix E). The pulmonary function test was conducted by a certified pulmonary function technologist. Instructions regarding the testing process, demands, and feedbacks were given to participants throughout the entire process. Participants were accompanied to the Endocrine and Metabolic Laboratory for a graded exercise stress test (Appendix F). The graded maximal exercise test was administered by an exercise physiologist and a physical therapist. Participants performed the exercise test by using an arm ergometer while sitting in their wheelchair while pedaling the arm crank. A ramp and platform were used for participants with low seat to floor height of the wheelchair. If participants used manual wheelchairs, a set of four blocks was positioned behind both side rear wheels and front casters to enhance stability while pedaling. Instructions such as pedaling speed and the use of the rating of perceived exertion scales were given at the beginning of the exercise test. Verbal reinforcement was provided by investigators during exercise test.

2.2.2.1 Socio-demographic survey and body composition measurements

Participant's basic information was collected from a socio-demographic survey, including age (calculated in years from participant's date of birth to the testing date), gender (categorized to male, female, and transgender), race (categorized to Caucasian, Black or African American, Native Hawaiian or Pacific Islander, American Indian or Alaskan Native, and other), education (was categorized to 8^{th} grade/less, 9^{th}-11^{th} grade, graduation from high school/GED, trade or

vocational school, some college/associate degree, college graduate (4 or 5 year program), some graduate school, and graduate degree), employment (was categorized to full-time, part-time, full-time student, part-time student, homemaker, retired, not retired but not currently employed, part-time volunteer, and other), income (was categorized to less than $10,000 a year, $10,000 to $19,999 a year, $20,000 to $34,999 a year, $35,000 to $49,999 a year, $50,000 to $74,999 a year, and $75,000 or more), primary mobility device (categorized to none, crutches/cane/walker, manual wheelchair, and power wheelchair), transportation method (categorized as own a car, have access to family car, rely on families or friends to drive, use Access, and use public transportation), self-reported smoking history (categorized as none and currently smoke/smoked in the past), living situation (categorized as live at home with family, live independently in own home, live independently in an apartment building, and live in supported living), self-reported health status (categorized as excellent, very good, good, fair, and poor), involvement of adaptive sport (categorized as none/involved in the past and involve now), and self-reported exercise hours (recording in hours per week). Type and manufacture of wheelchairs (categorized as no walking aid, K-0005, and power wheelchair) and seat cushions (categorized as no seat cushion, foam, gel, air, and mix) were also collected in this study as record.

Body composition measurements include height (measured in meters (m) from participant's head to heel by using a measuring tape), weight (measured in kilograms (kg) by using a wheelchair accessible scale - CR1000D (DETECTO - division of Cardinal Scale Manufacturing CO, Webb city, MO), arm span (measured in m from one side of participant's middle finger across the back through the other middle finger while participant was in the sitting position with arm spread out by using a measuring tape), waist circumference (measured in centimeters (cm) at umbilicus level while participant was in the sitting position by using a

measuring tape), hand grip strength (measured in kg by using JAMAR hand grip dynamometer (Sammons Preston Roylan Inc., Chicago, IL), triceps skinfold (measured in millimeters (mm) by using Lange skinfold caliper (Beta Technology Inc., Santa Cruz, CA). Because of using standard height to calculate body mass index (BMI) would underestimate the lung condition (Sherman et al., 1997) and overestimate BMI in non-obese spina bifida population due to short torsos of individuals with higher level of lesion (Nelson et al., 2007), the length of arm span was substitute for standard height in this study. BMI (expressed in units of kg/m^2) was calculated based on modified BMI formula with participant's weight divided by participant's arm span squared).

Physical activity was assessed by two functional measurements, one is the physical activity scale for individuals with physical disabilities (PASIPD) and the other is 10-meter wheel test. The PASIPD (expressed in MET-hour/day) is a seven-day recall physical activity questionnaire for people with physical disability (Washburn et al., 2002) and has been validated in physically disable populations (van der Ploeg et al., 2007, Washburn et al, 2002). The Ten-meter wheel test (10 MWT), was validated among manual wheelchair users and measures the time in seconds (sec) that it takes a participant to propel a manual wheelchair ten meters on a smooth level surface with regular propulsion speed (Askari et al., 2013).

Medical history variables were collected from an electronic medical record system, such as functional level of lesion (categorized to thoracic and non-thoracic), scoliosis (categorized as Cobb angle less than 30 degrees and Cobb angle larger than 30 degrees), spinal fusion (categorized as had spinal fusion or no spinal fusion), Chiari II malformation, Chiari II malformation surgery (categorized to presence of Chiari II malformation or no presence of Chiari II malformation), sleep apnea (categorized to presence of sleep apnea or no presence of sleep apnea), and history of lung condition (categorized as having a history of lung condition

including pneumonia, bronchitis, pleural effusion, and lung hypo-inflation, or no history of lung condition).

2.2.2.2 Pulmonary function tests

Pulmonary function tests (PFTs), including spirometry and lung volumes tests, were both measured by using the SensorMedics Autobox Respiratory Analyzer - V6200 (SensorMedics Corp., Yorba Linda, CA) (Figure 3). The SensorMedics Autobox Respiratory Analyzer is a plexiglass chamber with a captain seat and mouthpiece inside the box and is combined with a transducer and computer system that tests and reports the participant's pulmonary function. To reduce the risk of falling while the wheelchair user was transferring, a physical therapist and a trained investigator were directly next to each participant during the transfer and provided assistance. A transfer board was provided as needed. After the participant transferred into the chamber, the certified pulmonary function technologist explained and demonstrated the entire testing process. The pulmonary function test was conducted by a certified pulmonary function technologist in a laboratory setting with protocolized instructions to improve the reliability of the pulmonary function tests.

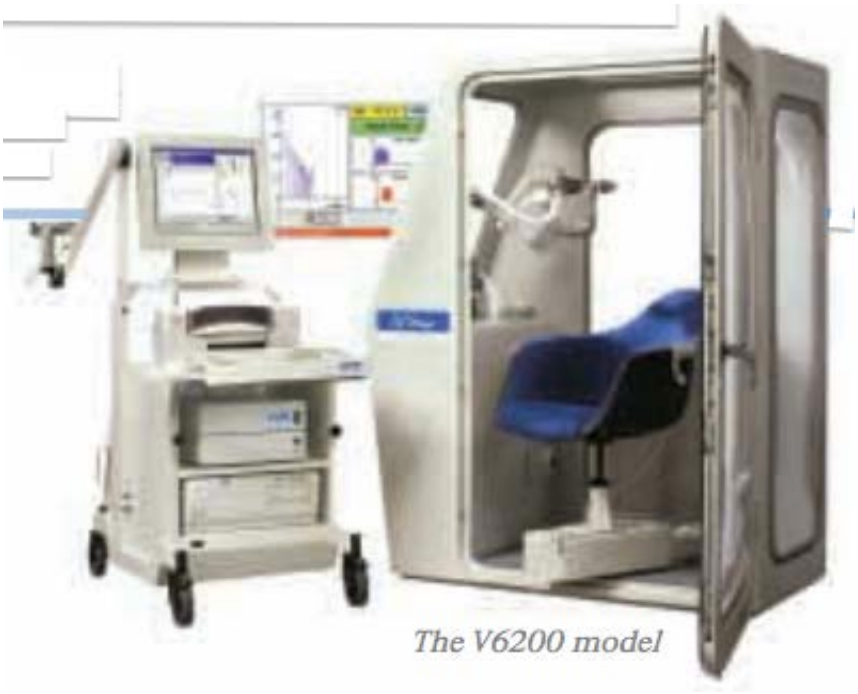

Figure 3. SensorMedics Autobox Respiratory Analyzer, V6200 model

Spirometry is a noninvasive test of ventilatory function that assesses the pulmonary system by providing measurements of the dynamic lung volumes and capacities (Pierce, 2005). Spirometry measures the amount and speed of air that can be inhaled and exhaled during forced expiration and inspiration, so that how effective and fast the lungs can be emptied and filled can be determined (Figure 4). Thus spirometry gives an overall assessment of lung function suitable for differentiation of various respiratory diseases, such as restrictive lung disease and obstructive lung disease. Measurements include forced vital capacity (FVC): a measurement of lung size and represents the volume of air in the lungs that can be exhaled following a deep inhalation; forced expiratory volume in one second (FEV_1): a measurement of how much air can be exhaled in one second following a deep inhalation; the ratio of FEV_1/FVC: the number represents the percentage of the lung size that can be exhaled in one second; FEF 25%-75%: the maximum

expiratory flow over the middle 50% of the vital capacity; peak expiratory flow (PEF): a person's maximum volume of expiration; and peak inspiratory flow (PIF): a person's maximum volume of inspiration. During the spirometry test, all of these measurements were reported and at least three trials and a maximum seven trials of testing were conducted to get two reproducible results.

Figure 4. Spirometry test

The lung volumes test measures how much air the lung can hold. The lung volumes test was conducted with the participant sitting in the closed chamber. Following the technician's instructions participants gently inhaled and exhaled against a closed shudder in the mouthpiece while wearing a nose clip and holding the cheeks (Figure 5). Three to five technically satisfactory panting maneuvers were recorded. In this test, functional residual capacity (FRC):

the measurement of the amount of air in your lungs at the end of a normal exhaled breath; total lung capacity (TLC): the measurement of the amount of air in your lung after you inhale as deeply as possible; vital capacity (VC): the measurement of the amount of air you can exhale after you inhaled as deeply as possible; expiratory reserve volume (ERV): the measurement of the difference between the amount of air in your lungs after a normal exhale and the amount after you exhale with force; and inspiratory capacity (IC): the measurement of the volume of air that can be taken into the lungs in a full inhalation were measured by using body plethysmography. Because residual volume (RV) cannot be obtained from the test, ERV was subtracted from TLC to derive RV.

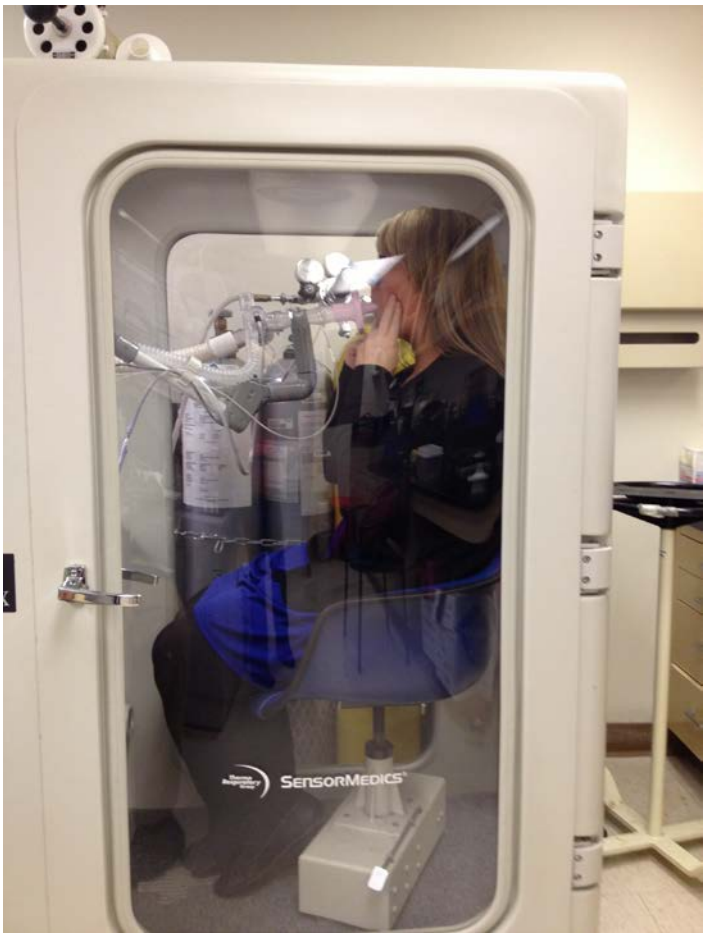

Figure 5. Lung volumes test

2.2.3 Data collection and data analysis

Socio-demographic information, body composition measurements, pulmonary function tests results, and graded maximal exercise test results were gathered within a one-time visit. The reference standards used for computing the PFTs values were the third National Health and Nutrition Examination Survey (NHANES III) and Crapo et al. (1981). The percent of predicted values were used in the analyses since these values are adjusted for age, height, weight, and gender. Arm span was used as a proxy for height. The interpretation of spirometry and lung volumes results for determination type of lung disease was made by a pulmonologist. Restrictive lung disease was identified as FVC < 80% or TLC < 80% with a FEV_1/FVC ratio > 70%. Obstructive lung disease was identified as FEV1 < 80% and FEV_1/FVC ratio < 70%. Descriptive results were presented with mean ± standard deviation or number and frequency. An independent *t* test was used to evaluate whether there was a significant difference between the two groups. All parameters that were obtained in this study were first analyzed using Pearson's correlation coefficient and then backward regression analysis was used to determine which parameter would strongly predict the PFTs results. Statistical analyses were performed using IMB SPSS version 22.0 for Windows (IBM Corporation, Armonk, NY); $p \leq 0.05$ were considered significant.

2.3 RESULTS

2.3.1 Demographic characteristics of all participants

The socio-demographic data of all participants are listed in Table 1. Twenty-nine participants (15 females) who had a diagnosis with spina bifida were recruited. Out of the 29 participants, two subjects (6.9%) were adolescents. The age of participants ranged from 17 to 71 years old (mean= 30 years, standard deviation (SD) = 12 years). The majority of participants were Caucasian (89.7%). Fifty-eight percent of participants completed college, and 8 participants were full-time students. Three participants (10.3%) reported working part-time and 31% of participants were unemployed. Thirteen (44.8%) participants lived at home with family; thirteen (44.8%) participants lived independently in their own home or in an apartment. The number of participants who lived in a supported living environment was 3 (10.3%). Further, 23 participants (79.3%) received 17.22 ± 22.24 hours/week (ranged from 2 - 81 hours/week) of paid or unpaid assistance from families or friends. As for transportation, 21 participants (72.4%) relied on family or friends or used public transportation; 8 participants (27.6%) owned a car or had access to family car. A manual wheelchair or power wheelchair was the primary mobility-assisted equipment used by twenty-two participants (75.8%). The type of all manual wheelchairs belonged to the K0005 category in the Centers for Medicare and Medicaid Services (CMS) the Healthcare Common Procedure Coding System (HCPCS) code set, which means these were ultra-lightweight and fully customized manual wheelchairs. Five participants (17.2%) used walking aids or braces, and 2 participants (7%) could walk without any walking aids or brace. The general health condition of the participants was self-reported as being in good condition or

better in 90% of participants. Only 3 participants reported as fair condition. The number of people who currently smoke or have smoked in the past was 8 (27.6%), and 21 (72.4%) participants had no smoking experience. Nineteen (65.5%) participants stated that they currently exercise and their average exercise time was 3.8 ± 2.8 hours per week (ranged of 0.5 - 9 hours/week).

Table 1. Socio-demographic data for all participants

n = 29			
Ordinal of dichotomous variables		n	Percentage (%)
Gender	Male	14	48.3
	Female	15	51.7
Race	Caucasian	26	89.7
	Black or African America	2	6.9
	Other	1	3.4
Education	9^{th}-11^{th} grade	4	13.8
	Graduated from high school / GED	4	13.8
	Trade or vocational school	4	13.8
	Some college / Association degree	14	48.3
	College graduated (4 or 5 year program)	3	10.3
Employment	Part-time job	3	10.3
	Student, full-time	8	27.6
	Homemaker	1	3.4
	Not currently employed, retired	1	3.4
	Not currently employed, not retired	9	31.0
	Volunteer, part-time	6	20.7
	Other	1	3.4
Income	Less than $10,000 a year	11	37.9
	$10,000 - $19,999 a year	5	17.2
	$20,000 - $34,999 a year	1	3.4
	$35,000 - $49,999 a year	3	10.3
	$50,000 - $74,999 a year	2	6.9
	$75,000 or more a year	1	3.4
	Do not know	6	20.7
Living situation	Live at home with family	13	44.8
	Live independently in own home	1	3.4
	Live in an apartment building independently	12	41.4
	Live in a supported living environment (not with family)	3	10.3

Table 1. (Continued)

Transportation	Own a car	7	24.1
	Have access to family car	1	3.4
	Rely on family or friends	7	24.1
	Use Access	13	44.8
	Use public transportation	1	3.4
Mobility-assisted equipment	None	2	6.9
	Lofstrand crutches	1	3.4
	Manual wheelchair	19	65.5
	Power wheelchair	3	10.3
	Walker	1	3.4
	Standard crutches	1	3.4
	Cane	1	3.4
	Other	1	3.4
Perceived health condition	Excellent	6	20.7
	Very good	10	34.5
	Good	10	34.5
	Fair	3	10.3
Smoking history	Not smoke	21	72.4
	Currently smoke or smoked in the past	8	27.6
Currently exercise	Not exercise	10	34.5
	Currently exercise	19	65.5

Continuous variables	Mean ±SD	Range
Age (year)	30.48 ± 12.51	17 - 71
Assistance time (hour/week)[1]	17.22 ± 22.24	2 - 81
Exercise time (hour/week)[2]	3.87 ± 2.79	0.5 - 9.0

[1] Assistance time: hours of assistance from other people per week, n= 23;
[2] Exercise time: self-reported hours of exercise per week, n= 19

Body composition measurements of all participants are listed in Table 2. All variables were analyzed by SPSS (Statistics Premium Edition version 22.0 for windows, IBM Corporation, Armonk, NY) and presented by mean ± SD and minimum value to maximal value. People with spina bifida tend to have a short statue due to scoliosis, undeveloped lower extremities, and lower extremity joint contractures, thus using height in the BMI calculation would underestimate the obesity rate in this population (Dosa et al., 2009). The length of arm span was used to calculate BMI. The method of categorizing BMI and disease risk for type 2 diabetes,

hypertension, and cardiovascular disease (CVD) was based on National Heart, Lung, and Blood Institute (National Institutes of Health, 2014) (Figure 6 & Table 3). Handgrip strength was measured in both the right hand (mean= 28.72, SE= 1.46) and left hand (mean= 27.31, SE= 1.60). The physical activity scale for individuals with disabilities (PASIPD) is a questionnaire that asks for self-reported physical activity in the past 7 days. Participants were asked to recall any physical activity involved in completing housework tasks, participating in recreational physical activity, and conducting work and caretaking duties. Every participant completed the ten-meter wheel test (10MWT). Participants who used manual wheelchairs (n = 24) during the 10MWT were reported separately in the list. A paired *t* test showed no significant difference between trial 1 and trial 2 for the 10MWT. The velocity of 10MWT was calculated by dividing distance (ten meters) by time (seconds).

