Who is Using Non-Invasive Ventilation? A Descriptive Study Examining the Population Enrolled in a Pediatric Non-Invasive Ventilation Program

by

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ABSTRACT

The use of non-invasive ventilation (NIV) in infants and children is increasing. This thesis examined the population of infants and children enrolled in a quaternary pediatric hospital's NIV program with the goal of describing the clinical characteristics of that population over a one-year period. It used a retrospective design and involved reviewing the charts of infants and children enrolled in the NIV program at the Stollery Children's Hospital in 2017. Demographic and clinical variables were collected, along with variables related to adherence to NIV therapy. For secure data storage purposes, a Research Electronic Data Capture (REDCap) database was created which also allowed for easy data analysis. Descriptive statistics, including mean or median and standard deviations or ranges, were provided for continuous variables and frequency distributions were used to summarize all categorical data. Independent t-tests and ANOVAs were also used to compare differences between groups. Findings included a comprehensive description of the population of infants and children enrolled in the Stollery's NIV program in 2017. This provided several directions for future research as well as implications for current practitioners who work with this population as well as the families of these infants and children.

PREFACE

This thesis is an original work by Mikelle Jenise Djkowich. The research project, of which this thesis is a part, received research ethics approval from the University of Alberta Health Research Ethics Board, Project Name "Who is Using Non-Invasive Ventilation? A Descriptive Study Examining the Population Enrolled in a Pediatric Non-Invasive Ventilation Program", Pro00088498, March 7, 2019.

Study data were collected and managed using Research Electronic Data Capture (REDCap), electronic data capture tools hosted at the University of Alberta. REDCap is a secure, web-based software platform designed to support data capture for research studies, providing 1) an intuitive interface for validated data capture; 2) audit trails for tracking data manipulation and export procedures; 3) automated export procedures for seamless data downloads to common statistical packages; and 4) procedures for data integration and interoperability with external sources (Harris et al., 2009; Harris et al., 2019).

The data analyzed in this thesis was used with the permission of and is owned by Alberta Health Services (AHS).

DEDICATION

I would like to dedicate this thesis to Caleb, my husband who so patiently waited and supported me while I pursued my dreams. I could not have done this without you.

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I would like to thank Dr. Shannon Scott, my supervisor, for her support and encouragement over the last two and a half years. Her passion for nursing scholarship was evident during the time we spent together, and I will be forever grateful that she was willing to share her time and expertise with me.

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I would like to acknowledge the financial support that I received from the Alberta Registered Nurses Educational Trust, Alberta Health Services, and from the University of Alberta. Thank you for allowing me to pursue my graduate studies and complete this research.

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CHAPTER ONE

Introduction

This thesis represents the final piece of my Master of Nursing degree. This research holds a special place in my heart; I am a registered nurse who works in pediatrics and have seen many infants and children benefit from non-invasive ventilation (NIV) therapy. I hope that this thesis will contribute to the body of knowledge surrounding NIV therapy in infants and children and help increase awareness about the needs of this unique population.

Organization of the Thesis

This thesis is comprised of four chapters. The first provides some background on NIV, information on the NIV program at the Stollery Children's Hospital, an overview of the existing literature surrounding NIV therapy in infants and children, the purpose of my study, and the significance of this as a research topic. Chapter two contains a detailed description of the methodology of the descriptive study that was conducted, including the sample, data collection and analysis strategies and any ethical implications. Chapter three focuses on the results of the study, including the statistical analysis and discussion. Chapter four provides a summary of my thesis work, conclusions drawn from the research, and outlines implications for practice and future research.

Background on Non-Invasive Ventilation

NIV is currently used in the pediatric population to treat a wide variety of sleep and respiratory disorders (Castro-Codesal et al., 2018a). It has been established that NIV provides the ventilatory assistance needed for infants and children who have complex medical conditions with respiratory involvement, often avoiding the need for tracheostomy and long-term invasive ventilation, as well as avoiding suboptimal treatment and death (Amin, et al., 2014; Castro-

Codesal et al., 2018a; Chatwin, Tan, Bush, Rosenthal, & Simonds, 2015; Edwards, Hsiao, & Nixon, 2005; Jarund, Dellborg, Carlson, Lauritzen, & Ejnell, 1999; Leboulanger, et al. 2010). Ultimately, NIV maintains airway patency and improves alveolar ventilation allowing for improvement in sleep (Hull, 2014; Marcus et al., 2012). For most infants and children, NIV can ultimately be managed at home as long as appropriate supports are present (Amaddeo, Frapin, & Faroux, 2016; Amin et al., 2014; McDougall, Adderley, Wensley, & Seear, 2013), allowing for fewer hospitalizations and improved quality of life, and for some an extended life span (Chatwin et al., 2015). While home NIV therapy is an excellent option for many infants and children, it requires extensive supports to be in place. Research surrounding adherence in the pediatric population has focused on factors impacting adherence to NIV therapy (Archbold, & Parthasarathy, 2009; DiFeo, et al., 2012; Ennis et al., 2015; Jambhekar et al., 2013; Machaalani, Evans, & Waters, 2016; Marcus et al., 2005; Marcus et al., 2012; O'Donnell, Bjornson, Bohn, & Kirk, 2006; Prashad et al., 2013; Ramirez et al., 2013; Uong, Epperson, Bathon, & Jeffe, 2007) as well as the effectiveness of behavioural interventions designed to improve adherence (Harford et al., 2012; King, Xanthopoous, & Marcus, 2014; Koontz, Slifer, Cataldo, & Marcus, 2003; Slifer, et al., 2007). There are also challenges identified with the use of NIV therapy in infants and children, including side effects such as nasal congestion, eye irritation, skin breakdown, and mid-face hypoplasia (Faroux et al., 2005; Li, Riley, & Guilleminault, 2000), as well as the limitations surrounding specialized equipment available for this population (Khirani et al., 2013; Ramirez et al., 2012).

This thesis adds to the literature on the use of NIV in infants and children by describing the population that requires NIV therapy at the Stollery Children's Hospital in Edmonton, Alberta. It describes the demographics, clinical characteristics of NIV therapy, and follow-up

and adherence patterns in this population. It is my hope that by describing this unique population, further research into this area will be feasible, supported and encouraged. By understanding the needs of infants and children who require NIV therapy, the supports and resources they require for success with therapy, and the common challenges experienced in this population, long-term program planning and resource allocation will be more feasible and better supported. The ultimate goal is that this thesis research will help support the ongoing provision of excellent care to these medically complex infants and children.