Table 2. Body composition measurements, physical activity score, and mobility test of all participants

n = 29			
Variables		Mean ± SD	Range
Weight (kg)		64.08 ± 17.71	39.8 - 97.6
Height (m)		1.49 ± 0.15	1.22 - 1.74
Arm span (m)		1.63 ± 0.10	1.50 - 1.88
Arm/height ratio		1.11 ± 0.10	0.96 - 1.41
BMI [1] (kg/m^2)		24.31 ± 7.56	12.09 - 40.81
Waist circumference (cm)		96.49 ± 19.61	69.0 - 140.6
Triceps skinfold (mm)		4.21 ± 2.97	1 - 12
Hand grip strength- right hand (kg)		28.72 ± 7.86	12 - 41
Hand grip strength- left hand (kg)		27.31 ± 8.63	6 - 42
PASIPD [2](MET-hour/day)		14.79 ± 11.05	1.54 - 54.81
10 MWT [3] (sec)	All participants	10.56 ± 4.34	4.66 - 27.63
	Manual wheelchair user [3]	9.67 ± 2.89	4.66 - 16.06
10 MWT speed [4] (m/sec)	All participants	1.06 ± 0.36	0.36 - 2.15
	Manual wheelchair user [4]	1.13 ± 0.35	0.62 - 2.15

[1]: BMI: body mass index calculated based on arm span
[2]: PASIPD, Physical Activity Scale for Individuals with Disabilities; MET: metabolic equivalent
[3]: Ten-meter wheel test (10MWT), n= 24
[4]: The average speed of 10MWT, n= 24

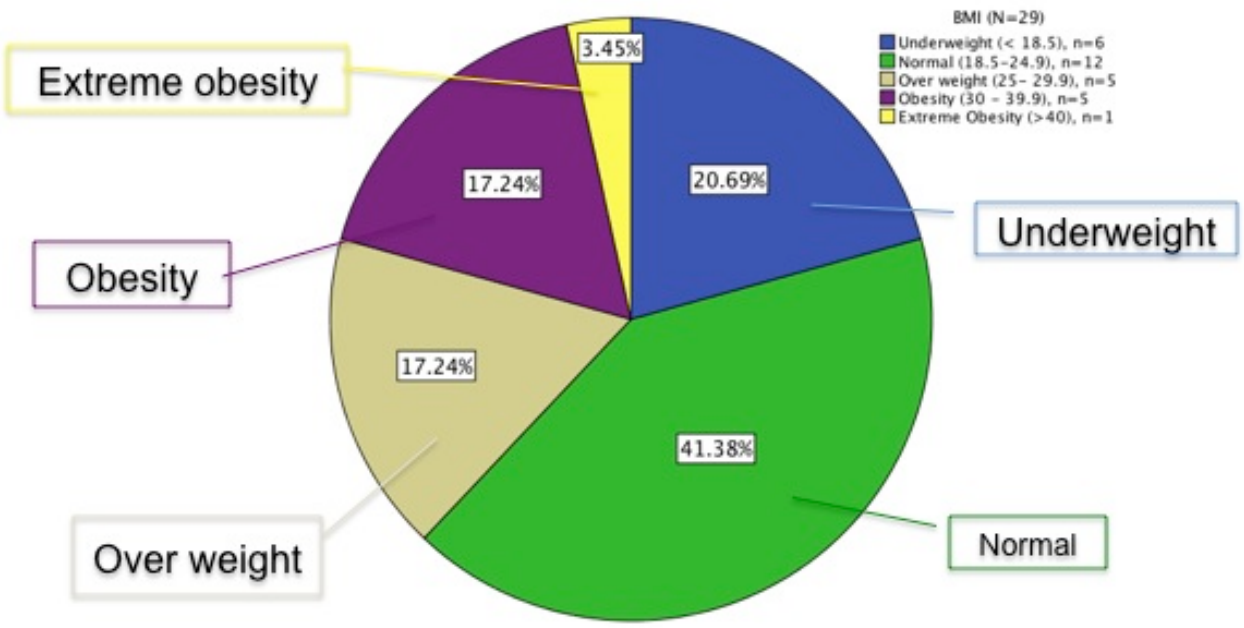

Figure 6. Classification of overweight and obesity by BMI

Table 3. Classification of overweight and obesity by BMI, waist circumference, and associated disease risks

	BMI	Obesity Class	Disease risk relative to normal weight and waist circumference	
			Men 102 cm or less Women 88 cm or less	Men > 102 cm Women > 88 cm
Underweight	< 18.5	-	5	1
Normal	18.5 - 24.9	-	10	2
Overweight	25.0 - 29.9	-	0	5 (high risk)
Obesity	30.0 - 34.9	I	0	2 (very high risk)
	35.0 - 39.9	II	0	3 (very high risk)
Extreme Obesity	40.0 +	III	1 (extremely high risk)	0
Classification of overweight and obesity by BMI, waist circumference, and associated disease risks. BMI was based on arm span. Disease risk for type 2 diabetes, hypertension, and CVD *Refer from the National Heart, Lung, and Blood Institute (National Institutes of Health, 2014)				

The medical records of all participants are listed in Table 4. The functional level of lesion was categorized into 2 groups, which are non-thoracic (n = 19, 65.5%) and thoracic lesion level (n = 10, 34.5%). The level of lesion was further coded at each lesion level in reverse order as an ordinal variable for regression analysis. For example, T7 was coded as twelve, T8 was coded as eleven, T9 was coded as ten, and the same coding rule for the following levels to normal, which was coded as 0. The Cobb angle of scoliosis was obtained from the electronic medical system and categorized into 2 groups as follows: 1) scoliosis less than 30 degrees or medical record note that stated the person had mild scoliosis (n = 25, 86.2%), and 2) scoliosis greater than 30 degrees or medical record note that stated the person had moderate or severe scoliosis (n = 4, 13.8%). Nine participants (31%) had spinal fusion surgery. Twelve participants (41.4%) had Chiari II malformation and among these participants, four of them (33.3%) had surgery. Out of 29 participants, five of them (17.2%) had sleep apnea. The records of lung disease history that were collected include pneumonia, bronchitis, pleural effusion, and lung hypo-inflation. Approximately 50% of participants had a history of such lung conditions in the medical records. These conditions primarily occurred following surgical procedures.

Table 4. Medical records of all participants

n = 29					
Variables			n		Percentage (%)
Functional level of lesion	Thoracic level	T7	1	10	34.5
		T8	1		
		T10	2		
		T11	1		
		T12	5		
	Non-thoracic level	L1	10	19	65.5
		L2	6		
		L3	1		
		L5	1		
		Normal	1		

Table 4. (Continued)

Scoliosis	Cobb angle less than 30 degrees	25	86.2
	Cobb angel equal or greater than 30 degrees	4	13.8
Spinal fusion	No spinal fusion	20	69
	Spinal fusion surgery	9	31
Chiari II malformation	No	17	58.6
	Yes	12	41.4
Chiari II malformation surgery	No	25	86.2
	Yes	4	13.8
Sleep apnea	No	24	82.8
	Yes	5	17.2
History of lung condition [1]	No	15	51.7
	Yes	14	48.3
[1]: History of lung condition includes pneumonia, bronchitis, pleural effusion, and lung hypo-inflation.			

2.3.2 Pulmonary function test results

A pulmonologist classified the participants based on the criteria of determining lung condition. One person (3.4%) had obstructed lung condition, with FEV_1 lower than 80% and FEV_1/FVC ratio lower than 70%. Fourteen people (48.3%) had restricted lung condition based on the FEV1/FVC ratio greater than 70% with FVC lower than 80%. Four participants (13.8%) had borderline restricted lung condition with FVC lower than 80% or TLC lower than 80% and FEV_1/FVC ratio greater than 70%. There was one participant whose FEV1/FVC ratio was 67%, FVC was 75%, and TLC was 72% that was classified as borderline for obstruction and restriction, so the pulmonologist determined this case as both restricted and obstructed (Figure 7). For analysis purposes, the lung condition categories were collapsed into three groups: 1). Restricted group (n = 19) that included people with restricted lung condition, borderline restricted lung condition, and both restricted and obstructed lung condition), 2). Non-restricted

group (n = 9), and 3). Obstructed group (n = 1). The results of PFTs for each group are shown in Table 5.

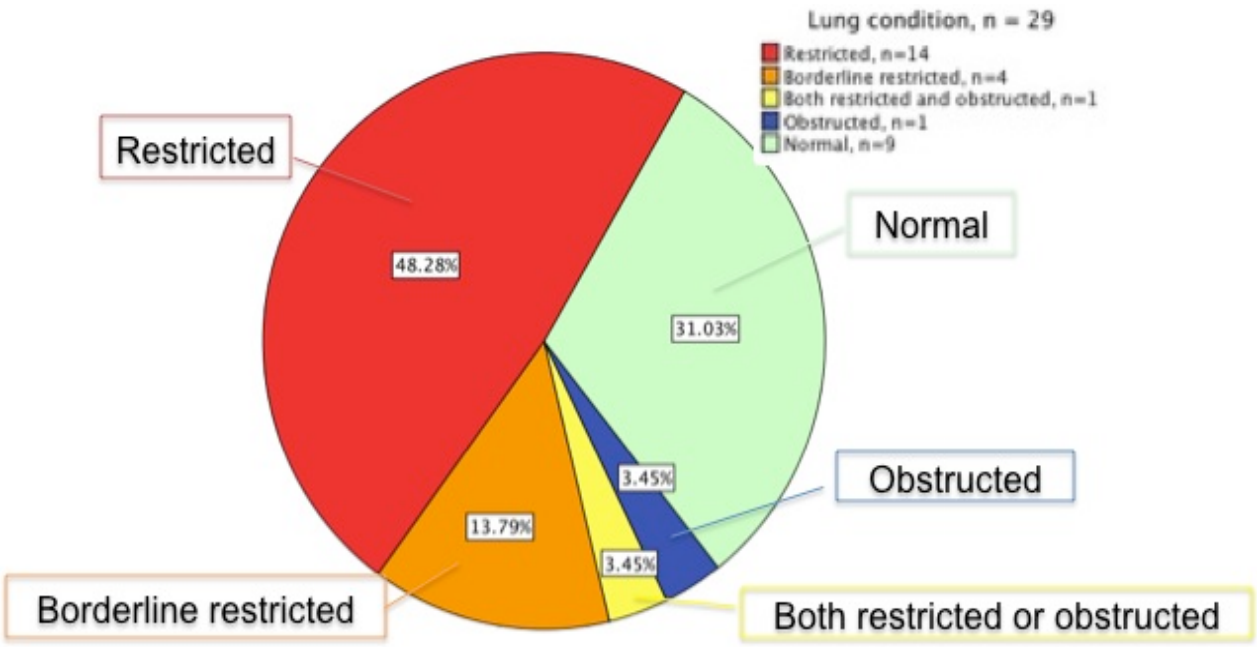

Figure 7. The pie chart of lung conditions for all participants

Table 5. Pulmonary function tests results of all participants

Variables	All participants (n = 29)		Type of lung condition				
			Restricted n = 19 (65.52%)		Non-restricted n = 9 (31.03%)		Obstructed n = 1 (3.45%)
Spirometry	**Mean ± SD**	**Range**	**Mean ± SD**	**Range**	**Mean ± SD**	**Range**	**Value**
FVC (%)	73.66 ± 28.60	20 - 127	57.74 ± 19.8	20 - 89	101.33 ± 12.7	84 - 125	127
FEV_1 (%)	73.76 ± 29.18	23 - 126	57.42 ± 20.19	23 - 100	103.11 ± 13.89	82 -126	120
FEV_1/FVC ratio (%)	86.07 ± 9.32	67 -100	86.16 ± 10.17	67 - 100	87.78 ± 5.63	80 - 94	69
FEF 25%-75% (%)	75.95 ± 31.81	23 - 136	60.53 ± 26.27	23 - 119	105.20 ± 17.73	80 - 136	103
PEF (liter/sec)	5.30 ± 2.45	1.82 - 10.51	4.31 ± 1.87	1.82 - 8.04	7.19 ± 2.53	3.51 - 10.51	7.13
PIF (liter/sec)	2.12 ± 1.05	0.64 - 4.59	1.9 ± 0.9	0.64 - 4.13	2.38 ± 1.21	0.77 - 4.59	3.76
MVV (Liter/minute)	83.28 ± 34.76	25.9 - 157.85	65.49 ± 23.54	25.9 - 109.55	119.39 ± 27.24	91 - 157.85	96.25
Lung volumes test	**Mean ± SD**	**Range**	**Mean ± SD**	**Range**	**Mean ± SD**	**Range**	**Value**
FRC (%)	76.52 ± 28.82	40 - 149	65.84 ± 17.87	40 - 116	95 ± 36.98	54 - 149	113
IC (%)	81.41 ± 31.03	26 - 128	64.32 ± 23.45	26 - 97	112.33 ± 9.57	97 - 126	128
TLC (%)	78.31 ± 24.4	35 - 126	64.16 ± 14.75	35 - 87	104.78 ± 14.59	85 - 126	109
ERV (%)	42.34 ± 33.62	2 - 136	32.68 ± 31.76	2 - 136	60.22 ± 32.29	16 - 121	65
RV (%)	92.93 ± 29.27	26 - 138	87.47 ± 26.4	26 - 134	104.22 ± 34.86	48 - 138	95
RV/TLC ratio (%)	32.83 ± 11.63	12 - 63	37.32 ± 10.76	14 - 63	23.33 ± 7.98	12 - 39	33

* FVC: forced vital capacity; FEV1: forced expiratory volume in one second; FEV_1/FVC: the ratio of forced expiratory volume in one second over forced vital capacity; FEF 25%-75%: maximum expiratory flow over the middle 50% of the vital capacity; PEF: peak expiratory flow; PIF: peak inspiratory flow; MVV: maximal voluntary ventilation, calculated from FEV_1 x 35; FRC: functional residual capacity; IC: inspiratory capacity; TLC: total lung capacity; ERV: expiratory reserve volume; RV: residual volume; RV/TLC: the ratio of residual volume over total lung capacity

Further analysis was conducted to compare the PFTs results between the restricted lung condition group and non-restricted lung condition group by using an independent *t* test. All PFTs were lower in the restricted group compared with non-restricted group. Significant differences were found in FVC, FEV1, FEF25-75%, FRC, IC, TLC, ERV, and RV/TLC between these two groups (Table 6).

Table 6. The independent *t* test result of PFTs between restricted and non-restricted group

PFTs variables	Restricted group (n=19) versus non-restricted group (n=9)*	
	t (degree of freedom)	p-value
FVC (%)	6.013 (26)	0.0001
FEV_1 (%)	6.109 (26)	0.0001
FEF 25% - 75%	4.585 (26)	0.0001
FRC (%)	2.844 (26)	0.009
IC (%)	5.870 (26)	0.0001
TLC (%)	6.829 (26)	0.0001
ERV (%)	2.132 (26)	0.043
RV/TLC ratio	-3.461 (26)	0.002
*Total n=28 due to one participants met criteria of obstructed lung condition, which was not included in this analysis; FVC: forced vital capacity; FEV1: forced expiratory volume in one second; FEF 25%-75%: maximum expiratory flow over the middle 50% of the vital capacity; FRC: functional residual capacity; IC: inspiratory capacity; TLC: total lung capacity; ERV: expiratory reserve volume; RV: residual volume; RV/TLC: the ratio of residual volume over total lung capacity		

2.3.3 The relationship between body composition measurements, physical activity, and PFTs results

An independent *t* test was used to examine whether there was significant difference of BMI, waist circumference, and physical activity between restricted (n = 19) and non-restricted (n = 9) groups. The restricted group had lower BMI values than the non-restricted groups (mean ± SD = 23.34 ± 7.23, 26.88 ± 8.38 kg/m^2, respectively), and PASIPD (mean ± SD = 17.06 ± 16.59, 13.91 ± 7.98 MET-hour/day, respectively). The restricted group had higher waist circumference (mean

± SD = 98.58 ± 20.13, 93.87 ± 19.60 cm, respectively) than the non-restricted group. There was no significant difference between these two groups in BMI ($t(26) = 1.153$, $p > 0.05$), PASIPD score ($t(26) = 0.686$, $p > 0.05$), and waist circumference ($t(26) = -0.583$, $p > 0.05$). The difference in the 10MWT scores of manual wheelchair users who were in the restricted and non-restricted groups was analyzed. Wheelchair users that in the restricted group (n = 18, mean ± SD = 10.11 ± 2.95 seconds) were slower than the non-restricted group (n = 5, mean ± SD = 8.57 ± 2.66 seconds) but there was no significant difference between these two groups ($t(21) = -1.05$, $p > 0.05$).

Correlation analysis was first used to examine the relationship between BMI, waist circumference, PASIPD, 10MWT, level of lesion, scoliosis, and PFTs results. All dependent variables used in regression analysis were normally distributed confirmed by Kolmogorov-Smirnov test. Predictors that could easily be gathered in the clinical setting, such as BMI, level of lesion, and spinal fusion were used as predictors in the backward regression analyses to predict FVC, FEV_1, TLC, and RV. Three significant models were found (Table 7). For model 1, the level of lesion and spinal fusion were significant predictors of FVC ($R^2 = 0.495$, $p < 0.0001$). The formula for model 1 was: FVC = 125.715 + (-7.881 x level of lesion) + (-10.131 x spinal fusion), showing that participants with higher level of lesion and those who had spinal fusion had lower FVC. For model 2, the level of lesion and spinal fusion were significant predictors of FEV_1 ($R^2 = 0.470$, $p < 0.0001$). The formula for model 2 was: $FEV_1 = 124.493 + (-7.538 \times \text{level of lesion}) + (-12.719 \times \text{spinal fusion})$, showing that participants with higher level of lesion and those who had spinal fusion had lower FEV_1. For model 3, the level of lesion and spinal fusion were significant predictors of TLC ($R^2 = 0.528$, $p < 0.0001$). The formula for model 3 was: TLC

= 123.225 + (-6.665 x level of lesion) + (-11.421 x spinal fusion), showing that participants with higher level of lesion and who had spinal fusion had lower TLC.

Table 7. Linear regression models of PFTs results

Model	DVs	Predictor	Unstandardized Coefficients B (SE)	t	p-value
Model 1					
$R^2 = 0.495$	FVC (%)	(Constant)	125.715 (11.444)	10.986	0.0001
$p < 0.0001$		Level of lesion	-7.881 (1.813)	-4.346	0.0001
$F (2,26) = 12.740$		Spinal fusion	-10.131 (8,877)	-1.141	0.264
Model 2					
$R^2 = 0.470$	FEV_1 (%)	(Constant)	124.493 (11.962)	10.408	0.0001
$p < 0.0001$		Level of lesion	-7.538 (1.895)	-3.977	0.0001
$F (2,26) = 11.529$		Spinal fusion	-12.719 (9.279)	-1.371	0.182
Model 3					
$R^2 = 0.528$	TLC (%)	(Constant)	123.225 (9.434)	13.062	0.0001
$p < 0.0001$		Level of lesion	-6.665 (1.495)	-4.459	0.0001
$F (2,26) = 14.568$		Spinal fusion	-11.421 (7.318)	-1.561	0.131

*DVs: dependent variables; SE: standardized error; FVC: forced vital capacity; FEV1: forced expiratory volume in one second; TLC: total lung capacity

Backward logistic regression was also employed to explore predictors of lung conditions (restricted lung condition versus non-restricted lung condition) (Table 8). BMI and spinal fusion were dropped from the model and functional level of lesion remained a significant predictor ($p = 0.045$, Nagelkerke $R^2 = 0.667$), showing that participants with higher level of lesion had an odds of 23.6:1 for having restricted lung condition.

Table 8. Binary logistic regression of restricted lung condition versus non-restricted lung condition

	B (SE)	Wald	Odds ratio	95% CI	
				Lower	Upper
BMI	-0.244 (0.147)	2.741	0.748	0.587	1.046
Spinal fusion	3.769 (2.085)	3.266	43.325	0.727	2580.620
Level of lesion	3.161 (1.579)	4.006*	23.603	1.068	521.656

* $p < 0.05$; B: Co-efficient; SE: Standard Error; CI: Confidential interval; Wald: Wald statistic

2.4 DISCUSSION

Hypothesis 1 was supported by the results showing that 65% of participants (n=19) met the criteria of restricted lung condition. Hypothesis 2 was also supported by the results showing that restricted lung condition group had lower PFTs than non-restricted lung condition group. Our results were slightly higher than previous studies. Seven subjects (58%) with myelomeningocele were reported to have restrictive lung disease and generally lower lung function than a controlled group (Sherman et al., 1997). Restrictive pattern of lung function was also found in patients with myelomeningocele but the prevalence was not reported (Patel et al., 2011). The higher prevalence of restricted lung condition among our participants may be due to mobility and age. More than 75% of participants in this study used manual wheelchairs or power wheelchairs as their primary mobility equipment, whereas none of the participants were wheelchair users in Sherman's study. Another explanation for higher rates of restricted lung conditions in out study could be due to the age of our participants. Changes that occur to the lungs with age include reduced lung volumes and alteration in trunk muscles that aid respiration. In this study, the range of participant's age was seventeen to seventy-one years whereas the age range in Sherman's study was ten to seventeen years. In the present study, two participants were age seventeen and neither met the criteria of restricted lung condition. Among participants whose were older than eighteen, more than seventy percent met the criteria of restricted lung condition. The level of lesion also plays a role in pulmonary function. In a study of twenty participants with spinal cord injuries with injury levels ranging from C5 to T11, people with lower thoracic injuries tended to have higher PFTs values than people with cervical and higher thoracic injuries (Battikha et al., 214). In the present study, participants' level of lesions ranging from T7 to S1 and were stratified

into the categories of thoracic lesion level and non-thoracic lesion level. Between these two groups, the thoracic level of lesion group had higher prevalence of restricted lung condition (90%) than the non-thoracic group (52.6%). Given that mobility, age, and level of lesion were correlated with restricted lung condition, future studies are needed to establish a central database to enhance the pulmonary function status of people with spina bifida.

Knowing that restricted lung condition is prevalent in this population, further analyses were conducted to find significant predictive models for PFTs results. Hypothesis 3, hypothesis 4, and hypothesis 5 were supported by the results by the finding that the level of lesion and spinal fusion were significant predictors for FVC, FEV_1, and TLC. Body mass index (BMI) was not a significant predictor for PFTs. In this study, arm span was used to calculate BMI based on a previous study that showed that the standard height would result in underestimation of the severity of lung disease (Sherman et al., 1997). Previous study showed that obesity has been shown as a risk factor of impaired lung function (Ciprandi et al., 2014), but this was not supported by the results of this study. Many different methods have been used to measure BMI and other than used the methods that we chose of using arm span. Some studies used a correction factor but we did not, it is not clear from the literature whether BMI using arm span is the most accurate measure. Thus, future studies should investigate whether BMI based on arm span is an accurate measure of body composition for the spina bifida population by comparing BMI based on arm span to the gold standard of Dual-energy X-ray absorptionmetry (DEXA). On the other hand, the finding of an inverse correlation between level of lesion and pulmonary function tests results was consistent with previous studies (Battikha et al., 2014; Cooper et al., 1993). The Cobb angle of scoliosis is determined by an x-ray and some participants did not have x-ray available. Some individuals had severe scoliosis that the radiologist reported was not measurable

using the Cobb angle. Consequently, the Cobb angles were not available on all participants. Whether a person had spinal fusion surgery or not was available in each person's medical record, however, this measure still does not provide a complete understanding of the role of scoliosis. Future studies should obtain x-ray films with Cobb angle determined by one radiologist for greater accuracy. We also explored factors that we thought might be predictors for restrictive or non-restrictive lung conditions. In our secondary analysis, we found that the odds of having pulmonary function values that indicate pulmonary restriction was 23.6 times higher in those who had spina bifida with higher lesion levels. Since it is more likely that people with spina bifida with thoracic level lesions are at risk for pulmonary restriction, then in the future it may become a more common practice for physicians to refer people with higher lesions for baseline pulmonary function testing. Finally, it is important to note that the only predictive equations that were available for us to use were those that were designed for able-bodied people; therefore, our results may be over or underestimating pulmonary restriction. A national study would be needed to collect pulmonary function measures in a large representative population with spina bifida of various heights, weights, ambulatory status, and other factors so that predictive equations could be developed for this population.

3.0 PHYSIOLOGICAL RESPONSES DURING GRADED EXERCISE STRESS TEST AMONG ADOLESCENTS AND ADULTS WITH SPINA BIFIDA

3.1 INTRODUCTION

Physical exercise requires the integration of the cardiovascular and pulmonary systems to respond to the increased respiratory demands of the contracting muscles. People with normal cardiovascular and lung functions rarely limit their exercise capacity due to a pulmonary disease. However, people who have pulmonary diseases were found to limit their exercise capacity (Truwit, 2003). Pulmonary restriction was prevalent in our cohort. In order to know how restrictive lung disease affects the exercise capacity of people with spina bifida, a graded exercise stress test on an arm ergometer was conducted. Cardiopulmonary exercise test (CPET) is a method of measuring the ability of the cardiovascular and respiratory systems to perform their functions through the measurement of gas exchange at the airway during maximal exercise (Wasserman et al., 2005). The CPET is usually performed on a stationary bicycle with a facemask, which is connected to a metabolic cart that measures gas exchange during exercise test. However, people with spina bifida have lower extremity functional limitation that would prohibit them from pedaling a stationary bicycle. Thus, the arm cycle ergometer was used for the graded exercise stress test.

Studies have shown that arm cycle exercise is less efficient than leg cycle exercise and has approximately 70% of maximal oxygen intake (VO_{2max}) compared to that of leg cycle exercise. In 2003, van den Berg-Emons conducted a study in the Netherlands that investigated body composition and physical activity among adolescents and young adults with myelomeningocele (n= 14, mean ± SD = 18 ± 4 years). The results showed that peak VO_2 was lower in the non-ambulatory group (mean ± SD = 22.5 ± 7.5 ml/kg/min) than the ambulatory group (mean ± SD = 30.1 ± 6.2 ml/kg/min); but no statistical significance between the two groups was observed. Buffart et al. (2008) also reported health-related physical fitness information among adolescents and young adults with myelomeningocele (n= 50, mean ± SD = 21.2 ± 4.5 years). The non-ambulatory group (mean ± SD = 19.2 ± 6.8 ml/kg/min) had a lower peak VO_2 than both the household ambulatory group (mean ± SD = 22.3 ± 6.6 ml/kg/min) and the community ambulatory group (mean ± SD = 29.0 ± 7.7 ml/kg/min). However, these previous studies did not compare the peak VO_2 between subjects who used arm cycle ergometer and who used leg cycle ergometer. Sherman et al. conducted a study in children with myelomeningocele (n= 12, mean ± SD = 13.1 ± 2.7 years) in 1997. They used an arm ergometer for exercise testing and found that subjects had significantly lower peak VO_2 (mean ± SD = 13.8 ± 4.8 ml/kg/min) than a healthy age-matched group (mean ± SD = 21.3 ± 7.5 ml/kg/min) ($p < 0.02$). In this study, they also found that 58% of participants (7 out of twelve participants) had restrictive ventilatory impairment on the basis of reduced TLC and normal FEV_1/FVC ratio and one had pulmonary limitation to exercise.

Limited studies have assessed pulmonary limitations during a graded exercise stress test among people with spina bifida. The first section of this present study was to investigate pulmonary function among adolescents and adults with spina bifida, which was present in

chapter two; the second section examines the exercise capacity in this population and in order to evaluate the relationship between pulmonary function and exercise capacity in this population.

3.1.1 Aims and hypotheses

Aims:

1. To investigate the exercise capacity of adolescents and adults with spina bifida.
2. To examine the relationship among body composition, physical activity, pulmonary function, and exercise capacity of adolescents and adults with spina bifida.

Hypotheses:

1. More than 60% of participants will have a pulmonary limitation to exercise.
2. Body mass index (BMI), level of lesion, and spinal fusion will be the most significant predictors of peak oxygen consumption (peak VO_2).
3. The Ten Meter Walk Test will be significantly correlated with peak VO_2.

3.2 METHODS

3.2.1 Recruitment procedures

This section was part of a large research project that investigated the pulmonary function among adolescents and adults with spina bifida. Please refer to 2.2.1.

3.2.2 Experimental protocol

This section was part of a large research project that investigated the pulmonary function among adolescents and adults with spina bifida. Please refer to 2.2.2.

3.2.2.1 Demographic and baseline measurements

This section was part of a large research project that investigated the pulmonary function among adolescents and adults with spina bifida. Please refer to 2.2.2.1.

3.2.2.2 Graded exercise stress test

Participants performed a symptom limited arm exercise stress test in an exercise test laboratory using an electronically braked arm ergometer (Saratoga Silver I Rand-Scot Inc., Colorado, U.S.A.). The graded exercise stress test was administered by an exercise physiologist and a physical therapist. All participants performed exercise stress test in a sitting position either with their own manual chairs, power wheelchairs or a standard office chair with armrest (Figure 8). The temperature if the room for the tests was 21.49 ± 1.60 degrees Celsius and relative humidity was 25.25 ± 2.95. The crank of the arm ergometer was situated at each participant's shoulder height in the beginning of test. A ramp and a platform were used for adjusting the seat-to-floor height when needed. Back support was added by using dense foams for participants who wanted to sit upright or get closer to the arm ergometer. A set of blocks was placed behind the front and rear wheels to stabilize the manual wheelchair while participants performed the exercise.

Figure 8. Setting of graded arm ergometry exercise stress test

A latex-free mask (Hans-Rudolph Inc., Kansas, U.S.A.) and a Polar heart rate monitor band (Polar Electro, Kemple, Finland) were placed on the participant's face and xiphoid process respectively. The mask was connected to a metabolic cart (Moxus Modular VO2 System, AEI Technologies, Inc., Pennsylvania, U.S.A.) that was calibrated before each test and recorded the participant's ventilation (VE) (expressed in L/min), breathing rate (expressed in breath/min), peak heart rate (peak HR) (expressed in beat/min, or bpm), peak oxygen consumption (peak VO_2) (expressed in L/min and ml/kg/min) and respiratory exchange ratio (RER). After a minute warm up at 10 Watts (W), the exercise test began and participants pedaled at 70 revolutions per minute (RPM) throughout the test while the work rate increased by 10 W each minute until the participant was exhausted. A metronome and verbal encouragement were provided to help participants maintain the 70-RPM pace throughout the test. The participants were then asked to state their level of perceived exertion on the Borg Scale and the WHEEL Scale during the last 15

seconds of each minute at the end of each workload until the end of the test. The graded exercise stress test concluded when the participant asked to stop or felt they could not go any further. The maximal workload (Watt) that each participant completed, the rating of perceived exertion at each phase, and the self-reported reason for stopping exercise were collected in the exercise chart.

3.2.3 Data collection and data analysis

Socio-demographic information, body composition measurements, pulmonary function tests results, and graded maximal exercise test results were gathered during a one-time visit. The percentage of predicted values used for analyses were adjusted for age, height, weight, and gender. Arm span was used as a proxy for height. Ventilation (VE), breathing rate, peak heart rate (peak HR), peak oxygen consumption (peak VO_2), and respiratory exchange ratio (RER) were recorded from the metabolic cart and presented as mean ± SD in the results section. The RER is generally considered an indication of achievement of maximal exercise. If the value is less than 1, this would reflect submaximal cardiovascular effort or a pulmonary limitation to exercise (Balady et al., 2010). In this study, RER < 1.00 was used to determine that the participants did not achieved the maximal exercise test. A pie chart was used to present the distribution of lung conditions in participants who had RER < 1 group and participants who had RER ≥1 groups. Also, an independent *t* test was used to analyze the differences between restricted and non-restricted groups that was determined from previous PFTs results.

To study the relationship among body composition, physical activity, PFTs results, and peak oxygen consumption, multiple linear regression analyses was used. Statistical analyses

were performed using IMB SPSS version 22.0 for Windows (IBM Corporation, Armonk, NY); $p \leq 0.05$ were considered significant.

3.3 RESULTS

3.3.1 Demographic characteristics of all participants

This section was part of a large research project that investigated the pulmonary function among adolescents and adults with spina bifida. Please refer to 2.3.1 and table 1, table 2, and table 3.

3.3.2 Physiological responses during exercise test

The exercise capacity data including ventilation, breathing rate, peak heart rate, peak VO_2, and RER were measured and recorded by the metabolic analyzer. The value of peak heart rate percentage of predicted maximum (peak HR % of predicted) was adjusted for age, gender, and arm span substitute for the height. The results of the physiological responses during the exercise test are listed in Table 9. There was one missing data of the peak heart rate (HR) and peak HR % of predicted resulting from equipment error. The maximal workload that each participant completed was recorded in the chart and expressed as mean ± SD. The reasons for stopping the test were obtained from the participants at the end of the test. More than 70% of participants reported that muscle fatigue was the reason that kept them from pedaling at the end of the test. Among these participants, five (83.3%) of them met the criteria for restricted lung condition and one participant reported that breathing was the reason for stopping the test.

Table 9. Graded arm ergometry exercise stress test results of all participants

n = 29			
Variables		Mean ± SD	Range
Ventilation (liter/min)		43.41 ± 22.95	16.75 - 136.14
Breathing rate (breath/min)		48.55 ± 13.98	26 - 80
Peak heart rate[1] (beats/min)		150.75 ± 29.16	102 - 193
Peak hear rate percent of predicted [2]		78.93 ± 14.00	55 - 101
Peak VO_2 [3]	(liter/min)	1.26 ± 0.69	0.42 ± 3.48
	(milliliter/kg/min)	19.53 ± 8.92	7.3 - 48.4
Peak VO_2 percent of predicted (%)		59.33 ± 23.62	26- 137
Peak RER [4]		1.07 ± 0.19	0.76 - 1.62
Maximal workload (Watt) [5]		50 ± 31.75	20 - 190

[1]: Peak heart rate, n = 28. One missing data due to equipment error
[2]: Peak heart rate percent of predicted, n = 28. One missing data due to equipment error
[3]: Peak VO_2, the peak oxygen consumption during maximal exercise test
[4]: Peak RER: peak respiratory exchange ratio during maximal exercise test
[5]: Maximal workload: the highest and complete workload that each participant achieved during graded arm ergometry exercise stress test

3.3.3 The relationship between body composition, physical activity, pulmonary function results, and oxygen consumption

The peak oxygen consumption (peak VO_2, in L/min) was used as the dependent variable. Using the previous findings in pulmonary function tests, an independent *t* test was conducted to investigate whether there was significant difference between the restricted group and non-restricted group (Table 10). The results showed that the restricted group had lower peak VO_2 (mean ± SD = 1.13 ± 0.698 L/min) than the non-restricted group (mean ± SD = 1.53 ± 0.69 L/min) but there was no significant difference between these two groups ($t(26) = 1.447$, $p > 0.05$). The peak RER of the restricted group was lower (mean ± SD = 1.06 ± 0.21) than non-restricted group (mean ± SD = 1.11 ± 0.15), but there was no significant difference between these two groups ($t(26) = 0.694$, $p > 0.05$). Figure 9 and figure 10 show pie charts of lung condition for RER < 1 and RER ≥ 1 groups. Among the participants who did not reach the

maximal exercise test with RER < 1 (n = 10), 90% of them had restricted or borderline restricted lung condition. Moreover, in participants who had RER ≥ 1 group, more than 50% of them had a restricted lung condition.

Table 10. Oxygen consumption and respiratory exchange ratio for lung condition groups

Variable		Restricted group (n = 19)		Non-restricted group (n = 9)		Obstructed (n = 1)
		Mean ± SD	Range	Mean ± SD	Range	Value
Peak VO_2 [1]	(l/min)	1.13 ± 0.69	0.42 - 3.48	1.54 ± 0.69	0.97 - 3.12	1.07
	(millilter/kg/min)	18.04 ± 8.28	7.3 - 37.4	22.58 ± 10.42	12.3 - 48.4	20.4
Peak VO_2 percent of predicted (%)		54.87 ± 26.08	26 - 137	66.94 ± 16.61	45 - 100	75.6
Peak RER [2]		1.06 ± 0.21	0.76 - 1.62	1.11 ± 0.15	0.89 - 1.45	1.10

[1]: Peak VO_2, the peak oxygen consumption during maximal exercise test
[2]: Peak RER: peak respiratory exchange ratio during maximal exercise test