Definition of Terms

Some common terms that will be used throughout this thesis are defined here:

(1) NIV is used to describe therapies where ventilatory assistance is delivered outside the airway. NIV is usually delivered via a machine connected to a mask interface that provides positive pressure into the patient's airway. Common choices of interfaces include nasal and full-face masks. For the purpose of this thesis research, NIV will be used to refer to an umbrella of therapies including continuous positive airway pressure (CPAP) and bilevel positive airway pressure (BPAP) (Castro-Codesal et al., 2018a).

(2) CPAP is a continuous pressure applied into the airway through the entire breathing cycle with the aim of keeping airway patency and lung recruitment at the end of expiration. CPAP can be provided either invasively or non-invasively, but when used in this thesis refers to the non-invasive modality. It requires that the patient breathe spontaneously and generate enough respiratory effort to provide alveolar ventilation (Castro-Codesal et al., 2018a).

(3) BPAP references the application of a higher positive pressure into the airway during inspiration to increase alveolar ventilation and a lower pressure during expiration to maintain lung recruitment and airway patency. BPAP therapy improves alveolar ventilation and carbon

dioxide removal with a reduction in respiratory effort. This type of therapy can also include a backup respiratory rate, and will provide machine-triggered breaths in the absence of a patient's breath or if breathing becomes too shallow (Essouri et al., 2005).

(4) A mask or interface refers to the equipment that rests on the infant or child's face and delivers positive airway pressure. There are a wide variety of masks available, from nasal to full-face options (Castro-Codesal, Olmstead, & MacLean, 2019). Figures 1.1-1.4 provide a visual depiction of the available types of mask interfaces.

Figure 1.1 – Nasal Mask
(Castro-Codesal et al., 2019)

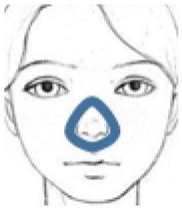


Figure 1.2 – Nasal Pillow
(Castro-Codesal et al., 2019)

Figure 1.3 – Total Face Mask
(Castro-Codesal et al., 2019)

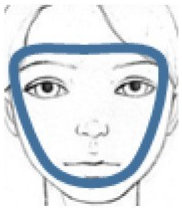
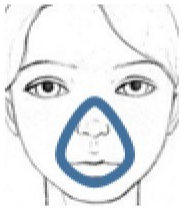


Figure 1.4 – Oro-Nasal Mask
(Castro-Codesal et al., 2019)



(5) Headgear refers to the straps used to keep the NIV mask in place. Typically, headgear involves a combination of head or chins traps designed to minimize discomfort and optimize placement of the NIV equipment (Castro-Codesal et al., 2019; Slifer et al., 2007).

(6) Adherence is frequently used within this thesis to refer to the usage of NIV therapy by infants and children. Definitions of adherence vary in the literature, but for the purposes of this thesis acceptable adherence is the use of NIV for at least for hours a night (Machaalani et al., 2016).

The Non-Invasive Ventilation Program at the Stollery Children's Hospital

The Stollery Children's Hospital is a quaternary pediatric hospital located in Edmonton, Alberta with a large catchment area including Northern Alberta, Northern British Columbia (BC), Northern Saskatchewan, the North West Territories (NWT), Nunavut and the Yukon. Some of the children enrolled in the Stollery's NIV program are likely from geographic locations outside of the Edmonton area.

The NIV program at the Stollery is comprehensive in nature. It includes a multidisciplinary team that works with each infant and child who requires NIV therapy. This multidisciplinary group includes a nurse practitioner, two respirologists and a respiratory therapist who consult with other specialties including occupational therapists, social workers, and child life specialists to provide care to this population. The Stollery's program provides a unique service to help ensure that NIV therapy is as effective as possible. This is the only program with government-funded sleep laboratory services, which allows for diagnostic polysomnography studies for characterization of sleep-related breathing disorders to be run. Infants and children enrolled in the NIV program also have access to titration polysomnography studies that allow for titration of ventilator settings to ensure optimal treatment of their respiratory abnormalities. They are followed closely once NIV is initiated, with periodic assessment of their underlying respiratory disease, overall health status, NIV equipment and overall adaptation to therapy. The program also offers custom adaptation of the headgear that holds the mask as well as optimization of mask fit for each infant or child who requires NIV. This adaptation is often done as an attempt to reduce side effects, such as air leak and skin breakdown; these side effects can negatively impact adherence, with the goal of customization being to improve adherence. Occupational therapists and therapy assistants are trained to

perform headgear adaptation, as well as to optimize mask fit. This may include things like custom straps to assist with fit of the equipment, and gel pads to offload pressure areas. Child life specialists are also involved and will assist with strategies to help desensitize the infant or child to the headgear and mask.

The Stollery's NIV program is an outpatient program. The preferred location for NIV initiation is as an outpatient. Infants and children who are started on NIV during a hospital admission are transitioned to the NIV program at the time of discharge. Regardless of where NIV is initiated, infants and children are followed as outpatients in the NIV clinic for the duration of their therapy or until they transition into adult care or medical care at other centres or in other provinces. Follow-up visits happen in the outpatient NIV clinic and are overseen by a nurse practitioner. At each clinic visit, any issues experienced with therapy are discussed with the team as well as the infant or child's respiratory and overall health status. This may include things like side effects, issues with mask or headgear fit, reappearance of respiratory and non-respiratory symptoms, and if the infant or child is adhering to the therapy. If necessary, additional modifications are made to the NIV equipment at each visit. Adherence information is gathered and tracked using objective data downloaded from the NIV machines which is stored digitally. This data can be accessed remotely by practitioners prior to follow-up visits, allowing the identification of any problems as well as individualized teaching and education to be provided at those visits.

Literature Review

Non-Invasive Ventilation Use in Infants and Children

The use of NIV to treat a variety of respiratory conditions in infants and children is increasing. Amin et al. (2014) examined the pediatric population in a Canadian NIV program

over a period of twenty years and found that the number of infants and children enrolled grew exponentially, from two to 156. This suggests that the use of NIV in infants and children is becoming more widely used and accepted, with better supports in place for this population. In the province of Alberta, Castro-Codesal et al. (2018b) examined longitudinal trends in NIV use in infants and children. They found that NIV incidence steadily increased over a ten-year period, from less than two children per 100 000 started on NIV between 2005-2008, to eight per 100 000 started on NIV between 2011-2014. The indications for NIV therapy in infants and children will be examined in the next section.