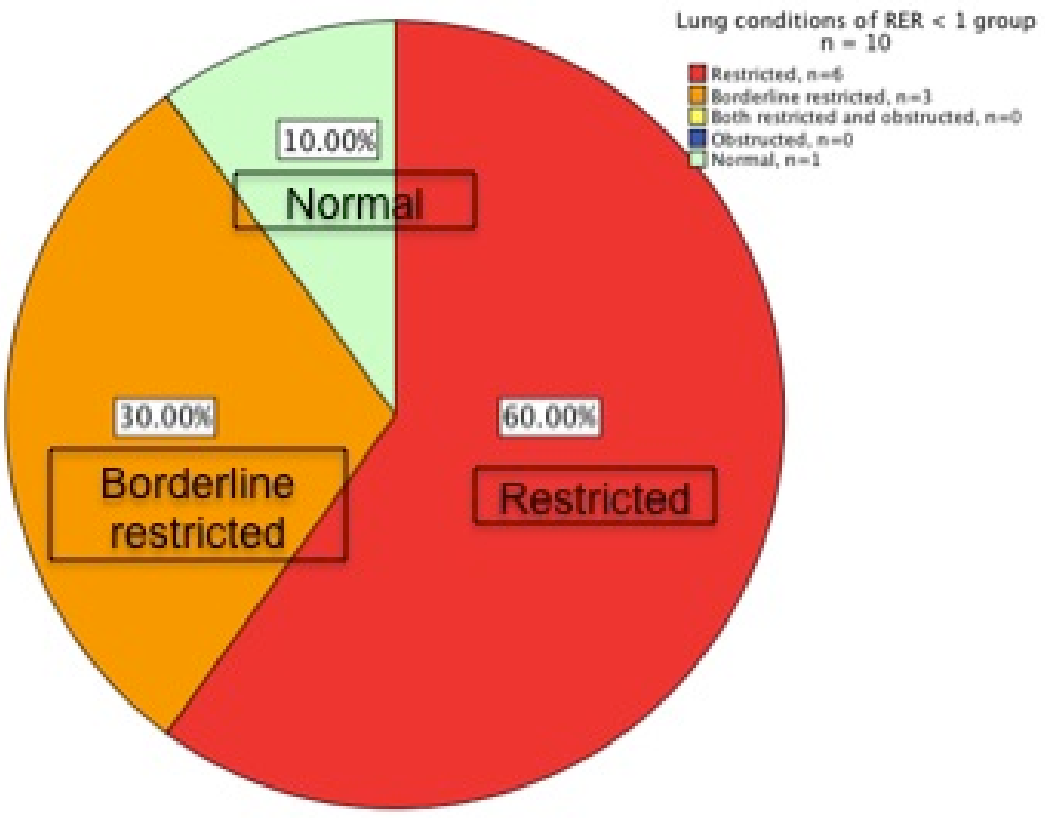

Figure 9. Lung conditions of participants whose RER < 1

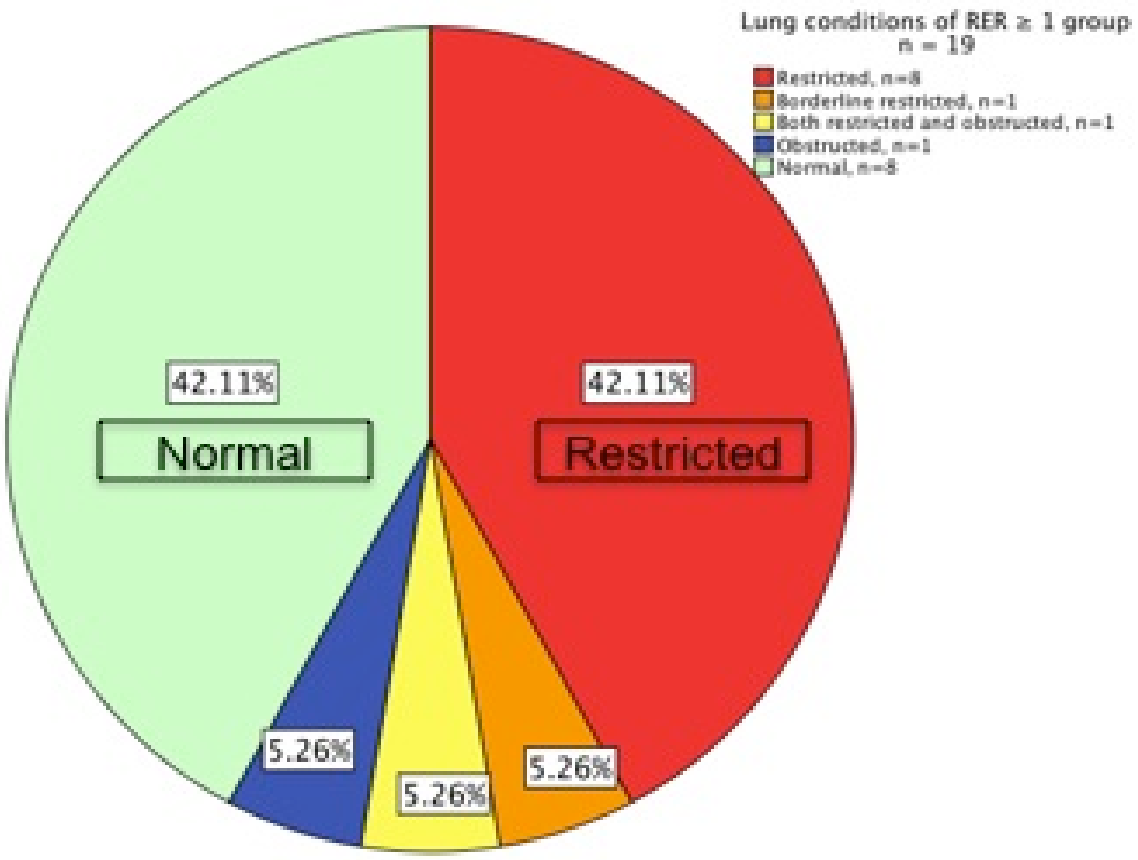

Figure 10. Lung conditions of participants whose RER ≥ 1

Given that gender was known as a predictor of peak VO_2 among people with spina bifida from a previous study (Buffart et al., 2008), hierarchical multiple regression was conducted to predict whether BMI, level of lesion, and spinal fusion were predictors of peak VO_2. After controlling for gender, the variables BMI, level of lesion, and spinal fusion explained more than 34.7% of the variance in peak VO_2 (Table 11). Gender, BMI, the functional level of lesion, and spinal fusion were significant predictors of peak VO_2 (R^2= 0.5, p = 0.02). The formula for the model was: peak VO_2 = 0.561 + (-0.822 x gender) + (0.056 x BMI) + (-0.048 x functional level of lesion) + (0.183 x spinal fusion). The relationship between 10MWT and peak VO_2 was also analyzed by regression (Table 12) and a significant correlation was found (R^2= 0.27, p = 0.08).

Table 11. Regression models for peak VO_2

Model	DVs	Predictor	Unstandardized Coefficients B (SE)	t	p-value
$R^2 = 0.153$ $p = 0.036$ $F (1,27) = 4.870$	Peak VO_2	(Constant)	1.529 (0.173)	8.865	0.0001
		Gender	- 0.529 (0.240)	-2.207	0.036
$R^2 = 0.500$ $p = 0.002$ $F (4,24) = 5.998$	Peak VO_2	(Constant)	0.561 (0.462)	1.213	0.237
		Gender	-0.822 (0.216)	-3.802	0.001
		BMI	0.056 (0.015)	3.806	0.001
		Level of lesion	-0.048 (0.045)	-1.066	0.297
		Spinal fusion	0.183 (0.239)	0.764	0.453
*DVs: dependent variables; SE: standardized error					

Table 12. The relationship between 10MWT and peak VO_2

Model	DVs	Predictor	Unstandardized Coefficients B (SE)	t	p-value
$R^2 = 0.277$ $p = 0.08$ $F (1,22) = 8.424$	Peak VO_2	(Constant)	2.592	5.454	0.0001
		10MWT	-0.137	-2.902	0.008
*DVs: dependent variables; SE: standardized error					

3.4 DISCUSSION

Hypothesis 1 required further analysis of variables gathered during the exercise test such as minute ventilation (VE) and maximal voluntary ventilation (MVV) to more specifically answer this question. However, descriptive data for this study showed that 34% of participants reached an RER < 1. The number of participants who reached an RER < 1 was ten (34.5%). It is worthy to note that participants who had RER ≥ 1 group, more than 50% of participants also met the criteria of restricted lung condition. This finding may suggest that RER may not be specific enough or may not be the only factor that is involved in determining whether pulmonary restriction played a role in exercise tolerance.

In the present study, the mean value of peak VO_2 for all participants was 19.53 ml/kg per minute during arm ergometry, which was lower than other studies of with adolescents and adults with spina bifida (22.6 ± 8.2 ml/kg per minute in Buffart et al. 2009 and 27.3 ± 7.4 ml/kg per minute in Van den Berg-Emons et al., 2003). Studies have shown that people with spina bifida tend to have an inactive lifestyle due to lower extremity functional limitation, increased body fat, and poor exercise capacity (Buffart et al., 2008, Dosa et al., 2009, Nelson et al., 2007, van den Berg-Emons et al., 2003) so finding a low peak VO_2 is not surprising. However, comparisons should be made with caution because peak VO_2 was measured either during leg cycle ergometry or arm ergometry depending on the persons mode of ambulation in the previous studies, and peak VO_2 from arm ergometry is thought to be 70% of that obtained from cycle ergometry. Also, previous studies enrolled participants with lower levels of lesion and the majority of participants were ambulatory, which would considerably affect peak VO_2. Thus, it may not be fair to compare the value of peak VO_2 percent of predicted (peak VO_2%) that was found in our study to other studies whose participants had higher functional status. But the value of peak VO_2 percent of predict (59.33%) still be informative and consistent with previous studies that demonstrated that people with spina bifida had lower exercise capacity. Future studies related to exercise testing should include a larger population of wheelchair users.

For hypothesis 2, BMI, level of lesion, and spinal fusion were able to predict peak VO_2 that was consistent with previous studies (Battikha et al., 2014, Buffart et al., 2008). In Buffart's study (2008), they used gender, ambulatory status, and muscle strength as predictors and explained 55% of peak VO_2. In the present study, the functional level of lesion, BMI, and spinal fusion were used in the second step of hierarchical regression and explained more 34.7% variances of peak VO_2. The level of lesion was based on motor level and provided more accurate

information in regards to functional status than the neurological level, however, due to small sample size this model may explained less variances of peak VO_2.

For hypothesis 3, the 10MWT was significantly correlated with peak VO_2, showing that participants who had lower 10MWT results (were faster) had higher peak VO_2. The value of 1.06 m/s is considered a safe velocity to cross an intersection (Cowan et al., 2008). Out of 19 participants who used manual wheelchair as primary mobility equipment, 13 participants (68.4%) participants had velocity greater than 1.06, meaning that their functional activity and independence, such as doing grocery shopping in community or going outside for occupational purpose, would be affected. Further investigations on propulsion force, pattern, and frequency in this population should be conducted in order to gain more information and clinical suggestions (i.e. prevention of upper-extremity injury or enhancement of functional activity) for people with spina bifida who used manual wheelchair as primary their mobility methods. Another physical activity that measured in the present study was the PASIPD score. Participants in this study reported PASIPD score (14.79 MET-hour/day versus 14.01 MET-hour/day) similar to previous study (Buffart's et al., 2008) but a lower score compared to Washburn's study (19.8 MET-hour/day). This finding is in line with a previous study (Buffart et al., 2008) that revealed that people with spina bifida tend to be hypo-active, but should be interpreted with caution since this study was conducted over the course of two-years and test scores might have been underestimated due to weather changes. In the present study, we also compared the socio-demographic data to a retrospective cohort study (Roach et al., 2011) and found that more participants in the present study lived independently (44.8% versus 30%) but fewer participants drove and had access to a car (27.5% versus 54%). Although these differences may due to

participants' living arrangements and support system, socio-demographic data presented in this study still provides valuable information.

3.4.1 Future studies

In the present study, restrictive lung conditions were found to be prevalent in adolescents and adults with spina bifida and this result was consistent with previous studies. The level of lesion and spinal fusion were factors that were found to be significant predictors of PFTs results. However, the record of spinal fusion was not representative of the Cobb angle of scoliosis. Future studies are needed to investigate the pulmonary function before and after spinal surgery. Finally, there are not predictive equations that can be applied to people with disabilities who are wheelchair users for arm ergometry exercise stress tests. Future studies are needed that test a large population to develop predictive equations for VO_{2max} for people with spina bifida.

3.4.2 Clinical significances

The results of this study showed that participants who had higher level of lesion had lower PFTs results and tend to have restricted lung condition and lower aerobic capacity. Also, the 10MWT was inversely correlated with aerobic capacity. Health care providers, such as physical therapist and occupational therapist, should consider that people with spina bifida with higher level of lesions might have pulmonary restriction and adjust exercise prescriptions accordingly and monitor vitals, including chest wall excursion and oxygen saturation. Also, with further research to determine a cutoff time that would indicate that a person is at risk for decreased ability to

complete activities of daily living, therapist could use the 10MWT an a tool when evaluating a client for a manual wheelchair.

4.0 VALIDITY OF THE WHEEL SCALE AMONG ADOLESCENTS AND ADULTS WITH SPINA BIFIDA

4.1 INTRODUCTION

Depending on the lesion level, spina bifida involves motor and sensory loss. Secondary conditions, such as cardiovascular disease, obesity, and metabolic syndrome were related to individual's mobility (Buffart et al., 2008). Previous studies also indicated that people with chronic conditions and obesity might develop a negative cycle of inactivity due to functional limitations of extremities and increase of body fat (Schwimmer et al., 2003). The hypo-activity life-style was also noticed among people with spina bifida in that their daily physical activity and aerobic fitness were low and body fat was high (Buffart et al., 2008b). Therefore, attention to physical fitness and adaptive exercise in health science has been increasing in recent decades.

Adaptive exercise devices, such as an arm ergometer, have been shown to improve aerobic fitness in people with spinal dysfunction (Hick et al., 2003, Widman et al., 2006). However, tools of knowing exercise intensity for clinician to understand subjective symptoms are limited. The use of perceived exertion scale (RPE) along with arm ergometry exercise testing has been suggested as a useful tool for regulating exercise intensity in people with spinal cord injury (Goosey-Tolfrey at al., 2010), however, there is no RPE scale developed or validated in wheelchair users. In this study, there is a concentrated focus on the spina bifida population,

specifically focused on people who have a diagnosis of myelomeningocele and whose primary mobility equipment is the wheelchair. Furthermore, the Borg Scale was used as a standard criterion for validating the newly developed WHEEL Scale.

4.1.1 Rating of perceived exertion scale

In order to know the perception of an individual's physical strain during exercise, a Swedish psychologist, Gunnar Borg, developed the first scale for measuring perceived exertion in the early 1960s. The theoretical rationale underlying the application of RPE is the functional interdependence of perceptual and physiological responses during exercise. His assumption was that the overall RPE integrates various information from body systems, including peripheral working muscles and joints, cardiovascular and respiratory functions, and nervous system. Then all the responses during exercise integrated into a configuration of perceived exertion (Borg, 1982, Robertson, 2004). Based on the assumption, in 2001, Robertson used an effort continua model of perceived exertion to explain the relationship between an individual's subjective responses during exercise, which involves three main effort continua: physiological, perceptual, and performance (Figure 11) (Robertson, 2004).

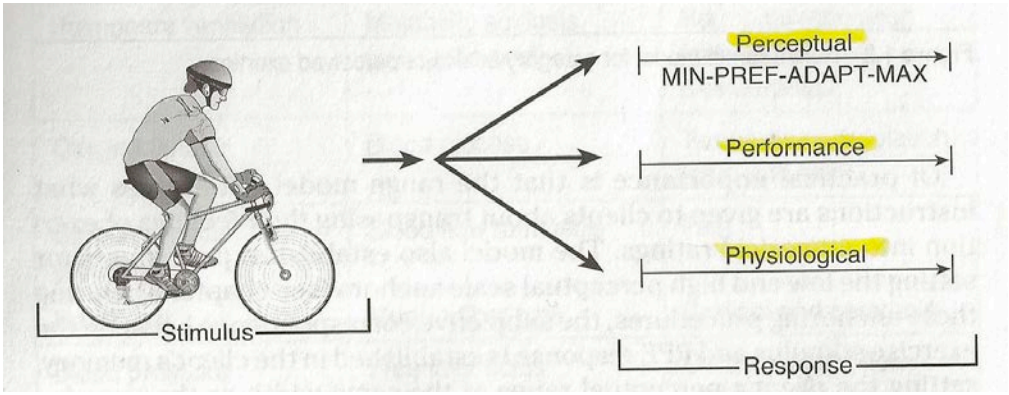

Figure 11. Effort continua model of perceived exertion (Robertson, 2004)

Robertson adopted this model from Borg (1998) and described that the physiological demands of exercise performance and the perceptual exertion associated with that performance. This indicated that a perceptual response provides the same information about the exercise performance as physiological response does. It can be said that both perceptual and physiological continua would change in response to increasing the exercise intensity increases. Therefore, prescribing or regulating exercise intensity of exercise can be based on the perceptual exertion and physiological response. The fundamental argument for measuring perceived exertion allowed scientists to establish a tool that had the same metric qualities as was used in physics and physiology. Then "psychophysical ratio-scaling methods" was introduced for measuring perceptual intensities. This method contained an absolute zero and the same distance between all values.

4.1.1.1 The Borg Scale

The Borg 6-20 Scale (Figure 12) is a tool for estimating an individual's physiological demand and regulating exercise intensity. It's a numeric scale with verbal descriptors ranging from "very, very light" corresponding with 7, to "very, very hard" corresponding with 19. The Borg 6 - 20 Scale was validated in able-bodied users under various exercise conditions, Some investigators adopted the scale for use in other population, including able-bodied adults (Dishman et al., 1994), and people with disabilities (Grange et al., 2002; Goosey-Tolfrey et al., 2010). Though the Borg Scale was adopted to use under various conditions of physical exertion, the verbal descriptors on the Borg Scale were developed for an adult population and may not be as useful for children or for individuals who have cognitive impairment (Robertson et al., 2005).

Borg Scale of Rating of Perceived Exertion (RPE)

6	
7	Very, very light
8	
9	Very light
10	
11	Fairly light
12	
13	Somewhat hard
14	
15	Hard
16	
17	Very hard
18	
19	Very, very hard
20	

Figure 12.The Borg 6 - 20 Scale

4.1.1.2 The OMNI Scale

Due to the lack of suitable verbal descriptors and pictorial format in the Borg 6-20 Scale, Robertson and colleagues validated the adult version of the OMNI Scale (2004) and developed the child version of the OMNI Scale (2000). The Child OMNI Scale contained easier language than in the Borg Scale. OMNI is an acronym for the word omnibus, which referred to a category scale having broadly generalizable measurement properties (Robertson, 2004). The Child OMNI Scale pairs a numerical response raging from 0 to 10 with drawings of a person exercising at different intensities and verbal descriptors (Figure 13) and has been validated (Robertson et al.,

2000, Robertson et al., 2004, Robertson et al., 2005, Utter et al., 2002). The advantage of the OMNI Scale is that those who are unable to read or understand the numeric scale can still choose a level of exertion based on the pictorial format. However, the OMNI Scales are depicted for ambulatory individuals or ambulatory individuals exercising on equipment.

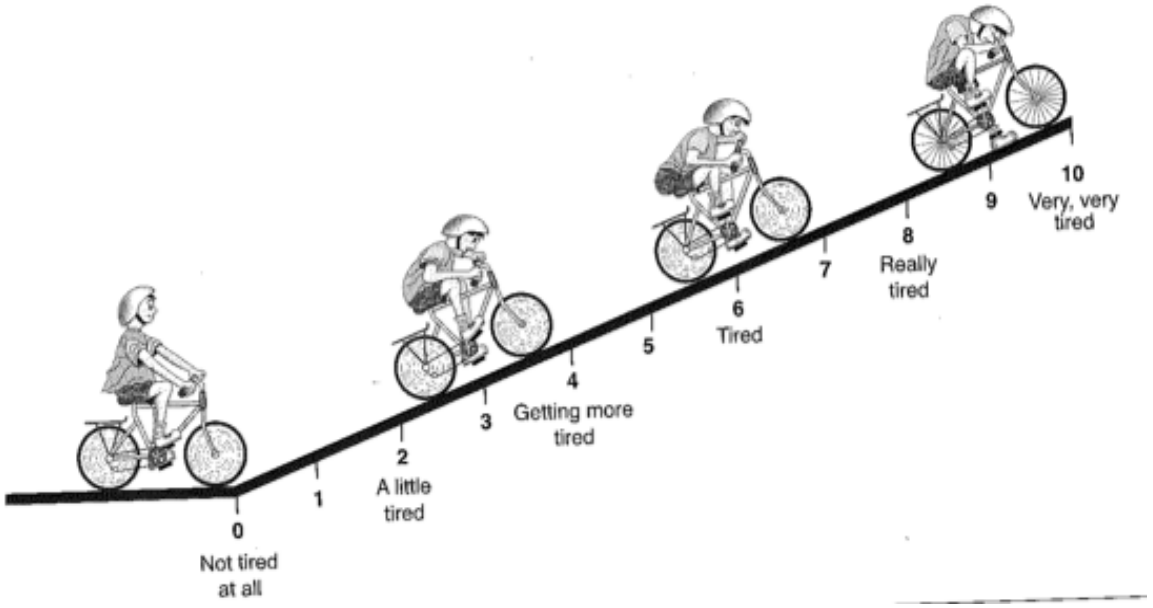

Figure 13.The Child OMNI Scale, cycle format (Robertson, 2004)

4.1.1.3 The WHEEL Scale

Manual wheelchair users may not be able to relate their feelings of exertion to these scales that were designed for people without disabilities. People with spina bifida who are life-long wheelchair users may have this issue. Also, their health care providers, such as physical therapists or exercise personnel trainers, may have issues when seeking for exercise intensity tool. In order to provide exercise prescription for people with spina bifida, the new rating of perceived exertion scale (the WHEEL Scale) (Figure 14) was adopted from the Child OMNI Scale and validated in this study.

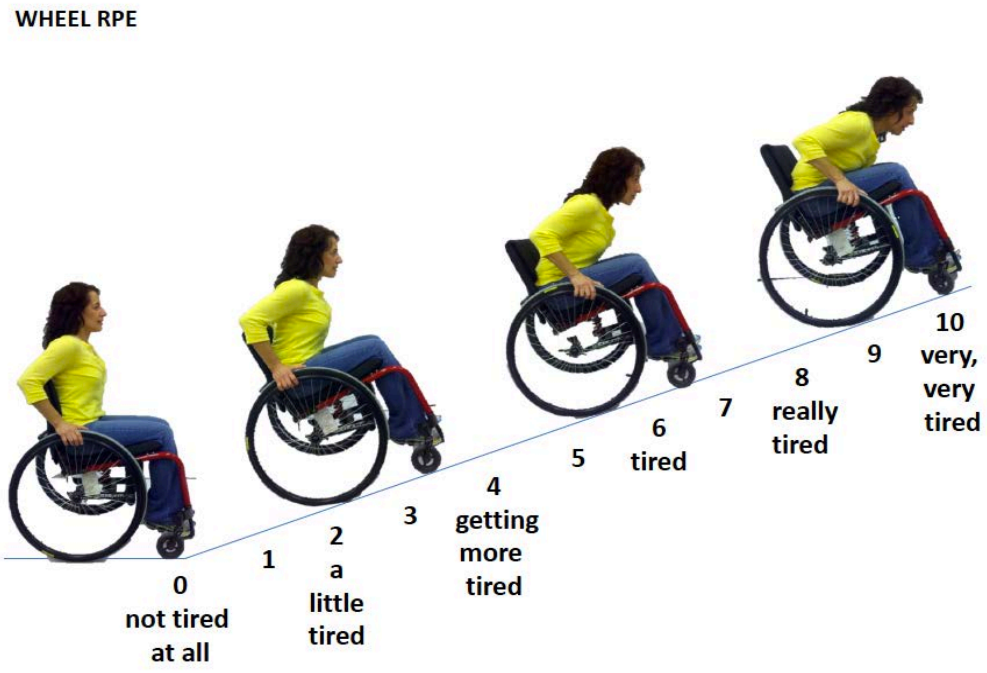

Figure 14. The WHEEL Scale

4.1.2 Aims and hypotheses

Aims:

1. To examine the concurrent validity of the newly developed WHEEL Scale.
2. To investigate the construct validity of the newly developed WHEEL scale.

Hypotheses:

1. A significant positive correlation will be seen between RPE derived from the Borg 6-20 scale and relative heart rate (HR) and relative oxygen consumption (VO_2).
2. A significant positive correlation will be seen between RPE derived from the WHEEL scale and relative HR and relative VO_2.

3. A significant positive correlation will be seen between RPE derived from the Borg scale and RPE from the WHEEL scale.

4.2 METHODS

4.2.1 Recruitment procedures

This study was part of a large research project that investigated the pulmonary function among adolescents and adults with spina bifida. Please refer to 2.2.1.

4.2.2 Experimental protocol

This study was the third part of a large research project and the data were derived from the graded exercise stress test, which was administered by an exercise physiologist and a physical therapist (Figure 15). Please refer to the detail testing procedure section 3.2.2.2. All participants were told about the scaling system in the Borg Scale and the WHEEL Scale. Then the participants were asked to state their level of perceived exertion on the Borg Scale and the WHEEL Scale sequentially each minute at the end of each workload until the end of the test.

Figure 15. Obtained the RPE Scales during the graded arm ergometry exercise stress test

4.2.3 Data collection and data analysis

Descriptive statistics for socio-demographic data and physiological data were presented as a mean ± SD for variables that were continuous and as a count (percentage) for variables that were categorical. Although the physiological variables (HR and VO_2) were collected breath by breath with the metabolic cart, we used values for every 20 seconds. The values were averaged at 20 seconds and 40 seconds of each exercise minute. The concurrent validity of the WHEEL Scale was determined using correlation analysis. For hypothesis 1 and hypothesis 2, Kendall's Tau correlation analysis was used to determine whether physiological variables (relative peak VO_2 and relative peak HR) increased linearly and positively as workload increased during the test. For hypothesis 3, a scatter plot of the WHEEL Scale versus the Borg Scale was constructed to

show the dispersion of the data points for each subject around the best-fit line. A linear regression (R^2) was also used to examine how strong the Borg Scale can be explained by the WHEEL Scale. The level of statistical significance was set at $p \leq 0.05$. Statistical analyses were performed using IMB SPSS version 22.0 for Windows (IBM Corporation, Armonk, NY).

4.3 RESULTS

4.3.1 Demographic characteristics of all participants

This section was part of a large research project that investigated the pulmonary function among adolescents and adults with spina bifida. Please refer to 2.2.2.1.

4.3.2 The relationship between Borg RPE, relative HR, and relative VO_2

The relationships between RPE obtained from the Borg Scale and relative heart rate (HR) and relative oxygen consumption (VO_2) are listed in Table 13. Figure 16 showed positive trends between the Borg RPE and relative HR and the Borg RPE relative VO_2 as workload increased.

Table 13. The correlation between Borg RPE, relative HR, and relative VO_2

n = 29				
Variable	Relative HR		Relative VO_2	
	r	*p*	*r*	*p*
Borg RPE	0.395	.0001**	0.415	.0001*
r: Kendall's tau_b correlation coefficient **: Correlation is significant at the 0.01 level (2-tailed)				

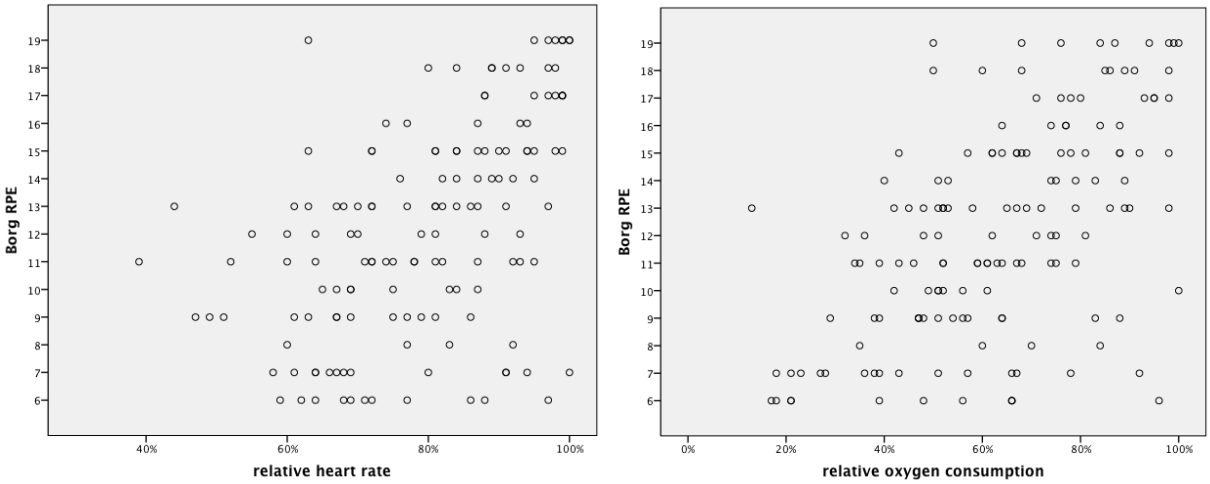

Figure 16. Scatter plots of Borg RPE, relative HR, and relative VO_2 at each workload for all participants

4.3.3 The relationship between WHEEL RPE, relative HR, and relative VO_2

The relationships between RPE obtained from the Borg Scale and relative heart rate (HR) and relative oxygen consumption (VO_2) are listed in Table 14. The RPE obtained from the Borg Scale during the exercise test was significant correlated with relative HR (r = 0.398, p < 0.0001) and relative VO_2(r = 0.403, p < 0.0001). Figure 17 showed positive trends between the Borg RPE and relative HR and the Borg RPE relative VO_2 as workload increased.

Table 14. The correlation between WHEEL RPE, relative HR, and relative VO_2

n = 29				
Variable	Relative HR		Relative VO_2	
	r	*P*	*r*	*p*
WHEEL RPE	0.398	.0001**	0.403	.0001**
r: Kendall's tau_b correlation coefficient **: Correlation is significant at the 0.01 level (2-tailed)				

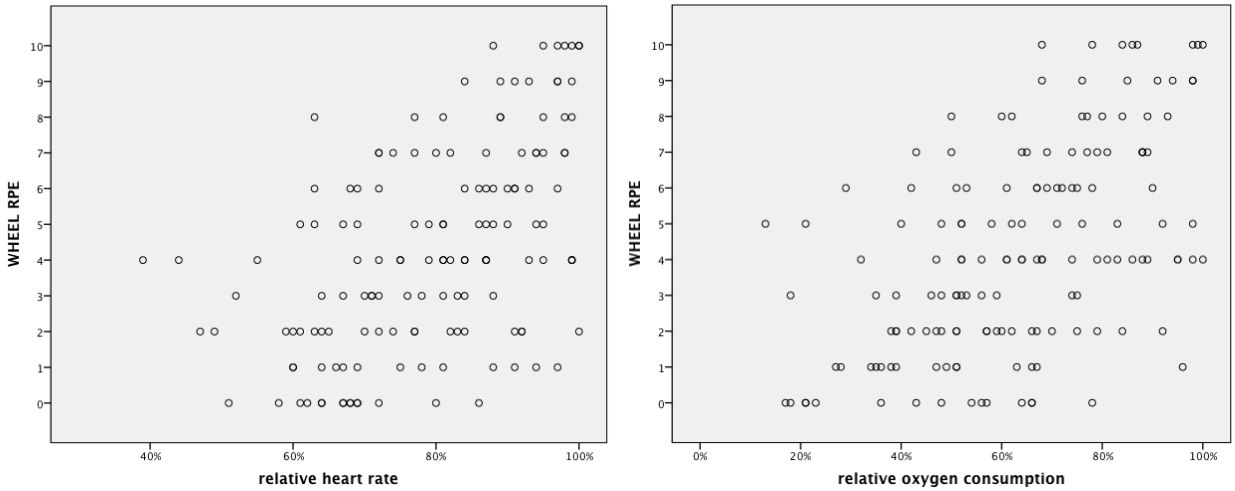

Figure 17. Scatter plots of WHEEL RPE, relative HR, and relative VO_2 at each workload for all participants

4.3.4 The relationship between RPEs obtained from the Borg Scale and the WEEEL scale

A scatter plot showed a significant correlation between the WHEEL Scale and the Borg Scale (Figure 18). Also, a linear regression was used to investigate the relationship between the Borg Scale and the WHEEL Scale (Table 15). The results showed that the WHEEL Scale significantly predicted the Borg Scale, $b = 0.868$, $t = 21.517$, $p < 0.0001$. The WHEEL Scale also explained a significant proportion of variance in the Borg Scale, $R^2 = 0.754$, $F(1, 151) = 463.013$, $p < 0.0001$.

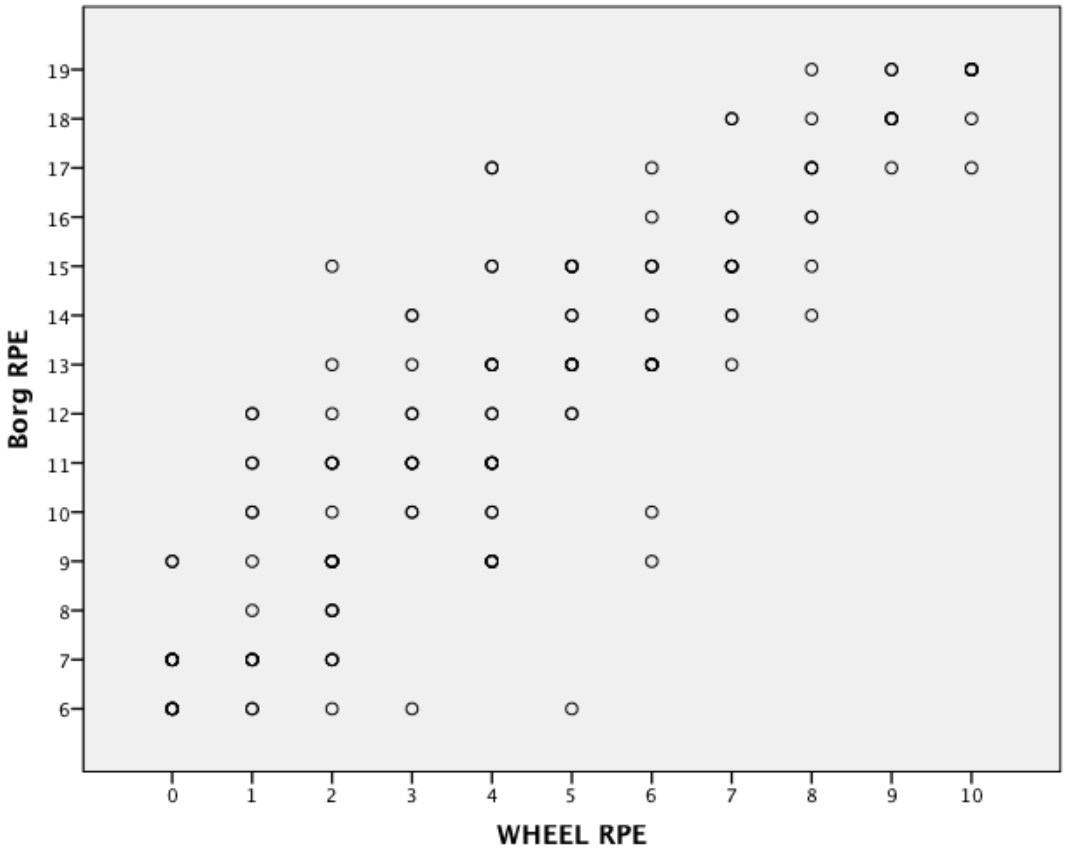

Figure 18. Scatter plot of WHEEL RPE and Borg RPE

Table 15. Linear regression of WHEEL RPE and Borg RPE

Model	DVs	Predictor	Unstandardized Coefficients B (SE)	t	p-value
$R^2 = 0.754$ p = 0.001 F (1,151) = 463.013	WHEEL RPE	(Constant)	7.207 (0.277)	26.013	0.0001
		Borg RPE	1.158 (0.054)	21.518	0.0001
*DVs: dependent variables; SE: standardized error					

4.4 DISCUSSION

In this study, we tested the concurrent and construct validity of a newly developed WHEEL Scale based on a symptom limited arm ergometry exercise stress test. For hypothesis 1 and hypothesis 2, the results showed modest positive correlations between physiological variables and both RPE scales. However, the correlation coefficients were small in comparison to previous studies that used the OMNI Scale in able-bodied individuals (Nakamura et al, 2009; Balasekaran et al, 2012; Krause et al, 2012). One possible explanation would be the small sample size in this study. Another explanation may be due to variability among participants in their exercise tolerance, which may have affected the results of the exercise testing. For example, the number of self-reported hours of exercise per week varied among participants from 0 to 9 hours. The findings were consistent with previous study (Buffart et al., 2008) that individuals with spina bifida who had lower levels of daily physical activity had significantly lower levels of aerobic fitness. This explanation was also supported by further analyses of differences between the values of relative VO_2 and relative HR among participants who had RER < 1 and who had RER ≥ 1. Participants who had RER < 1 had higher relative HR (mean ± SD = 89.59 ± 7.06 %) than who had RER ≥ 1 (mean ± SD = 75.79 ± 14.05%) and there was significant difference between these two groups ($t(149) = 5.875$, $p > 0.0001$). Participants who had RER < 1 had higher relative VO_2 (mean ± SD = 71.46 ± 20.69%) than who had RER ≥ 1 (mean ± SD = 61.24 ± 20.85%) and there was significant difference between these two groups ($t(157) = 2.665$, $p = 0.009$). Also, during the exercise stress test, participants used their upper extremities to pedal arm ergometer, which was performed by the small muscles of upper body when compared to large muscles of lower body that are used for leg ergometry exercise testing. It may be that participants did not

reach the levels of VO_2 that would be achievable with leg ergometry. Also, this is the first time many participants were exposed to the WHEEL and Borg scales; thus further training may have increased the correlation coefficients.

Hypothesis 3 was supported by the results showing that the relationship between the Borg Scale and the WHEEL Scale was highly correlated, which is consistent with findings in other studies that used the OMNI Scale (Robertson et al., 2004) and indicates high internal consistency, meaning they measured the same construct. Last, although this study used a small sample size, the experimental protocol and testing procedure were standardized to improve reliability (Robertson et al., 2005; Krause et al., 2012).

4.4.1 Future studies

Future studies should include practical trial of RPE scales during exercise tests and should be conducted in a larger population to confirm its validity and the inter- and intra-rater reliability.

4.4.2 Clinical significances

The WHEEL Scale has potential for use by physical therapists or exercise physiologists to monitor exercise intensity in people with spina bifida during therapeutic exercise. The benefit of using the WHEEL Scale is that it is easy to learn and it is possible to use without interrupting the exercise flow. Also, the WHEEL Scale would be a low cost method of monitoring exercise intensity in a clinical setting.

5.0 CONCLUSIONS

5.1 PULMONARY FUNCTION AND EXERCISE CAPACITY AMONG INDIVIDUALS WITH SPINA BIFIDA

This is a preliminary study that investigated the relationship between pulmonary function and exercise capacity among people with spina bifida. From session 2 and session 3, the results showed that restrictive lung condition was prevalent among people with spina bifida. Significant models to predict pulmonary function results were established by using the functional level of lesion and spinal fusion. Significant models to predict peak oxygen consumption in this population were established using body mass index (BMI), the functional level of lesion, and spinal fusion. Ninety percent of participants who were not able to perform the maximal exercise test met the criteria of restricted lung condition that was considered pulmonary limitation to exercise. An alternative analysis such as logistic regression would be used to determine the probability of restrictive lung condition among adolescents and adults with spina bifida.

Lower exercise capacity and higher risk of cardiovascular disease (CVD) were also found in this population that was in consistence with previous studies. In sight of functional level of lesion and spinal fusion were significant predictors for PFTs and knowing that participants who had higher level of lesion had higher probability of restrictive lung condition, these results provide clinicians or health care providers more information regarding to the lung functions in

this population by using these easy to get records. Also, given that people with spina bifida would be long-term wheelchair users, 10MWT could be used as a clinical tool in terms of assessing their propulsion velocity, frequency, and pattern. Future study could investigate on collecting more sample size in this population, then the center data pool could be used to compared between individuals and clinicians would be able to provide training or suggestions of reducing upper-extremity injury and enhancing functional activity.