Indications for non-invasive ventilation. NIV is used for a wide range of respiratory disorders in infants and children. Research surrounding indications for NIV therapy has focused mainly on obstructive sleep apnea and spinal muscular atrophy, with other conditions being less well studied (Castro-Codesal et al., 2018a; Chatwin et al., 2015). NIV has been successfully used in infants and children with obstructive sleep apnea, particularly those who are also obese as they typically do not respond as well to tonsillectomy and adenoidectomy (Downey, Perkin, & MacQuarrie, 2000; Jarund et al., 1999; Kuhle, Urschitz, Eitner, & Poets, 2009; Leonardis, Robinson, & Otteson, 2013; Marcus et al., 2012; Sudarsan, Paramasivan, Arumugam, Murali, & Kameswaran, 2014). It has also been used to improve morbidity in infants and children with chronic lung disease such as cystic fibrosis (Armstrong, 2013; Faroux, Boffa, Desguerre, Estournet, & Trang, 2003; Flight et al., 2012; Moran, Bradley, & Piper, 2017). Neuromuscular diseases that result in weakened respiratory muscles, including muscular dystrophy and spinal muscular atrophy, are also an indication for NIV therapy (Chatwin, Bush, & Simonds, 2011; Faroux et al., 2003; Simonds, 2003, 2006). Infants and children may experience nocturnal hypoventilation that requires treatment due to other conditions, including conditions secondary to

cancer (Rosen, & Brand, 2011), metabolic disorders (Nakra, Bhargava, Dzuira, Cario, & Bazzy-Assand, 2008; Nashed et al., 2009), achondroplasia (Julliand et al., 2012), trisomy 21 (Brooks et al., 2015), attention-deficit hyperactivity disorder (Johnstone, Tardiff, Barry, & Sands, 2001), and other disorders of the central nervous system (CNS) affecting the drive to breath (Grychtol, & Chan, 2018; Markstrom, Sundell, Stenberg, & Katz-Salamon, 2008; Ramesh, Boit, & Samuels, 2008), which can also be treated using NIV. NIV has also been used to treat pulmonary hypertension, a complication often related to congenital heart disease in infants and children, although the evidence in the pediatric population is very limited (Bunn, Roberts, & Thomson, 2004). Ultimately, it is agreed that the use of NIV in infants and children can be a successful alternative to more aggressive forms of treatment, including surgical intervention, tracheotomy or intubation and invasive mechanical ventilation (Amin et al., 2014; Castro-Codesal et al., 2018a; Chatwin et al., 2015; Edwards et al., 2005; Hull, 2014; Jarund et al., 1999; Leboulanger et al., 2010). At one Canadian centre, 17% of infants and children required invasive ventilation, compared to 83% being successfully managed with NIV (Amin et al., 2014). The successful use of NIV is also linked to a decrease in morbidity and mortality in this population (Amaddeo et al., 2016; Amin et al., 2014; Castro-Codesal et al., 2018a,b; Chatwin et al., 2015; Edwards et al., 2005; Hull, 2014; McDougall et al., 2013; Wallis, Paton, Beaton, & Jardine, 2011). A decrease in morbidity suggests that the use of NIV therapy may improve the quality of life in infants and children with a variety of medical conditions, reducing the number of hospitalizations they experience and allowing them to spend more time at home (Chatwin et al., 2015). In order to successfully care for infants and children who require NIV therapy at home, the coordination of specialized supports and services is required.

Non-invasive ventilation at home. Long-term use of NIV in chronically ill infants and children has resulted in a re-examination of where care is provided and has caused a shift in care setting, from the hospital to home (Amaddeo et al., 2016; Amin et al., 2014; Chatwin et al., 2015; Edwards et al., 2005; Hull, 2014; McDougall et al., 2013; Wallis et al., 2011). Guttmann, Cohen, and Moore (2009) suggest that appropriate health human resource planning is required, especially for infants and children with chronic, complex medical needs; it is also important to recognize that as the pediatric population requiring NIV expands, additional outpatient resources and caregiver training will be required (Edwards et al., 2005). Infants and children requiring NIV need caregivers that are competent and actively involved in their care; the responsibility of caring for this population at home should not be minimized. Caregiver burden has been identified as a significant issue that is associated with this population. Families with medically complex infants and children experience social and financial hardships, as well as sleep disturbances due to the amount of around-the-clock care they require (Keilty, Cohen, Spalding, Pullenayegum & Stremler, 2018; Thomson et al., 2016). There is agreement that infants and children requiring NIV need an experienced and comprehensive multidisciplinary team that can actively follow their care and provide support (Edwards et al., 2005; Ennis et al., 2015; Jambhekar et al., 2013; Machaalani et al., 2016; McDougall, et al., 2013; O'Donnell et al., 2006; Ramirez et al., 2013; Wallis et al., 2011). This includes extensive homecare and respite support, as well as specialists and a multidisciplinary team that is easily accessible, which may present a challenge depending on geographic location and available resources (Edwards et al., 2005; McDougall et al., 2013). An additional complicating factor is the fact that as children who require NIV therapy age, they will eventually have to transition from a pediatric to an adult NIV program.

Transition to adult services. The transition from pediatric to adult care has also been examined (Amin et al., 2014; Castro-Codesal et al., 2018a,b; Chatwin et al., 2015; McDougall et al., 2013; Wallis et al., 2011), with an increased number of children making this transition as a result of the increased life-expectancy of many complex medical conditions due to successful treatment with NIV. Castro-Codesal et al. (2018b) examined the transition from pediatric to adult care in the province of Alberta and found that the number of children making this transition continues to rise. From 2005-2008, 10 per 100 000 on NIV made the transition to adult services compared to 28 per 100 000 in 2011-2014. This increase provides an additional challenge, demanding additional resources and ongoing support from the healthcare system. The transition from pediatric to adult services can be difficult for patients and their families, especially as patients take on a more independent role in the decision-making process. Families may benefit from a transition program to ease this process and minimize health risks during this critical time period. Additionally, adult practitioners may require extra education to become more familiar with childhood disorders and the care that they require (McDougall et al., 2013).