5.2 USING THE WHEEL SCALE AS AN EXERCISE INTENSITY TOOL AMONG INDIVIDUALS WITH SPINA BIFIDA

The WHEEL Scale was developed for people with spina bifida who use manual wheelchairs when regulating exercise intensity. The concurrent validity and construct validity of the WHEEL Scale was proved by significant correlations between relative oxygen consumption (relative VO_{2peak}), relative hear rate (relative HR), and the Borg Scale during arm ergometry testing. In this study, the WHEEL RPE did not have as high of a correlation with physiological variables and with Borg RPE as previous studies have shown. Future studies were needed with a large cohort of people with spina bifida to test inter and intra-rater reliability. Also, given that hypo activity and lower rate of exercise were observed in this population, an alternative experimental protocol would be suggested in terms of providing training on using both RPE scales.

5.3 LIMITATIONS AND FUTURE WORK

There are few limitations in the present study. Though there were significant models for predicting pulmonary function results and exercise capacity, small sample size would affect the external validity meaning that it would not be able to generalize the results to entire population of spina bifida. Also, with this small sample size, the analysis methods that could be used were limit and the results should be interpreted with caution. However, even with small sample size, there were some significant findings that are worthy to notice, such as the lower rate of exercise habit and higher risk of CVD among participants with adolescents and adults with spina bifida. Another limitation would be determining the pulmonary limitation to exercise. In present study, respiratory exchange ratio (RER) was used to determine whether participants achieved maximal exercise then further analyzed their lung condition. Future study should investigate more in-depth of exercise physiological variables, such as using the slope of VE/VO_2 and collaborating with pulmonologist and exercise physiologist to exam the data in a comprehensive way.

Given that restricted lung condition, lower rate of aerobic capacity was prevalent among participants with adolescents and adults with spina bifida, further studies regards to management of cardiopulmonary function and exercise intervention should be emphasized. Using the WHEEL scale for regulating exercise intensity with training protocol for people with spina bifida is needed in future study. Also, differentiated RPE for arms and breathing should be collected in order to determine the reason for stopped the exercise test. Therefore, future studies with a large sample size are needed in order to test inter and intra-rater reliability of the WHEEL scale.

APPENDIX A

MACARTHUR COMPETENCE ASSESSMENT TOOL FOR CLINICAL RESEARCH

Evaluation to Sign Consent Form

Participant Initials: _____ Interviewer: _____ Date: _____

Please rate the participant's answers to questions in the below 5 areas using the following scale:

0 = inadequate understanding 1 = partial understanding 2 = adequate understanding

1. Purpose of the Project Rating: 0 1 2

What is the purpose of the research project that I described to you?

2. Activities involved in Participation Rating: 0 1 2

How many study visits are you asked to participate in?

How long will your participation in this research be if you decide to stay until the end?

3. Benefits of participation Rating: 0 1 2

In what way may you benefit by volunteering to participate in this study?

What might health care workers learn if people decide to be in this research project?

4. Risks/Discomforts of participation Rating: 0 1 2

Can you tell me about the possible risks of participating in this project?

5. Ability to withdraw Rating: 0 1 2

What will you do if you decide that you no longer want to participate in this study?

Total Score: _____ Total score must be 8 or higher to participate in research.

Adapted from Appelbaum, P. S., & Grisso, T. (2001). *MacArthur Competence Assessment Tool for Clinical Research*. Sarasota, FL: Professional Resource Exchange.

APPENDIX B

PATIENT SOCIODEMOGRAPHIC SURVEY

I'd now just like to get some basic information about you and your health.

1. Gender: (Check one)
_____ Male (1)
_____ Female (2)
_____ Transgender (3)

2. Are you of Spanish, Hispanic, or Latino origin or descent? (Check one)
____Yes, Spanish/Hispanic/Latino (1)
____No, not Spanish/Hispanic/Latino (2)
____Don't Know (88)
____Refused (99)

3. What is your race? (Check one)
____White (1)
____Black or African American (2)
____Asian (3)
____Native Hawaiian or Pacific Islander (4)
____American Indian or Alaskan Native (5)
____Other (specify) (6)
____Do not Know (88)
____Refused (99)

4. What is the highest level of education you completed? (Check one)
____8th grade/less (1)
____9th -11th grade (2)
____Graduated from high school/GED (3)
____Trade or Vocational school (4)
____Some college/Associate degree (5)
____College graduate (4 or 5 year program) (6)

____Some graduate school (7)
____Graduate degree (8)
____Other (9), specify _____
____Do not Know (88)
____Refused (99)

5. What is your current employment status? (Check one)
____Employed at a job for pay full-time (1)
____Employed at a job for pay part-time (2)
____Student, full-time (3)
____Student, part-time (4)
____Homemaker (5)
____Not currently employed retired (6)
____Not currently employed, not retired (7)
____Volunteer, part time (8)
____Other (9), specify _____
____Do not Know (88)
____Refused (99)

6. Which of the following categories best describes your total annual household income before taxes? (Check one)
____Less than $10,000 a year (1)
____$10,000 – $19,999 a year (2)
____$20,000 – $34,999 a year (3)
____$35,000 – $49,999 a year (4)
____$50,000 – $74,999 a year (5)
____$75,000 or more (6)
____Do not Know (88)
____Refused (99)

7. In general, would you say your health is: (Check one)
____Excellent (1)
____Very good (2)
____Good (3)
____Fair (4)
____Poor (5)
____Do not Know (88)
____Refused (99)

8. Compared to one year ago, how would you rate your health in general now? (Check one)
____Much better now than one year ago. (1)
____Somewhat better now than one year ago. (2)
____About the same as one year ago. (3)
____Somewhat worse now than one year ago. (4)
____Much worse than one year ago. (5)
____Do not Know (88)
____Refused (99)

9. What is your primary assistive device? (Check one)
____None (1)

____Lofstrand Crutches (2)
____Manual Wheelchair (3) TYPE: _____ CUSHION: _____
____Power Wheelchair (4) TYPE: _____ CUSHION: _____
____Walker (5)
____Cane or Canes (6)
____Crutches (7)
____Other (8), specify _____
____Do not Know (88)
____Refused (99)

10. What transportation do you have available to you? (Check one)
____Own a car (1)
____Have access to family car (2)
____Rely on family or friends to drive you (3)
____Use Access (4)
____Use Public transportation (5)
____None (6)
____Do not Know (88)
____Refused (99)

11. How did you get here today? (Check one)
____Own car (1)
____Family car (2)
____Friends car (3)
____Access (4)
____Public transportation (5)
____Other (6), specify _____
____Do not Know (88)
____Refused (99)

12. Do you currently smoke? (Check one)
____Yes (1)
____No (2)
____Do not Know (88)
____Refused (99)

13. Have you smoked in the past? (Check one)
____Yes (1)
____No (2)
____Do not Know (88)
____Refused (99)

14. Does anyone in your family smoke in the house?
____Yes (1)
____No (2)
____Do not Know (88)
____Refused (99)

15. What are your living arrangements? (Check one)
____Live at home with family (1)

____Live independently in own home (2)
____Live in an apartment building independently (3)
____Live in supported living (not with family) (4)
____Do not Know (88)
____Refused (99)

16. What is your level of spinal lesion? (Check one)
____Cervical (1)
____Thoracolumbar (2)
____Lumbar (3)
____Lumbosacral (4)
____Sacral (5)
____Do not Know (88)
____Refused (99)

17. How many hours of paid assistance do you receive each week? _____(Total number)

18. How many hours of unpaid assistance (i.e., family, friends) do you receive each week? _____(Total number)

19. What type of orthotics do you wear, if any?
____SMO's (1)
____AFO's (2)
____KAFO's (3)
____TLSO (4)
____Other (5), specify_____
____None (6)
____Do not Know (88)
____Refused (99)

20. How many hours per day do you wear your leg or foot orthotics? _____ (Total number)

21. How many hours per day to you wear your TLSO? _____ (Total number)

22. Are you involved in adaptive sports currently and/or have you been involved in the past?
____Yes, involved now (1)
____Yes, involved in the past (2)
____Yes, involved in the past and now (3)
____No (4)
____Do not Know (88)
____Refused (99)

23. Do you currently exercise (beyond participation in adaptive sports)?
____Yes (1)
____No (2)
____Do not Know (88)
____Refused (99)

24. If YES, do you exercise, how many hours per week on average do you exercise? _____ (Total Time)

25. Do you like to exercise?
____Yes (1)
____No (2)
____Somewhat (3)
_____ Do not Know (88)
_____ Refused (99)

26. Do you think exercise can improve your health?
____Yes (1)
____No (2)
____Somewhat (3)
____Do not Know (88)
____Refused (99)

APPENDIX C

BODY COMPOSITION MEASURMENTS

Body Composition:

- Gender: _____
- Height:

  R't _____ cm / _____ inch

  _____

  L't _____ cm / _____ inch

  _____

  Average _____ cm / _____ inch
- BMI (kg/m^2): _____
- Waist Circumference ____ cm / ____ inch
- Hand grip: (R't) ____ kg; (L't) ____ kg
- Type of Wheelchair _____
- Weight:

  Wheelchair + person: ____ kg / ____ lbs

  Wheelchair: _____ kg / _____ lbs

  _____

  _____

  Subject: _____ kg / _____ lbs
- Arm Span _____ cm / _____ inch
- 10 M Wheel test (1) ____ sec; (2) ____ sec
- Triceps Skinfold _____
- Type of Seat Cushion _____

APPENDIX D

PHYSICAL ACTIVITY SCALE FOR INDIVIDUALS WITH PHYSICAL DISABILITIES

Instructions: This questionnaire is about your current level of physical activity and exercise. Please remember there are no right or wrong answers. We simply need to assess your current level of activity.

Leisure Time Activity

1. During the past 7 days how often did you engage in *stationary activities* such as reading, watching TV, computer games, or doing handcrafts?
   1. Never (Go to question #2)
   2. Seldom (1–2d)
   3. Sometimes (3–4d)
   4. Often (5–7d)

   What were these activities?

   On average, how many hours per day did you spend in these *stationary activities*?
   1. Less than 1hr
   2. 1 but less than 2hr
   3. 2–4hr
   4. More than 4hr
2. During the past 7 days, how often did you *walk*, *wheel*, *push outside* your home *other than specifically for exercise*. For example, getting to work or class, walking the dog shopping, or other errands?
   1. Never (Go to question #3)
   2. Seldom (1–2d)
   3. Sometimes (3–4d)
   4. Often (5–7d)

   On average, how many hours per day did you spend wheeling or pushing outside your home?
   1. Less than 1hr
   2. 1 but less than 2hr
   3. 2–4hr
   4. More than 4hr
3. During the past 7 days, how often did you engage in *light sport or recreational activities* such as bowling, golf with a cart, hunting or fishing, darts, billiards or pool, therapeutic exercise (physical or occupational therapy, stretching, use of a standing frame) or other similar activities?
   1. Never (Go to question #4)
   2. Seldom (1–2d)
   3. Sometimes (3–4d)
   4. Often (5–7d)

   What were these activities?

   On average, how many hour per day did you spend in these *light sport or recreational* activities?
   1. Less than 1hr
   2. 1 but less than 2hr
   3. 2–4hr
   4. More than 4hr
4. During the past 7 days, how often did you engage in *moderate sport and recreational* activities such as doubles tennis, softball, golf without a cart, ballroom dancing, wheeling or pushing for pleasure or other similar activities?
   1. Never (Go to question #5)
   2. Seldom (1–2d)
   3. Sometimes (3–4d)
   4. Often (5–7d)

   What were these activities?

   On average, how many hours per day did you spend in these *moderate sport and recreational* activities?
   1. Less than 1hr
   2. 1 but less than 2hr
   3. 2–4hr
   4. More than 4hr
5. During the past 7 days, how often did you engage in *strenuous sport and recreational* activities such as jogging, wheelchair racing (training), off-road pushing, swimming, aerobic dance, arm cranking, cycling (hand or leg), singles tennis, rugby, basketball, walking with crutches and braces, or other similar activities
   1. Never (Go to question #6)
   2. Seldom (1–2d)
   3. Sometimes (3–4d)
   4. Often (5–7d)

   What were these activities?

   On average, how many hours per day did you spend in these *strenuous sport or recreational* activities?
   1. Less than 1hr
   2. 1 but less than 2hr
   3. 2–4hr
   4. More than 4hr
6. During the past 7 days, how often did you do any exercise *specifically to increase muscle strength and endurance* such as lifting weights, push-ups, pull-ups, dips, or wheelchair push-ups, etc?
   1. Never (Go to question #7)
   2. Seldom (1–2d)
   3. Sometimes (3–4d)
   4. Often (5–7d)

   What were these activities?

   On average, how many hours per day did you spend in these *exercises to increase muscle strength and endurance*?
   1. Less than 1hr
   2. 1 but less than 2hr
   3. 2–4hr
   4. More than 4hr

Household Activity

7. During the past 7 days, how often have you done any *light housework*, such as dusting, sweeping floors or washing dishes?
   1. Never (Go to question #8)
   2. Seldom (1–2d)
   3. Sometimes (3–4d)
   4. Often (5–7d)

   On average, how many hours per day did you spend doing *light housework*?
   1. Less than 1hr
   2. 1 but less than 2hr
   3. 2–4hr
   4. More than 4hr
8. During the past 7 days, how often have you done any *heavy housework or chores* such as vacuuming, scrubbing floors, washing windows, or walls, etc?
   1. Never (Go to question #9)
   2. Seldom (1–2d)
   3. Sometimes (3–4d)
   4. Often (5–7d)

   On average, how many hours per day did you spend doing *heavy housework or chores*?
   1. Less than 1hr
   2. 1 but less than 2hr
   3. 2–4hr
   4. More than 4hr
9. During the past 7 days, how often you done *home repairs* like carpentry, painting, furniture refinishing, electrical work, etc?
   1. Never (Go to question #10)
   2. Seldom (1–2d)
   3. Sometimes (3–4d)
   4. Often (5–7d)

   On average, how many hours per day did you spend doing *home repairs*?
   1. Less than 1hr
   2. 1 but less than 2hr
   3. 2–4hr
   4. More than 4hr
10. During the past 7 days how often have you done *lawn work or yard care* including mowing, leaf or snow removal, tree or bush trimming, or wood chopping, etc?
    1. Never (Go to question #11)
    2. Seldom (1–2d)
    3. Sometimes (3–4d)
    4. Often (5–7d)

    On average, how many hours per day did you spend doing *lawn work*?
    1. Less than 1hr
    2. 1 but less than 2hr
    3. 2–4hr
    4. More than 4hr
11. During the past 7 days, how often have you done *outdoor gardening*?
    1. Never (Go to question #12)
    2. Seldom (1–2d)
    3. Sometimes (3–4d)
    4. Often (5–7d)

    On average, how many hours per day did you spend doing *outdoor gardening*?
    1. Less than 1hr
    2. 1 but less than 2 hr
    3. 2–4hr
    4. More than 4hr
12. During the past 7 days, how often did you *care for another person*, such as children, a dependent spouse, or another adult?
    1. Never (Go to question #13)
    2. Seldom (1–2d)
    3. Sometimes (3–4d)
    4. Often (5–7d)

    On average, how many hours per day did you spend *caring for another person*?
    1. Less than 1hr
    2. 1 but less than 2hr
    3. 2–4hr
    4. More than 4hr

Work-Related Activity

13. During the past 7 days, how often did you *work for pay or as a volunteer*? (Exclude work that mainly involved sitting with slight arm movement such as light office work, computer work, light assembly line work, driving bus or van, etc.)
    1. Never (Go to END)
    2. Seldom (1–2d)
    3. Sometimes (3–4d)
    4. Often (5–7d)

    On average, how many hours per day did you spend *working for pay or as a volunteer*?
    1. Less than 1hr
    2. 1 but less than 4hr
    3. 5 but less than 8hr
    4. 8hr or more

Scoring: PASIPD

Item multipliers

1. Not scored
2. 2.5
3. 3.0
4. 4.0
5. 8.0
6. 5.5
7. 1.5
8. 4.0
9. 4.0
10. 4.0
11. 4.0
12. 1.5
13. 2.5

Average Hours Per Day Calculation for Items 2–12

Category	Reported (hr/d)	Average (hr/d)
Seldom (1–2d)	<1	.11
	1–2	.32
	2–4	.64
	>4	1.07
Sometimes (3–4d)	<1	.25
	1–2	.75
	2–4	1.50
	>4	2.50
Often (5–7d)	<1	.43
	1–2	1.29
	2–4	2.57
	>4	4.29

Average Hours Per Day Calculation for Item 13

Category	Reported (hr/d)	Average (hr/d)
Seldom (1–2d)	<1	.12
	1–4	.64
	5–8	1.39
	>8	1.93
Sometimes (3–4d)	<1	.28
	1–4	1.5
	5–8	3.11
	>8	4.5
Often (5–7d)	<1	.49
	1–4	2.57
	5–8	5.57
	>8	7.71

NOTE. PASIPD score = sum of item multiplier × average hours per day over items 2–13.

APPENDIX E

PULMONARY FUNCTION TESTS CHECKLIST

E.1 SPIROMETRY

Measures: Forced vital capacity (FVC), forced expiratory volume in one second (FEV1), and FEV1/FVC.

Equipment Calibrated		NOTES
Familiarize participant with equipment and technician		
Participant transfers to box		
Type of transfers	D / A / I	
Level of Assistance	SBA / CGA / Min / Mod / Max	
Position participant		
Posture		
Supports		
Cushion	CAT / Subject own	
D= Dependent Transfer; A= Assisted Transfer; I= Independent Transfer SBA= Stand By Assistance; CGA= Contact Guard Assistance; Min= Minimal Assistance; Mod= Moderate Assistance; Max= Maximal Assistance		
Instruction/Demonstration		
Explanation of Test Provided		
Demonstrate/provide person with opportunity to try breathing maneuver without the mouthpiece.		