Adherence to Non-Invasive Ventilation

Adherence to NIV therapy has widely been examined in the pediatric population (Archbold, & Parthasarathy, 2009; DiFeo et al., 2012; Ennis et al., 2015; Harford et al., 2012; Jambhekar et al., 2013; King et al., 2014; Koontz et al., 2003; Machaalani et al., 2016; Marcus et al., 2005; Marcus et al., 2012a; O'Donnell et al., 2006; Prashad et al., 2013; Ramirez et al., 2013; Slifer et al., 2007; Uong et al., 2007), with some evidence suggesting that adherence rates might be better than those seen in the adult population (Ramirez et al., 2013). Acceptable adherence is often defined as usage of NIV for at least four hours per night, with variable numbers on how many nights per week should meet this threshold ranging from three to seven (Archbold, &

Parthasarathy, 2009; Ennis et al., 2015; Machaalani et al., 2016; Marcus et al., 2005; O'Donnell et al., 2006; Uong et al., 2007). These criteria are somewhat arbitrary and, at the best, adult-based (Archbold, & Parthasarathy, 2009; Ennis et al., 2015; Machaalani et al., 2016; Marcus et al., 2005; Marcus et al., 2012a; O'Donnell et al., 2006; Prashad et al., 2013; Uong et al., 2007), considering the amount of recommended sleep in children greatly changes over time. However, this information is provided by the NIV machine via downloads and is often used in the pediatric population due to the lack of a better definition for adherence to NIV therapy in infants and children. Early adherence patterns can be used to predict long-term adherence to NIV therapy (Archbold, & Parthasarathy, 2009), and therefore can provide valuable information to practitioners working with this population. Understanding of patterns of NIV use can promote an open discussion with the infant or child and their family surrounding any potential barriers to NIV therapy and adherence, as well as proactive solutions to this issue. The quality of the evidence and the data available vary, but there is agreement that certain factors influence the likelihood of adherence to NIV therapy. Specifically, fewer side effects (Amaddeo et al., 2016; Ennis et al., 2015; King et al., 2014; Uong et al., 2007), good mask fit (Ennis et al., 2015; Hull, 2014; Li et al., 2000; Marcus et al., 2005; O'Donnell et al., 2006; Ramirez et al., 2012), involvement and competency of parents (DiFeo et al., 2012; Edwards et al., 2005; King et al., 2014; Koontz et al., 2003; Machaalani et al., 2016; O'Donnell et al., 2006; Prashad et al., 2013), and excellent coordination of specialized care (Edwards et al., 2005; Ennis et al., 2015; Jambhekar et al., 2013; Machaalani et al., 2016; O'Donnell et al., 2006; Ramirez et al., 2013) are predictors of improved adherence to NIV therapy in infants and children.

Behavioural interventions focused on helping children get used to the NIV equipment, adjust NIV settings, and creating a new routine that includes the use of NIV therapy have also

been examined in the context of adherence, with some improvement noted post-intervention (Harford et al., 2012; King et al., 2014; Koontz et al., 2003; Slifer et al., 2007). All of these programs utilized a multidisciplinary approach, reinforcing the need for an integrated care team to manage the complex nature and care needs of this population. Although available information about adherence to NIV therapy varies, evidence suggests that early interventions that help to minimize complications and improve adherence are an important part of managing NIV therapy in infants and children.

Complications Related to Non-Invasive Ventilation

The presence of NIV side effects can be disturbing and have been shown to decrease adherence among infants and children requiring NIV therapy. Side effects related to NIV therapy may be seen immediately, or they may develop over time. Immediate side effects may include eye irritation (Amaddeo et al., 2016; Amin et al., 2016; Hull, 2014; Marcus et al., 2012b), nasal congestion (Amaddeo et al., 2016; Amin et al., 2016; Marcus et al., 2005; Marcus et al., 2012b; Pavone, Verillo, Calderelli, Ullmann, & Cutrera, 2013), and gastric insufflation (Amin et al., 2016; Pavone et al., 2013). Long-term side effects may include the development of midface hypoplasia (Amaddeo et al., 2016; Amin, Al-Saleh, & Narang, 2016; Faroux et al., 2005; Li et al., 2000; Pavone et al., 2013), and skin breakdown (Amaddeo et al., 2016; Amin et al., 2016; Faroux et al., 2005; Hull, 2014; Marcus et al., 2012b; Ramesh et al., 2008; Ramirez et al., 2012). Skin breakdown and eye irritation are two of the most common side effects of NIV therapy and are typically are related to poor mask fit (Amaddeo, et al., 2016; Amin et al., 2016; Faroux et al., 2003; Hull, 2014; Ramesh et al., 2008); therefore, it can be argued that if equipment fit is improved, the potential for side effects is decreased, potentially resulting in increased adherence in infants and children requiring NIV therapy.

Fitting NIV equipment, including headgear and masks, for infants and children can prove to be especially challenging, as most equipment is designed for the adult population. Currently, the type of mask chosen often depends on the age of the child and the type of ventilatory support required (Amin et al., 2016; Castro-Codesal et al., 2019; Ramirez et al., 2012), with changes made later on if necessary. Available mask interface categories include nasal and full-face, with a nasal mask being preferred. Nasal masks are favoured, as they allow the child to communicate, minimize the risk of aspiration, and allow for secretion management if necessary (Castro-Codesal et al., 2019). Full-face masks provide an option for children who do not sleep with their mouths closed, but present additional challenges, including an increased aspiration risk if the infant or child cannot remove the mask on their own (Ramirez et al., 2012). There is discussion about customization of mask interfaces for infants and children, but options so far are limited and not widely used (Castro-Codesal et al., 2019; Lanza et al., 2019). One study (Slifer et al., 2007) specifically identified poor mask fit as an issue and suggested that mask and headgear modifications might be beneficial for children requiring NIV therapy.

Summary

The use of NIV in medically complex infants and children is increasing. There are many benefits to this, including a decrease in morbidity and mortality as well as improved quality of life. However, the provision of this therapy also has its challenges. Adherence to NIV can present a challenge, particularly if side effects from the therapy are present. In order for infants and children to benefit from NIV therapy, the coordinated provision of specialized care from a multidisciplinary team is required.

Purpose of the Study

This descriptive study was designed to characterize the population of infants and children seen at the Stollery Children's Hospital that require NIV support. It aims to answer the following research question: What are the clinical characteristics of a population of infants and children enrolled during a one-year period in a NIV program at a quaternary pediatric hospital? This was achieved through descriptive analysis of the demographic and clinical characteristics, follow-up practices and adherence patterns of each infant and child enrolled in the Stollery's NIV program for a one-year period post NIV initiation. Between groups analyses were also done to further illuminate the relationships between various clinical characteristics with the goal being to add another layer to the analysis in order to better characterize and understand this unique population.