Demonstrate/provide person with opportunity get on the mouthpiece correctly and attach the nose clip		
*Start Test/Trial 1***		
Stable baseline values obtained with no leaks		
Record 1st maneuver		
Visually inspect volume/time and flow/volume loop (exclude if "flow volume curve does not show rapid rise to peak flow, and smooth descending line, without evidence of cough or glottis closure." [1]		
Repeat test	Y N	
Trial 2**		
Stable baseline values obtained with no leaks		
Record 1st maneuver		
Visually inspect volume/time and flow/volume loop (exclude if "flow volume curve does not show rapid rise to peak flow, and smooth descending line, without evidence of cough or glottis closure." [1]		
Repeat test	Y N	
Trial 3**		
Stable baseline values obtained with no leaks		
Record 1st maneuver		
Visually inspect volume/time and flow/volume loop (exclude if "flow volume curve does not show rapid rise to peak flow, and smooth descending line, without evidence of cough or glottis closure." [1]		
Repeat test	Y N	
Trial 4**		
Stable baseline values obtained with no leaks		
Record 1st maneuver		
Visually inspect volume/time and flow/volume loop (exclude if "flow volume curve does not show rapid rise to peak flow, and smooth descending line, without evidence of cough or glottis closure." [1]		
Repeat test	Y N	
Trial 5**		
Stable baseline values obtained with no leaks		

Record 1st maneuver		
Visually inspect volume/time and flow/volume loop (exclude if "flow volume curve does not show rapid rise to peak flow, and smooth descending line, without evidence of cough or glottis closure." [1]		
Repeat test	Y N	
Trial 6**		
Stable baseline values obtained with no leaks		
Record 1st maneuver		
Visually inspect volume/time and flow/volume loop (exclude if "flow volume curve does not show rapid rise to peak flow, and smooth descending line, without evidence of cough or glottis closure." [1]		
Repeat test	Y N	
Trial 7**		
Stable baseline values obtained with no leaks		
Record 1st maneuver		
Visually inspect volume/time and flow/volume loop (exclude if "flow volume curve does not show rapid rise to peak flow, and smooth descending line, without evidence of cough or glottis closure." [1]		
Repeat test	Y N	

**CRITERIA:

For spirometry we will confirm we have good trials if FVC and FEV1 are within 0.15 L of each other. For those with low FVC or FEV1 (anything < 1L) they have to be within 0.10 L.
May need 7-8 trials to get 2 reproducible results.
Need 6 seconds exhalation time and/or end of test criteria met (i.e. less than 25ml/sec flow).
Do a 3rd test even if get 2 reproducible tests.

E.2 LUNG VOLUMES:

Measures: Functional residual capacity (FRC) – volume of gas present in lung at end-expiration during tidal breathing; Residual volume (RV) – volume of gas remaining in lung after max exhalation; Total lung capacity (TLC) = FRC + RV – volume of gas in lungs after maximal inspiration (sum of all volumes)

Adjust to patient so he or she is sitting comfortably in chamber and can reach the mouthpiece with neck in neutral position – not too much flexion or extension		
(Mary) We're measuring how much air is in your lungs. **We're going to shut the door and wait one minute for the temperature to stabilize. Then I'll tell you to go on the mouthpiece, put the clips on your nose, and hold your cheeks so air will not fill them later. I'll tell you to breathe normally until we have a stable baseline, then I'll warn you that the shutter is closing. When that happens I want you to pull then push against that shutter as gently as you can for 2-3 efforts. I'll guide you like this: (demonstrate with your hand in/out, pull/push). When the shutter releases, I'll tell you to breathe in as deeply as you can, then exhale as far as you can, then breathe in deeply as possible and come off the mouthpiece. Remember to listen to me; I'll guide you through it.**		**Note: we can have them take 2-3 normal breaths before the slow vital capacity maneuver)**
Trial 1		
"The door will be closed now."	**Door is closed first, then subject goes on mouthpiece**	**Step 1**
Allow time for thermal transients to stabilize and the patient to relax		**Step 1**
Try it now: Go on the mouthpiece, put the nose clip on, now hold your cheeks. Breathe in and out to my command, watch my hand. It's a gentle pull/push.	**(Note: have them breathe normally at first at their rhythm.)**	**Step 2**
Instruct patient to put *"mouthpiece on; breath normally"*	**Get 3-5 tidal breaths; 10 is usually too many. That will tire them and keep them in the box too long.**	
"put your hands on your cheeks		
"and on the next breath in….Breath in/out, in/out (pair with hand motion up/down)	**This should be at their resting TV and freq.** **(Note: shutter is closed until they've reached a preset pressure change-about 2-3 breaths.)**	
"Feel the shutter…..push/ pull….keep this up"	Person instructed to perform series of gentle pants between 0.5 and 1.0 Hz (if > 1.5 then errors, if < 0.5 causes problems with controlled leak of plethysmograph system –metronome suggested, we will use hand signal/verbal cue.	
"Ok you can come off"	Permit the person to come off of the mouthpiece and provide a rest between maneuvers. If person is short of breath allow he or she to take 2-3 tidal breaths after panting	**They may take 2-3 normal breaths, then perform a SVC maneuver-described above.**

	maneuver	
Trial 2		
"The door will be closed now."	**Door is closed first, then subject goes on mouthpiece**	**Step 1**
Allow time for thermal transients to stabilize and the patient to relax		**Step 1**
Try it now: Go on the mouthpiece, put the nose clip on, now hold your cheeks. Breathe in and out to my command, watch my hand. It's a gentle pull/push.	**(Note: have them breathe normally at first at their rhythm.)**	**Step 2**
Instruct patient to put *"mouthpiece on; breath normally"*	**Get 3-5 tidal breaths; 10 is usually too many. That will tire them and keep them in the box too long.**	
"put your hands on your cheeks		
"and on the next breath in….Breath in/out, in/out (pair with hand motion up/down)	**This should be at their resting TV and freq.** **(Note: shutter is closed until they've reached a preset pressure change-about 2-3 breaths.)**	
"Feel the shutter…..push/ pull….keep this up"	Person instructed to perform series of gentle pants between 0.5 and 1.0 Hz (if > 1.5 then errors, if < 0.5 causes problems with controlled leak of plethysmograph system –metronome suggested, we will use hand signal/verbal cue.	
"Ok you can come off"	Permit the person to come off of the mouthpiece and provide a rest between maneuvers. If person is short of breath allow he or she to take 2-3 tidal breaths after panting maneuver	**They may take 2-3 normal breaths, then perform a SVC maneuver-described above.**
"Ok you can come off"	Permit the person to come off of the mouthpiece and provide a rest between maneuvers. If person is short of breath allow he or she to take 2-3 tidal breaths after panting maneuver	**They may take 2-3 normal breaths, then perform a SVC maneuver-described above.**
Trial 3		
"The door will be closed now."	**Door is closed first, then subject goes on mouthpiece**	**Step 1**
Allow time for thermal transients to stabilize and the patient to relax		**Step 1**
Try it now: Go on the mouthpiece, put the nose clip	**(Note: have them breathe normally at first at their**	**Step 2**

on, now hold your cheeks. Breathe in and out to my command, watch my hand. It's a gentle pull/push.	**rhythm.)**	
Instruct patient to put *"mouthpiece on; breath normally"*	**Get 3-5 tidal breaths; 10 is usually too many. That will tire them and keep them in the box too long.**	
"put your hands on your cheeks		
"and on the next breath in….Breath in/out, in/out (pair with hand motion up/down)	**This should be at their resting TV and freq. (Note: shutter is closed until they've reached a preset pressure change-about 2-3 breaths.)**	
"Feel the shutter…..push/ pull….keep this up"	Person instructed to perform series of gentle pants between 0.5 and 1.0 Hz (if > 1.5 then errors, if < 0.5 causes problems with controlled leak of plethysmograph system –metronome suggested, we will use hand signal/verbal cue.	
"Ok you can come off"	Permit the person to come off of the mouthpiece and provide a rest between maneuvers. If person is short of breath allow he or she to take 2-3 tidal breaths after panting maneuver	**They may take 2-3 normal breaths, then perform a SVC maneuver-described above.**
Trial 4		
"The door will be closed now."	**Door is closed first, then subject goes on mouthpiece**	**Step 1**
Allow time for thermal transients to stabilize and the patient to relax		**Step 1**
Try it now: Go on the mouthpiece, put the nose clip on, now hold your cheeks. Breathe in and out to my command, watch my hand. It's a gentle pull/push.	**(Note: have them breathe normally at first at their rhythm.)**	**Step 2**
Instruct patient to put *"mouthpiece on; breath normally"*	**Get 3-5 tidal breaths; 10 is usually too many. That will tire them and keep them in the box too long.**	
"put your hands on your cheeks		
"and on the next breath in….Breath in/out, in/out (pair with hand motion up/down)	**This should be at their resting TV and freq. (Note: shutter is closed until they've reached a preset**	

	pressure change-about 2-3 breaths.)	
"Feel the shutter…..push/ pull….keep this up"	Person instructed to perform series of gentle pants between 0.5 and 1.0 Hz (if > 1.5 then errors, if < 0.5 causes problems with controlled leak of plethysmograph system –metronome suggested, we will use hand signal/verbal cue.	
"Ok you can come off"	Permit the person to come off of the mouthpiece and provide a rest between maneuvers. If person is short of breath allow he or she to take 2-3 tidal breaths after panting maneuver	**They may take 2-3 normal breaths, then perform a SVC maneuver-described above.**
Trial 5		
"The door will be closed now."	**Door is closed first, then subject goes on mouthpiece**	**Step 1**
Allow time for thermal transients to stabilize and the patient to relax		**Step 1**
Try it now: Go on the mouthpiece, put the nose clip on, now hold your cheeks. Breathe in and out to my command, watch my hand. It's a gentle pull/push.	**(Note: have them breathe normally at first at their rhythm.)**	**Step 2**
Instruct patient to put *"mouthpiece on; breath normally"*	**Get 3-5 tidal breaths; 10 is usually too many. That will tire them and keep them in the box too long.**	
"put your hands on your cheeks		
"and on the next breath in….Breath in/out, in/out (pair with hand motion up/down)	**This should be at their resting TV and freq. (Note: shutter is closed until they've reached a preset pressure change-about 2-3 breaths.)**	
"Feel the shutter…..push/ pull….keep this up"	Person instructed to perform series of gentle pants between 0.5 and 1.0 Hz (if > 1.5 then errors, if < 0.5 causes problems with controlled leak of plethysmograph system –metronome suggested, we will use hand signal/verbal cue.	
"Ok you can come off"	Permit the person to come off of the mouthpiece and provide a rest between maneuvers. If person is short of breath allow he or she to take 2-3 tidal breaths after panting maneuver	**They may take 2-3 normal breaths, then perform a SVC maneuver-described above.**

** CRITERIA: For restrictive conditions we will see low RV. We can use this formula: TLC – VC = RV to determine RV. TLC variability < 5% between tests. For VC, look at 2 trials to make sure they are close (less than 0.15 L between best and second best).
3-5 technically satisfactory panting maneuvers recorded.
Should see series of superimposed straight lines with slight space due to thermal drift
A minimum of 3 FRCpleth values that agree within 5% (difference between highest and lowest value/mean is ≤ 5%)
According to ACSM the following are characteristics of how severe restrictive the lung condition is and is based on the preset of predicted values. When restrictive component is presents, TLC is required in order to confirm restriction is present.

Stage	Characteristics
Mild	FVC less than the lower limit of normal but ≥ 70% of predicted
Moderate	FVC 60% to 69% of predicted
Moderately Severe	FVC 50% to 59% of predicted
Severe	FVC 34% to 49% of predicted
Very Severe	FVC < 34% of predicted

APPENDIX F

GRADED MAXIMAL EXERCISE TEST CHECKLIST AND RECORD FORM

F.1 GRADED MAXIMAL EXERCISE TEST

Following the pulmonary function test, subjects will return to the 8TH floor of Montefiore with one of the co-investigator where the participant will undergo a graded maximal exercise test (Theresa and/or Yu-Ting to transport)

1. **EXPLANATION OF TEST TO PARTICIPANT: The following explanation will be provided to each subject by one of the co-investigators** (Theresa):
    - This will be a <u>one-time</u> graded maximal exercise test.
    - We will find out more about your physical fitness and how your body uses the oxygen that moves through your body during exercise.
    - To measure how your body uses oxygen, we will ask you to wear a mask over your mouth and nose that lets you breathe. (*Show the Hans Rudolph mask and how it fits over the mouth and nose, can let the person hold the mask and try to place it).*
    - While you breathe in and out, this machine (point to the AEI Moxus system) will measure the oxygen in your breath and other things, like how fast your heart is beating.
    - During the test we will be asking you to tell us how hard you feel you are working on these 2 scales (WC RPE scale and BORG scale).
    - Attach Polar monitor and watch
    - Ask participant “are you comfortable”
2. **EXPLANATION OF RPE TO PARTICIPANT** (Theresa):

- *"During the exercise test we want you to <u>pay close attention</u> to how hard you feel the exercise work rate is. This feeling should reflect your total amount of exertion and fatigue, combining all sensations and feeling of physical stress, effort and fatigue. Don't concern yourself with any one factor such as [arm] pain, shortness of breath or exercise intensity, but try to concentrate on your total inner feeling of exertion. <u>Try not to underestimate or overestimate your feelings of exertion; be as accurate as you can."</u>*
- We will be holding this scale up so you can see it while you are pedaling during the test.
- You will be ask to pick a number on this scale that reflects how exerted you feel.
- I will read the list of numbers and you can nod your head when I reach the correct one so that you do not have to take your hands off the wheel. If you feel I don't understand you, just say the number.
- For exercise testing where you are giving it your all, you may reach the higher numbers.
- Do you need to use the restroom before we get started?

3. **SET UP ON SARATOGA** (Theresa and Yu–Ting)
   - Assist person up ramp – 1 person standing behind participant (Yu-Ting or Theresa)
   - Fit to Saratoga (Yu-Ting or Theresa) **
   - Ask the person to do a few turns of the crank, and ask if they are comfortable. Place blocks under wheels after set up on manual wheelchair wheels
   - Ask if person wants the headrest, if so follow instructions for fitting it above
   - Ask if person can see the screen – if they can't, then angle the screen down, but make sure it clears the arm cranks as they move
   - Ask, *"Can you see the screen of the Saratoga"*– if they can't, then angle the screen down, but make sure it clears the arm cranks as they move.
   - Ask participant "are you comfortable"
   - Input information into <u>*Saratoga computer screen*</u>
     - Select 1 (program) and scroll to W (watts mode)
     - Press ENTER
     - Display on R will blink for weight. Enter Weight, using up/down arrows, press ENTER. Display on R will blink for time, use up/down arrows to enter 40 minutes, press ENTER.
     - Watts flashes – should be on 10, if not use up/down
     - Arrows to set to 10 (lowest wattage), press ENTER
   - Instruction on use of Saratoga and <u>*Demonstrate speed and metronome*</u>
4. **FIT MASK** (Theresa and Yu-Ting)
   - Pick smallest, most comfortable mask, may use Hans measures

- Check mask seal (your hand over mask) and ask subject and ask, “Please breathe in and out, do you feel any leaks”
- Apply mask that fits the best and headgear that matches with it
- Check fit
- Tighten straps or get smaller mask if leaks occur as person is asked to breathes out hard as investigator covers opening in mask
- Ask participant “are you comfortable”

5. **EXPLANATION OF TEST:**
   - Explain to subject that we will be reaching in to change setting Watts several times during the test
   - “We will start the exercise testing. You will be asked to crank the Saratoga at a set rate of speed (29-30 on speed display which equals 70 RPM). While you are cranking you will be breathing through a mask that will tell me about how your body is using oxygen. Expect to sweat and to be winded or out of breath. We want you to give us your very best effort at cranking the arms of the bike for as long as you possibly can. We want you to pedal until you absolutely have to stop. You are going to feel really tired. We are trying to get everything out of you that we can so we need your very strongest effort. You will be sweating and breathing fast because you will be putting out an effort similar to if you were in a race to win it.”
6. **INSTRUCT PERSON TO NOTIFY US OF ANY ADVERSE REACTIONS DURING THE EXERCISE**
   - “That being said, “It is also important that you let me know if you have any symptoms such as chest pain or heart palpations, extreme shortness of breath, lightheadedness, nausea or feeling confused and we will stop the test.”
   - “During the test I prefer you don’t talk because that will affect your test, unless you need to let me know of any of the symptoms I described, but we will be talking to you throughout the test. We will be asking you for a number on the Borg scale. I will read the numbers to you and you will give me a clear yes nod of your head if I hit the correct number.”
7. **RESTING MEASURES** include: (Yu-Ting will record manually)
   - Record resting heart rate from EKG or the Polar monitor
   - Blood pressure two times, take the average of the two
     - Calculate max HR 220-age
8. **GRADED MAXIMAL EXERCISE TEST** (Steve conducting test, Theresa will stay at person’s side and change Saratoga watts/ ask RPE, Yu-Ting/technician manually records data)
   - Participants will be encouraged to crank the arm ergometer at a set rate of speed (70 rpm) and to try to match the speed on the Saratoga of 29.7 and the cadence of

the metronome (matching the cadence while the resistance is increased every 1 minutes *(manually increased by investigator)*

- 15s before 1 minute mark, ask for RPE body, lungs and manually record + HR, speed (29.7) and VO2
- Metronome *(KORG or the one Steve has)* will be used as an auditory or visual cue to help participants maintain a steady pace. Also, a speed display on R of screen (29.7 = 70 W).
- Watch R (watch for 1.2) and VO2 changes (should increase over time), watch HR (watch for nearing HR max)
- Provide encouragement but don't tell them when I am changing resistance

9. MEASURES

- During the test the following will be recorded: Time, RR, Vt, Ve, VO2, VCO2, VO2/kg, RER, HR, Mix O2, MixCO2, VE/VO2, Ti, Ttot, Ti/Ttot, Vt/Ti, PetCO2, VeqO2, VeqCO2, VO2/BSA, VCO2/BSA

10. END POINT

- Increase in Watts every 1 minute until:
- Testing will stop prior to the end of the test if subjects show any adverse reactions or if they feel they cannot continue.
- Adverse reactions include but are not limited to: Chest pain or palpitations, extreme shortness of breath, lightheadedness or confusion, nausea or vomiting, or signs of peripheral ischemia.
- Investigator observes signs subject needs to stop
- Record reason for ending test in chart

11. **COOL DOWN**

- Drop Watts down to lower wattage (10) when test finished.
- Subject continues cranking the ergometer during cool down
- Record BP immediately after + HR from polar
- Record BP + HR again

12. TEST RESULT/EXERCISE INSTRUCTION:

- Provide a copy of the results of the test result to each participant for their physician
- We will provide the following information to participants:

  1. A copy of a summary of the results of the test to each participant.
  2. Suggested starting point for exercise.
  3. General information packet about physical activity including:
     a). Exercise Instruction sheet and personal exercise contract
     b). Options for adaptive exercise and resources on physical activity
     c). Contact information/brochures for the local adaptive sports networks and gyms

4. We can offer to be a resource for participants should he or she have questions or need a referral.

***Instructions for set up per the Saratoga guide (p. 10):** *"If you are using the adjustable height table, adjust its height such that the axis of the cranks is approximately at the height of your shoulders. At this height, your arms will be horizontal at the farthest extent of crank rotation. The red locking knob must be loosened for raising or lowering the table. Tighten it securely when at desired height. After adjusting table height, adjust your chair position such that your hand, when your arm is fully extended and while gripping the grip of choice, is comfortably seated on the grip. If you use a power chair, be sure to turn the chair power off and break your chair before starting to cycle. Before using, you will want to adjust your Saratoga Silver I headrest assembly. It can be raised and lowered using the 3/8" pull pin set in the proper hole and it can be adjusted horizontally by sliding the horizontal headrest tube into its receiver. While this item is utilized mainly by those without trunk stability, some with trunk stability find it helpful."*

F.2 GRADED MAXIMAL EXERCISE RECORD FROM

RHR (POLAR): _____ RHR (Metabolic cart): _____

RESTING BP 1) ____ / ____ 2) ____ / ____

POST BP 1) ____ / ____ 2) ____ / ____

	Time (min)	Watts (w)	RPE (Borg)	RPE (WC)	Speed (29.7 goal)	Time (on metabolic cart)	Comments
Warm-up	0-1	10					
Exercise	1-1	10					
	2-3	20					
	3-4	30					
	4-5	40					
	5-6	50					
	6-7	60					
	7-8	70					
	8-9	80					
	9-10	90					
	10-11	100					
	11-12	110					
	12-13	120					
Recovery	13-14	10					
	14-15	10					
	15-16	10					
	16-17	10					
	17-18	10					

Reason for Test Termination and other notes on test:

_____

_____

_____

_____

_____

BIBLIOGRAPHY

Abresch, R. T., McDonald, D. a, Widman, L. M., McGinnis, K., & Hickey, K. J. (2007). Impact of spinal cord dysfunction and obesity on the health-related quality of life of children and adolescents. *The Journal of Spinal Cord Medicine*, *30*(Suppl 1), S112–8. Retrieved from http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=2031984&tool=pmcentrez&rendertype=abstract

Adzick, N. S., Thom, E. A., Spong, C. Y., Brock III, J. W., Burrows, P. K., Johnson, M. P., … Farmer, D. L. (2011). A Randomized Trial of Prenatal versus Postnatal Repair of Myelomeningocele. *The New England Journal of Medicine*, *364*(11), 993–1004.