Significance of the Research

Results from this descriptive study will be used to inform future research surrounding NIV therapy at the Stollery. The existence of a database specifically designed to capture the pediatric population enrolled in the NIV program at the Stollery will allow for additional data collection and research to be undertaken quickly. The information gathered from this study will be used to educate and inform healthcare professionals who work with the pediatric population requiring NIV therapy, as well as families who care for these infants and children at home. In order for practitioners to provide excellent care to infants and children requiring NIV therapy, a solid understanding of this population, including the indications for and challenges with NIV therapy is required. For example, by understanding common indications for NIV therapy as well as for discontinuation of therapy, practitioners will be able to answer common questions from families about duration of therapy and help manage long-term expectations. Examining and describing adherence in this population will be particularly important, providing insight into

what factors may impact adherence to NIV therapy in infants and children, creating the potential to improve outcomes in this population. For example, if a large number of infants and children are discontinuing therapy due to non-compliance or non-tolerance, additional supports or training may need to be built into the NIV program to help ensure long-term success of therapy. The data from this study will also be able to inform resource allocation and future program planning, ensuring that this population continues to be properly supported. The goal of sharing this knowledge is that additional research into this area will be supported, and that these infants and children will receive improved quality of care.

Summary

This chapter has included background information on NIV therapy in infants and children, as well as some information about the NIV program at the Stollery Children's Hospital. It also included a literature review on NIV in infants and children in order to situate my thesis research within the existing literature. It concluded by presenting the purpose of my thesis research and its significance for this population. The following chapter contains a detailed explanation of the methodology of my thesis work.

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CHAPTER TWO

Methodology

Study Design

This was a single-centre, retrospective study conducted through a chart review of all infants and children initially enrolled in the NIV program at the Stollery Children's Hospital during 2017. The year 2017 was chosen for a time frame as it provided current and complete data in an attempt to accurately reflect the composition of infants and children enrolled in the Stollery's NIV program.

Ethics approval was obtained from the Health Ethics Research Board at the University of Alberta prior to data collection and analysis occurring. Operational approval was also obtained from Alberta Health Services (AHS) in order to access patient information through the electronic medial record (EMR) and to use it in this study. All research work that involved data collection and analysis was done onsite at the Stollery, on a secure, password protected computer in a locked office. All patient data was uploaded and securely stored in a Research Electronic Data Capture (REDCap) database specifically created for this project and was never accessed remotely. There was minimal risk involved as all patient data was de-identified prior to analysis.

Population

The sample included children who met the following criteria:

(1) Zero to eighteen years of age at the time of NIV initiation (the Stollery currently treats patients until they turn eighteen)

(2) Documented initiation of NIV use in 2017 (including CPAP and BPAP) and follow-up through the NIV program at the Stollery Children's Hospital

(3) Available adherence data from machine downloads. Adherence was captured at regular intervals as outlined in the data collection section

Infants and children were not excluded if they did not have a follow-up visit or adherence download available during each interval, as it was recognized that follow-up requirements for each infant or child may differ based on the success, failure, or complications of NIV therapy. If they did not have any follow-up or adherence data available, they were excluded from that specific portion of the analysis.

Data Collection

Data was collected from the charts of infants and children enrolled in the Stollery Children's Hospital NIV therapy program during 2017 and their subsequent follow-up visits for a one-year period post NIV initiation. This captured one year's worth of follow-up and adherence data for all infants and children enrolled in the Stollery's NIV therapy program in 2017.

Demographics and clinical variables. Data captured from the NIV clinic visits included the following information for the infants and children enrolled in the NIV program at the Stollery in 2017: Personal identifying information, date of birth, date of NIV initiation, date of clinic visit, type of NIV therapy (CPAP or BPAP), type of mask used (nasal or full-face), if custom headgear adaptation was performed, complications from the therapy including eye irritation, nasal congestion, and skin breakdown, and concerns with equipment fit, including mask fit concerns, mask leak concerns, and issues with headgear fit.

Further demographic and clinical information were extracted directly from the electronic medical record of each infant or child, including sex, geographic location, place of NIV initiation, if NIV was discontinued and the reason for discontinuation, type of clinic visit (zero to

three month, three to six month, six to nine month, or nine to twelve month), as well as the underlying diagnosis leading to NIV initiation. In order to determine geographic location, postal codes were used to identify infants and children who were from the Edmonton metro area, those from other areas in the province of Alberta and those from the neighbouring provinces and territories that the Stollery serves, including BC, Saskatchewan, Yukon, the NWT and Nunavut. The reason for NIV initiation included broad disease categories that encompassed the majority of reasons for NIV therapy. These categories included CNS disorders (ex. cerebral palsy, congenital hypoventilation, metabolic disorders), musculoskeletal disorders (ex. muscular dystrophy, spinal muscular atrophy, achondroplasia), upper airway obstruction caused by underlying disorders (ex. obesity, airway abnormalities, Trisomy 21, craniosynostosis), pulmonary disease (ex. cystic fibrosis), cardiac disease (ex. pulmonary hypertension, congenital heart disease), and cancer (ex. brainstem tumors). The reasons for discontinuing NIV therapy were categorized into groups as well, including non-compliance, death, transfer (to an adult program or to another site), escalation of therapy like tracheostomy or intubation, and improvement.

Follow-up and adherence variables. Follow-up data were collected from the electronic medical records of the NIV clinic visits at the Stollery, where a standard flowsheet was used. In-person follow-up visits for a one-year period post NIV initiation were included for each infant and child. For the purpose of this thesis, phone follow-up visits were not tracked, as documentation of phone visits differed based on the practitioner conducting the visit, making standard data collection difficult and beyond the scope of this project. Throughout the one-year period, intervals to capture follow-up data were chosen based on available information and standard follow-up procedures and scheduling at the NIV clinic including zero to three months, three to six months, six to nine months and nine to twelve months.

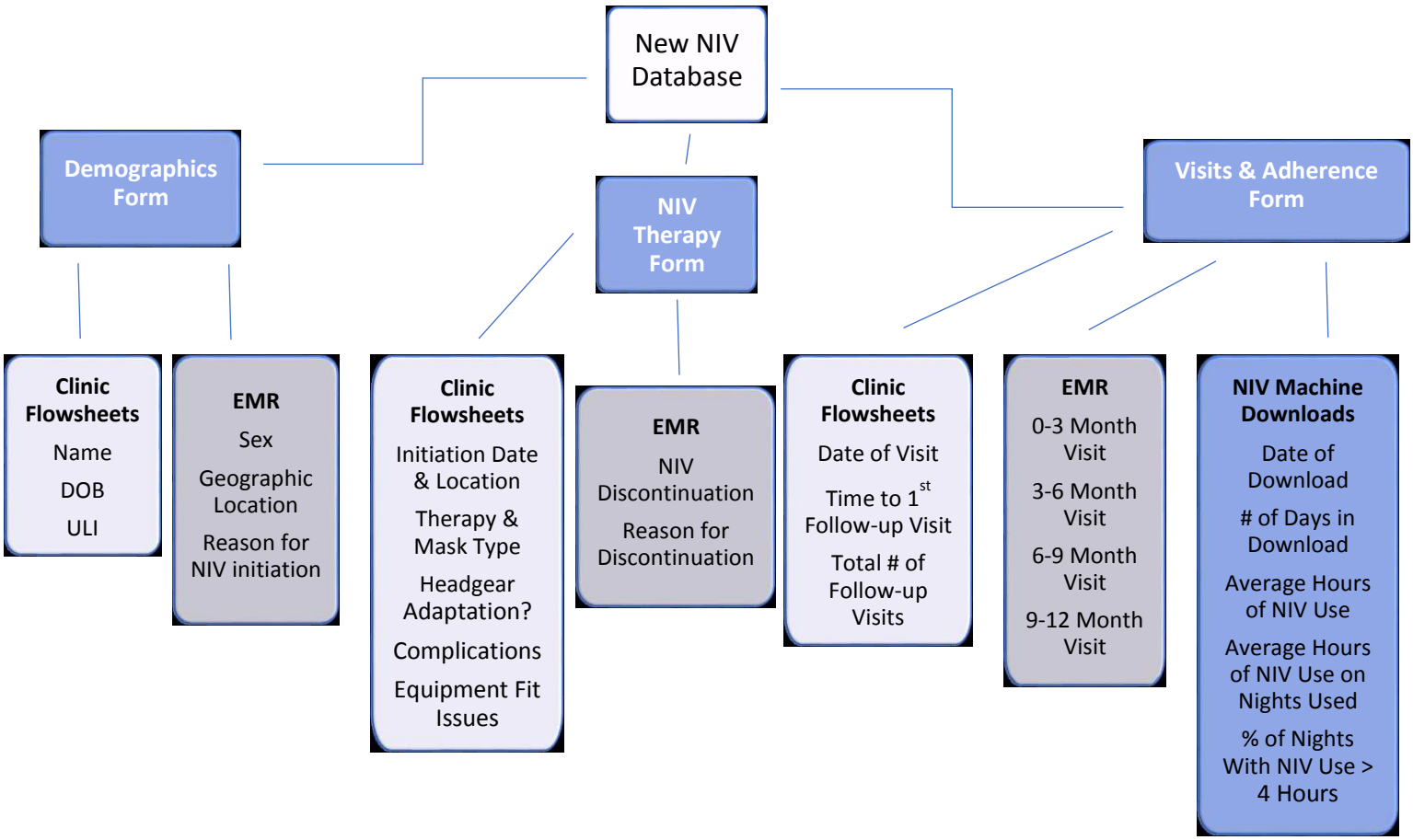
Objective adherence data were extracted from the NIV machine downloads to encompass a one-year period following NIV initiation for each infant and child. Adherence data was made available through NIV machine downloads which were found in a separate section of the EMR. This data included the date of the machine download, the number of days included in the download, the average number of hours of NIV use per night, the average number of hours of NIV on nights it was used, and the percentage of nights with NIV use greater than four hours. These downloads typically included adherence data from a total of thirty days, however the number of days was not consistent across the entire sample. Adherence to NIV therapy was examined at the same intervals as follow-up data. If more than one download was available during the interval, the one closest to the date of the follow-up visit was chosen. Second, overall mean adherence data was also calculated to describe adherence throughout the entire first year of enrollment in the NIV program for each infant and child who had adherence download data available. This variable was adjusted based on the number of adherence downloads available for analysis for each infant or child.

Data Storage and Database Creation

A pre-existing spreadsheet was made available through the information technology department at the Stollery Children's Hospital who compiled data from the clinic visits of infants and children who had NIV therapy initiated through the program from 2013-2018. Those who were initiated onto NIV therapy in 2017 were separated from the of the larger data set into a smaller spreadsheet, referred to as the NIV clinic flowsheet. This spreadsheet was stored on one computer, which was password protected, backed up to a secure offsite location and located in a locked office at the Stollery Children's Hospital. I then created a REDCap database that was designed specifically for this thesis research that allowed variables from different sources to be

securely stored in one location. The database was designed to be user friendly, with the hope that it would be utilized by the NIV program at the Stollery for ongoing research into this area. The data from the pre-existing spreadsheet was manually uploaded into REDCap and combined with other data obtained from the EMR and NIV machine downloads of each infant or child to create a comprehensive database. This simplified access to the data and allowed for easier data export and analysis. The database went through several revisions to ensure that the all applicable variables were included and entered in a way that allowed for data to be analyzed appropriately. The REDCap database that was created consisted of three forms: Demographics, NIV therapy, and visits and adherence data. These are located in the appendices at the end of the thesis. Figure 2.1 provides a visual overview of the database creation.

Figure 2.1 – Visual Depiction of REDCap Database Creation



Outcomes of Interest

The variables that were analyzed were chosen to provide a thorough understanding of the population enrolled in the Stollery's NIV program in 2017. These included sex, age at NIV initiation, geographic location, reason for NIV initiation, NIV initiation location, therapy and mask type, reported complications and issues with equipment fit, if custom headgear adaptation occurred, if NIV was discontinued and the reason for discontinuation. The time from NIV initiation to first follow-up visit and the number of follow-up visits within the first year of enrollment were also examined. Variables related to adherence to NIV therapy were also examined, including the average hours of NIV use, the average hours of NIV use on nights used and the percentage of nights with NIV use over four hours.

Data Analysis

Descriptive analysis of the data occurred using SPSS version 25. Descriptive statistics, including mean or median and standard deviations or ranges, were provided for continuous variables and frequency distributions were used to summarize all categorical data. Independent t-tests and ANOVAs were used to compare differences between groups. An alpha level of 0.05 was used for all statistical tests.