American College of Sports Medicine. (2010). *ACSM's Guidelines for Exercise Testing and Prescription*. Lippincott Williams & Wilkins.

American Lung Association. (2014). Retrieved July 02, 2014, from http://www.lung.org/lung-disease/pneumonia/

Askari, S., Kirby, R. L., Parker, K., Thompson, K., & O'Neill, J. (2013). Wheelchair propulsion test: development and measurement properties of a new test for manual wheelchair users. *Archives of Physical Medicine and Rehabilitation*, *94*(9), 1690–8. doi:10.1016/j.apmr.2013.03.002

Balady, G. J., Arena, R., Sietsema, K., Myers, J., Coke, L., Fletcher, G. F., … Milani, R. V. (2010). Clinician's Guide to cardiopulmonary exercise testing in adults: a scientific statement from the American Heart Association. *Circulation*, *122*(2), 191–225. doi:10.1161/CIR.0b013e3181e52e69

Balasekaran, G., Loh, M. K., Govindaswamy, V. V., & Robertson, R. J. (2012). OMNI Scale of Perceived Exertion: mixed gender and race validation for Singapore children during cycle exercise. *European Journal of Applied Physiology*, *112*(10), 3533–46. doi:10.1007/s00421-012-2334-8

Battikha, M., Sa, L., Porter, A., & Taylor, J. A. (2014). Relationship between pulmonary function and exercise capacity in individuals with spinal cord injury. *American Journal of Physical Medicine & Rehabilitation*, *93*(5), 413–421. doi:10.1097/PHM.0000000000000046

Borg, G. A. V. (1982). Psychophysical bases of perceived exertion. *Medicine & Science in Sports & Exercise*, *14*(5), 377–81.

Botto, L. D., Moore, C. A., Khoury, M. J., & Erickson, J. D. (1999). Neural-tube defects. *The New England Journal of Medicine*, *341*(20), 1509–1519.

Boulet, S. L., Yang, Q., Mai, C., Kirby, R. S., Collins, J. S., Robbins, J. M., … Mulinare, J. (2008). Trends in the postfortification prevalence of spina bifida and anencephaly in the United States. *Birth Defects Research. Part A, Clinical and Molecular Teratology*, *82*(7), 527–32. doi:10.1002/bdra.20468

Buffart, L. M., Roebroeck, M. E., Rol, M., Stam, H. J., & van den Berg-Emons, R. J. G. (2008). Triad of physical activity, aerobic fitness and obesity in adolescents and young adults with myelomeningocele. *Journal of Rehabilitation Medicine : Official Journal of the UEMS European Board of Physical and Rehabilitation Medicine*, *40*(1), 70–5. doi:10.2340/16501977-0135

Buffart, L. M., van den Berg-Emons, R. J., Burdorf, A., Janssen, W. G., Stam, H. J., & Roebroeck, M. E. (2008). Cardiovascular disease risk factors and the relationships with physical activity, aerobic fitness, and body fat in adolescents and young adults with myelomemningocele. *Archives of Physical Medicine and Rehabilitation*, *89*(11), 2167–73. doi:10.1016/j.apmr.2008.04.015

Buffart, L. M., van den Berg-Emons, R. J. G., van Wijlen-Hempel, M. S., Stam, H. J., & Roebroeck, M. E. (2008). Health-related physical fitness of adolescents and young adults with myelomeningocele. *European Journal of Applied Physiology*, *103*(2), 181–8. doi:10.1007/s00421-008-0684-z

Centers for Disease Control and Prevention. (2014). Spina Bifida Facts. *CDC*. Retrieved from Centers for Disease Control and Prevention

Ciprandi, G., Schiavetti, I., Bellezza Fontana, R., Sorbello, V., & Ricciardolo, F. L. M. (2014). Overweight and obesity as risk factors for impaired lung function in patients with asthma: A real-life experience. *Allergy and Asthma*, *35*(4), e62–e71.

Classification of Overweight and Obesity by BMI, Waist Circumference, and Associated Disease Risks. (2014). *National Institutes of Health*. Retrieved July 17, 2014, from http://www.nhlbi.nih.gov/health/public/heart/obesity/lose_wt/bmi_dis.htm

Cooper, R. a, Baldini, F. D., Langbein, W. E., Robertson, R. N., Bennett, P., & Monical, S. (1993). Prediction of pulmonary function in wheelchair users. *Paraplegia*, *31*(9), 560–70. doi:10.1038/sc.1993.91

Cowan, R. E., Boninger, M. L., Sawatzky, B. J., Mazoyer, B. D., & Cooper, R. a. (2008). Preliminary outcomes of the SmartWheel Users' Group database: a proposed framework for

clinicians to objectively evaluate manual wheelchair propulsion. *Archives of Physical Medicine and Rehabilitation*, *89*(2), 260–8. doi:10.1016/j.apmr.2007.08.141

Crapo, R., AH, M., & RM, G. (1981). Reference spirometric values using techniques and equipment that meet ATS recommendations. *American Review of Respiratory Disease*, *123*(6), 659–664.

De Groot, J. F., Takken, T., van Brussel, M., Gooskens, R., Schoenmakers, M., Versteeg, C., … Helders, P. (2011). Randomized controlled study of home-based treadmill training for ambulatory children with spina bifida. *Neurorehabilitation and Neural Repair*, *25*(7), 597–606. doi:10.1177/1545968311400094

Dicianno, B. E., Kurowski, B. G., Yang, J. M. J., Chancellor, M. B., Bejjani, G. K., Fairman, A. D., … Sotirake, J. (2008). Rehabilitation and medical management of the adult with spina bifida. *American Journal of Physical Medicine & Rehabilitation / Association of Academic Physiatrists*, *87*(12), 1027–50. doi:10.1097/PHM.0b013e31818de070

Dicianno, B. E., & Wilson, R. (2010). Hospitalizations of adults with spina bifida and congenital spinal cord anomalies. *Archives of Physical Medicine and Rehabilitation*, *91*(4), 529–35. doi:10.1016/j.apmr.2009.11.023

Dishman, R. K. (1994). Prescribing exercise intensity for healthy adults using perceived exertion. *Medicine and Science in Sports and Exercise*, *26*(9), 1087–94.

Dosa, N. P., Foley, J. T., Eckrich, M., Woodall-Ruff, D., & Liptak, G. S. (2009). Obesity across the lifespan among persons with spina bifida. *Disability and Rehabilitation*, *31*(11), 914–20. doi:10.1080/09638280802356476

Falvo, D. R. (2013). *Medical and Psychosocial Aspects of Chronic Illness and Disability* (p. 156). Jones & Bartlett Learning, LLC.

Friedman, D., Holmbeck, G. N., DeLucia, C., Jandasek, B., & Zebracki, K. (2009). Trajectories of autonomy development across the adolescent transition in children with spina bifida. *Rehabilitation Psychology*, *54*(1), 16–27. doi:10.1037/a0014279

Galeiras Vázquez, R., Rascado Sedes, P., Mourelo Fariña, M., Montoto Marqués, A., & Ferreiro Velasco, M. E. (2013). Respiratory management in the patient with spinal cord injury. *BioMed Research International*, *2013*, 168757. doi:10.1155/2013/168757

Goosey-Tolfrey, V., Lenton, J., Goddard, J., Oldfield, V., Tolfrey, K., & Eston, R. (2010). Regulating intensity using perceived exertion in spinal cord-injured participants. *Medicine and Science in Sports and Exercise*, *42*(3), 608–13. doi:10.1249/MSS.0b013e3181b72cbc

Grange, C. C., Bougenot, M. P., Groslambert, A., Tordi, N., & Rouillon, J. D. (2002). Perceived exertion and rehabilitation with wheelchair ergometer: comparison between patients with

spinal cord injury and healthy subjects. *Spinal Cord*, *40*(10), 513–8. doi:10.1038/sj.sc.3101353

Grisso, T., Appelbaum, P. S., & Hill-Fotouhi, C. (1997). The MacCAT-T: a clinical tool to assess patients' capacities to make treatment decisions. *Psychiatric Services (Washington, D.C.)*, *48*(11), 1415–9. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/9355168

Hicks, a L., Martin, K. a, Ditor, D. S., Latimer, a E., Craven, C., Bugaresti, J., & McCartney, N. (2003). Long-term exercise training in persons with spinal cord injury: effects on strength, arm ergometry performance and psychological well-being. *Spinal Cord*, *41*(1), 34–43. doi:10.1038/sj.sc.3101389

Krause, M. P., Goss, F. L., Robertson, R. J., Kim, K., Elsangedy, H. M., Krinski, K., & Da Silva, S. G. (2012). Concurrent Validity of An OMNI Rating of Perceived Exertion Scale for Bench Stepping Exercise. *Journal of Strength and Conditioning Research*, *26*(2), 506–512.

Liptak, G. S., Kennedy, J. a, & Dosa, N. P. (2010). Youth with spina bifida and transitions: health and social participation in a nationally represented sample. *The Journal of Pediatrics*, *157*(4), 584–8, 588.e1. doi:10.1016/j.jpeds.2010.04.004

Lynch, A., Ryu, J.-C., Agrawal, S., & Galloway, J. C. (2009). Power mobility training for a 7-month-old infant with spina bifida. *Pediatric Physical Therapy : The Official Publication of the Section on Pediatrics of the American Physical Therapy Association*, *21*(4), 362–8. doi:10.1097/PEP.0b013e3181bfae4c

Management of Myelomeningocele Study (MOMS). (2003). Retrieved July 06, 2014, from http://clinicaltrials.gov/show/NCT00060606

Melvin, E., George, T., Worley, G., Franklin, A., Mackey, J., Viles, K., … Speer, M. (2000). Genetic studies in neural tube defects . NTD Collaborative PubMed Commons. *Pediatric Neurosurgery*, *32*(1), 1–9.

Nakamura, Fabio Y.; Perandini, Luiz A.; Okuno, N. M. (2009). Construct and concurrent validation of omni– kayak rating of perceived exertion scale. *Perceptual and Motor Skills*, *108*, 744–758. doi:10.2466/PMS.108.3.744-758

National Health and Nutrition Examination Survey. (2014). Retrieved July 09, 2014, from http://www.cdc.gov/nchs/nhanes.htm

National Institute of Neurological Disorders and Stroke. (2014). Spina Bifida Facts Sheet. *NIH publication*. Retrieved from http://www.ninds.nih.gov/disorders/spina_bifida/detail_spina_bifida.htm

Nelson, M. D., Widman, L. M., Abresch, R. T., Stanhope, K., Havel, P. J., Styne, D. M., & McDonald, C. M. (2007). Metabolic syndrome in adolescents with spinal cord dysfunction. *The Journal of Spinal Cord Medicine*, *30 Suppl 1*(7), S127–39. Retrieved from

http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=2031992&tool=pmcentrez&rendertype=abstract

Newton, P. O., Faro, F. D., Gollogly, S., Betz, R. R., Lenke, L. G., & Lowe, T. G. (2005). Results of Preoperative Pulmonary Function Testing of Adolescents with Idiopathic Scoliosis. *The Journal of Bone and Joint Surgery*, *87-A*(9), 1937–1946.

Özek, M. M., Cinalli, G., & Maixner, W. J. (2008). *Spina Bifida: Management and Outcome*.

Parker, S. E., Mai, C. T., Canfield, M. a, Rickard, R., Wang, Y., Meyer, R. E., … Correa, A. (2010). Updated National Birth Prevalence estimates for selected birth defects in the United States, 2004-2006. *Birth Defects Research. Part A, Clinical and Molecular Teratology*, *88*(12), 1008–16. doi:10.1002/bdra.20735

Patel, J., Walker, J. L., Talwalkar, V. R., Iwinski, H. J., & Milbrandt, T. a. (2011). Correlation of spine deformity, lung function, and seat pressure in spina bifida. *Clinical Orthopaedics and Related Research*, *469*(5), 1302–7. doi:10.1007/s11999-010-1687-8

Pierce, R. (2005). Spirometry: an essential clinical measurement. *Australian Family Physician*, *34*(7), 535–9. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/15999163

Roach, J. W., Short, B. F., & Saltzman, H. M. (2011). Adult consequences of spina bifida: a cohort study. *Clinical Orthopaedics and Related Research*, *469*(5), 1246–52. doi:10.1007/s11999-010-1594-z

Robertson, R. J. (2004). *Perceived Exertion for Practitioners: Rating Effort with the OMNI Picture System*. Champaign, IL: Human Kinectics.

Robertson, R. J., Goss, F. L., Andreacci, J. L., Dub??, J. J., Rutkowski, J. J., Snee, B. M., … Metz, K. F. (2005). Validation of the Children's OMNI RPE Scale for Stepping Exercise. *Medicine & Science in Sports & Exercise*, *37*(2), 290–298. doi:10.1249/01.MSS.0000149888.39928.9F

Robertson, R. J., Goss, F. L., Boer, N. F., Peoples, J. a., Foreman, A. J., Dabayebeh, I. M., … Thompkins, T. (2000). Children's OMNI Scale of Perceived Exertion: mixed gender and race validation. *Medicine & Science in Sports & Exercise*, *32*(2), 452–8. doi:10.1097/00005768-200002000-00029

Robertson, R. J., Goss, F. L., Dube, J., Rutkowski, J., Dupain, M., Brennan, C., & Andreacci, J. (2004). Validation of the adult OMNI scale of perceived exertion for cycle ergometer exercise. *Medicine and Science in Sports and Exercise*, *36*(1), 102–8. doi:10.1249/01.MSS.0000106169.35222.8B

Schwimmer, J. B., Burwinkle, T. M., & Varni, J. W. (2003). Health-related quality of life of severely obese children and adolescents. *JAMA : The Journal of the American Medical Association*, *289*(14), 1813–9. doi:10.1001/jama.289.14.1813

Shaw, G. M., Velie, E. M., & Schaffer, D. (1996). Risk of neural tube defect-affected pregnancies among obese women. *JAMA : The Journal of the American Medical Association*, *275*(14), 1093–6. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/8601928

Sherman, M. S., Kaplan, J. M., Effgen, S., Campbell, D., & Dold, F. (1997). Pulmonary dysfunction and reduced exercise capacity in patients with myelomeningocele. *The Journal of Pediatrics*, *131*(3), 413–8. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/9329418

Simeonsson, R. J., McMillen, J. S., & Huntington, G. S. (2002). Secondary conditions in children with disabilities: spina bifida as a case example. *Mental Retardation and Developmental Disabilities Research Reviews*, *8*(3), 198–205. doi:10.1002/mrdd.10038

Spina Bifida Association. (2014). Retrieved November 01, 2014, from http://www.spinabifidaassociation.org/site/c.evKRI7OXIoJ8H/b.8277225/k.5A79/What_is_Spina_Bifida.htm

Staggenborg, A. (2009). *RESTRICTIVE PULMONARY DISORDERS: AN OVERVIEW* (Vol. 5798). Retrieved from www.RCECS.com

Tortori-Donati, P., Rossi, A., & Cama, A. (2000). Spinal dysraphism: a review of neuroradiological features with embryological correlations and proposal for a new classification. *Neuroradiology*, *42*(7), 471–91. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/10952179

Truwit, J. (2003). Pulmonary disorders and exercise. *Clinics in Sports Medicine*, *22*(1), 161–80. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/12613092

Utter, A. C., Robertson, R. J., Nieman, D. C., & Kang, J. (2002). Children's OMNI Scale of Perceived Exertion: walking/running evaluation. *Medicine & Science in Sports & Exercise*, *34*(1), 139–144. doi:10.1097/00005768-200201000-00021

Van den Berg-Emons, H., Bussmann, J., Meyerink, H., Roebroeck, M., & Stam, H. (2003). Body fat, fitness and level of everyday physical activity in adolescents and young adults with meningomyelocele. *Journal of Rehabilitation Medicine*, *35*(6), 271–275. doi:10.1080/16501970310012400

Van der Ploeg, H. P., Streppel, K. R. M., van der Beek, A. J., van der Woude, L. H. V, Vollenbroek-Hutten, M., & van Mechelen, W. (2007). The Physical Activity Scale for Individuals with Physical Disabilities: test-retest reliability and comparison with an acceleromter. *Journal of Physical Activity & Health*, *4*(1), 96–100. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/17489011

Verhoef, M., Lurvink, M., Barf, H. a, Post, M. W. M., van Asbeck, F. W. a, Gooskens, R. H. J. M., & Prevo, a J. H. (2005). High prevalence of incontinence among young adults with

spina bifida: description, prediction and problem perception. *Spinal Cord*, *43*(6), 331–40. doi:10.1038/sj.sc.3101705

Washburn, R. a., Zhu, W., McAuley, E., Frogley, M., & Figoni, S. F. (2002). The physical activity scale for individuals with physical disabilities: Development and evaluation. *Archives of Physical Medicine and Rehabilitation*, *83*(2), 193–200. doi:10.1053/apmr.2002.27467

Wasserman, K., Hansen, J. E., Sue, D. Y., Stringer, W. W., & Whipp, B. J. (2005). *Principles of exercise testing and interpretation : including pathophysiology and clinical applications* (4th ed.). Philadelphia: Lippincott Williams & Wilkins.

Widman, L. M., McDonald, C. M., & Abresch, R. T. (2006). Effectiveness of an upper extremity exercise device integrated with computer gaming for aerobic training in adolescents with spinal cord dysfunction. *The Journal of Spinal Cord Medicine*, *29*(4), 363–70. Retrieved from http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=1864855&tool=pmcentrez&rendertype=abstract

World Health Organization. (2001). International classification of functioning, disability, and health. Geneva: Word Health Organization. Retrieved from http://www.who.int/classifications/icf/en/

Young, N. L., McCormick, A. M., Gilbert, T., Ayling-Campos, A., Burke, T., Fehlings, D., & Wedge, J. (2011). Reasons for hospital admissions among youth and young adults with cerebral palsy. *Archives of Physical Medicine and Rehabilitation*, *92*(1), 46–50. doi:10.1016/j.apmr.2010.10.002