Independent t-tests were conducted to compare the total average hours of NIV use, the total average hours of NIV use on nights used and the total average percentage of nights with NIV use greater than four hours in infants and children who had custom headgear adaptation done versus those who did not, who had NIV initiated as an inpatient versus those who had it initiated as an outpatient, those who used CPAP versus BPAP therapy, those who used nasal masks versus full-face masks, those who reported complications with NIV therapy and those who did not, and those who reported a concern with equipment fit versus those who did not.

One-way ANOVAs were also conducted for between groups analysis. One-way ANOVAs were conducted to compare the effect of age at NIV initiation and the effect of reason for NIV discontinuation on the total average hours of NIV use, the total average hours of NIV use on days used and the total average percentage of nights with NIV use greater than four hours.

Summary

This chapter included a detailed overview of the methodology used in my thesis research. It provided the sample characteristics, data collection procedures and explained the creation of the REDCap database for this project. It concluded by outlining the plan for data analysis using the data that was collected. The following chapter contains the results of the study that was conducted.

CHAPTER THREE

Results

Demographic and Clinical Characteristics

During 2017, a total of 54 infants and children were initiated on NIV through the program at the Stollery Children's Hospital. 29 (54%) of them were male. 34 (63%) of the infants and children resided in the Edmonton metro area, with 17 (32%) living elsewhere in the province of Alberta, and three (6%) living outside of the province. NIV initiation happened at 6.6 years on average (SD = 5.4). 16 (29%) initiations occurred as infants, eight (16%) occurred during the pre-school age, 26 (48%) occurred between the ages of five and 15, and four (7%) occurred over the age of 15.

The underlying condition leading to NIV initiation was also examined, with 36 (67%) of the cases being due to upper airway obstruction. CNS disorders accounted for the second largest number of NIV initiations, with eight (15%). The most common age for initiation due to upper airway obstruction was between 10 and 15 years, with 10 (28%) of 36 initiations happening at this time. Initiation for upper airway obstruction was also common in infants, with nine (25%) of 36 initiations occurring in those less than six months old and five (14%) happening between six months and one year. Initiation for CNS disorders occurred most commonly between one and five years old, with four (50%) of eight initiations happening at this time.

While the majority of the infants and children initiated on NIV continued using the therapy during the study period, 11 (20%) of the 54 infants and children who started NIV therapy in 2017 also discontinued NIV therapy within one year. The reasons for NIV discontinuation varied. Improvement in the underlying condition to the point NIV was no longer required occurred in five out of 11 (46%) cases, death within the year accounted for three out of 11 (27%)

cases, a change to invasive ventilation occurred in one (9%) case of 11, and two out of 11 (18%) cases were transferred to other NIV programs. Infants and children who required NIV therapy because of upper airway obstruction were more likely to discontinue therapy within the year, with six (55%) of the 11 discontinuations falling into this category. The highest number of discontinuations happened in infants, with four (36%) of those who discontinued NIV during the year falling into this age category. The remainder of age categories did not appear to have any association between age at NIV initiation and NIV discontinuation, with three (27%) being pre-school age, two (18%) being between five and 15 years, and two (18%) being over 15 years. Table 2.1 provides an overview of the demographic and clinical characteristics of this population of infants and children.

Table 2.1 – Demographic and Clinical Characteristics of Infants and Children Enrolled in the NIV Program

Characteristic	*Total*	*Percentage*
Sex	*54*	*100*
Male	29	53.7
Female	25	46.3
Location	*54*	*100*
Edmonton Metro	34	63
Outside Edmonton	17	31.5
BC	0	0
Saskatchewan	1	1.9
NWT	2	3.7
Yukon	0	0
Nunavut	0	0
Age at Initiation	*54*	*100*
<6 months	12	22.2
6 months - <1 year	4	7.4
1 - <5 years	8	14.8
5 - <10 years	11	20.4
10 - <15 years	15	27.8
>15 years	4	7.4
Reason for NIV Initiation	*54*	*100*
CNS disorder	8	14.8
Musculoskeletal	3	5.6
Upper airway	36	66.7
Pulmonary disease	5	9.3
Cardiac disease	1	1.9
Cancer	1	1.9
Discontinuation	*54*	*100*
Yes	11	20.4
No	43	79.6
Reason for Discontinuation	*11*	*100*
Deceased	3	27.3
Transfer	2	18.2
Escalation	1	9.1
Improvement	5	45.5
Non-compliance	0	0

The characteristics of the type of NIV therapy at the Stollery Children's Hospital were also examined. 36 (67%) of the new NIV initiations in 2017 occurred in an outpatient setting.

CPAP therapy was used with 46 (85%) initiations, and nasal masks were used with the majority, 50 (92%), of infants and children. CPAP therapy was most commonly used with infants and children who required NIV therapy due to upper airway obstruction, with 34 (94%) of these cases using CPAP. 29 (54%) infants or children had custom headgear adaptation performed at one of their follow-up visits. Table 2.2 provides an overview of the characteristics of NIV therapy for infants and children enrolled in the Stollery's NIV program in 2017.

Table 2.2 – Characteristics of NIV for Infants and Children Enrolled in the NIV Program

Characteristic	*Total*	*Percentage*
Type of Therapy	*54*	*100*
CPAP	46	85.2
BPAP	8	14.8
Type of Mask	*54*	*100*
Nasal	50	92.6
Full face	4	7.4
Initiation Location	*54*	*100*
Outpatient	36	66.7
Inpatient	17	31.4
Not Documented	1	1.9

Complications	*54*	*100*
Yes	28	51.9
No	26	48.1
Concern with Equipment Fit	*54*	*100*
Yes	28	51.9
No	26	48.1
Headgear Adapt	*54*	*100*
Yes	29	53.7
No	25	46.3

Follow-Up Practices

There was a total of 100 in-person NIV follow-up clinic visits for 54 infants and children who were enrolled in the NIV program at the Stollery during 2017. One infant or child who had NIV therapy initiated 2017 did not have any documented in-person follow-up visits with the NIV clinic after enrollment in the program; this infant or child was booked for several follow-up appointments but failed to attend any of them. The number of in-person follow-up visits per infant and child within the first year of enrollment in the NIV program varied; 23 (43%) had one, with only three (6%) having four visits within the first year of NIV therapy. The majority of these visits happened in the first six months after initiation of NIV therapy, with 30 (30%) happening between zero and three months and 31 (31%) between three to six months. The mean number of days from the initiation of NIV to the infant or child's first follow-up visit was 95

days (SD = 74). Table 2.3 provides an overview of the follow-up practices at the Stollery in 2017.

Table 2.3 – Frequency of Clinical Multidisciplinary Follow-up for Infants and Children Enrolled in the NIV Program

Characteristic	*Total # Follow-up Visits*	*Percentage*
Clinic Visit	*100*	*100*
0-3 months	30	30
3-6 months	31	31
6-9 months	19	19
9-12 months	20	20

# Follow-up Visits Per Infant/Child	*54*	*100*
Zero	1	1.9
One	23	42.6
Two	16	29.6
Three	11	20.4
Four	3	5.6

At these follow-up clinic visits, 28 (52%) reported at least one NIV-related complication within the first year from NIV initiation, and nine of those (32%) had more than one; nine (17%) had eye irritation from air leak, 13 (24%) had persistent nasal congestion and 17 (31%) suffered from skin breakdown. Similarly, 28 (52%) reported at least one concern related to mask interface, and 14 of those (50%) reported more than one; there were 21 (39%) complaints of an issue with mask fit, eight (15%) complaints of an issue with mask leak, and 16 (30%) complaints related to headgear fit.

Adherence Data

There were 90 adherence downloads from NIV machines available for analysis for the 2017 year, with an average of 1.6 (SD = 0.96) downloads per patient. Three infants or children had no adherence information available for analysis at any of the intervals. The average number of reported days was 30 (SD = 19). The average total hours of NIV use, the average hours of NIV use on nights used and the percentage of nights with NIV use greater than four hours were examined at each follow-up interval for the infants and children enrolled in the NIV program at

the Stollery Children's Hospital in 2017 who had adherence data available. Table 2.4 contains the results of this analysis.

Table 2.4 – Mean Adherence Data for Each Follow-Up Interval

Time from NIV Initiation	*Mean Hours Used (SD)*	*Mean Hours Used on Nights Used (SD)*	*Mean % Nights with Use >4 Hours (SD)*
0-3 months	5 (3.9)	6 (3.8)	52 (39)
3-6 months	5 (3.6)	6 (3.7)	52 (40)
6-9 months	4 (2.7)	6 (3)	46 (33)
9-12 months	5 (3)	7 (2.9)	58 (34)

The total average adherence across the year for all the infants and children enrolled in the Stollery's NIV program was also examined. The total mean for average hours of use was five hours (SD = 3.5), the total mean for average hours of NIV use on nights used was six hours (SD = 3.6) and the total mean for percentage of nights with NIV use greater than four hours was 54 percent (SD = 37). Figures 3.1, 3.2 and 3.3 provide histograms that describe the total averages over the first year of enrollment in the Stollery's NIV program for each adherence variable.

Figure 3.1 – Proportion of Infants and Children Using NIV by Number of Hours of NIV Use (All Nights)

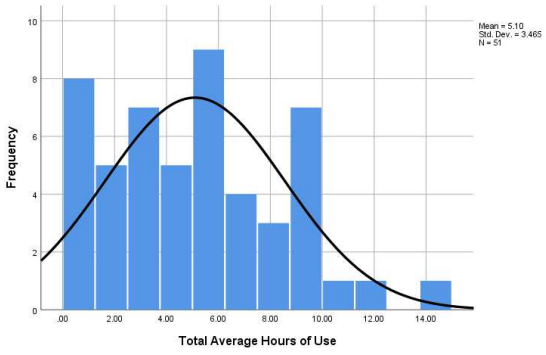


Figure 3.2 – Proportion of Infants and Children Using NIV by Number of Hours of NIV Use (Nights Used)

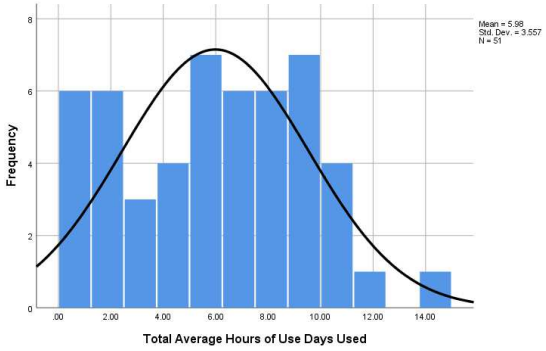
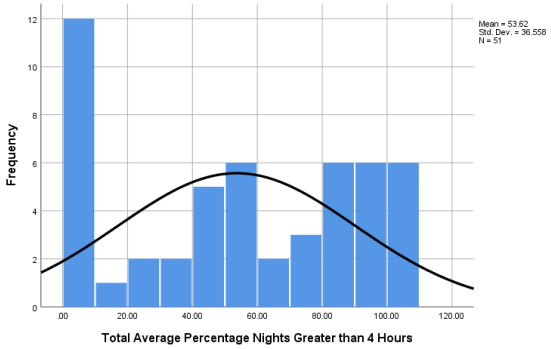


Figure 3.3 –Average Percentage of Nights with NIV Use Greater than Four Hours Across Entire Sample



Between Groups Analysis

No statistically significant difference was found in adherence data for between groups analysis, including custom headgear adaptation, inpatient versus outpatient initiation, type of NIV therapy, mask type, reported complications, age at NIV initiation or reason for NIV discontinuation.

Discussion

The data analyzed in this study provided a descriptive understanding of the population of infants and children enrolled in the NIV program at the Stollery Children's Hospital in 2017, their clinical and demographic characteristics, their follow-up during the first year in the program and their adherence to NIV therapy.

Demographic and Clinical Characteristics

The number of NIV starts at the Stollery in 2017 is large for a specialty service. The increased use of NIV in infants and children has been well documented in the literature, not only in Alberta (Castro-Codesal et al., 2018a), but across Canada (Amin et al., 2014; McDougall et al., 2013; O'Donnell et al., 2006), as well as in Australia (Edwards et al., 2005; Machaalani et al., 2016), and the United Kingdom (Chatwin et al., 2015; Wallis et al., 2011). NIV is one of the least invasive options that allows for long-term treatment of complex and chronic respiratory conditions outside of the hospital setting. The trend of increased use in infants and children may be reflective of the decision to initiate NIV as the first line of treatment, as well as the push for more healthcare services that can be delivered at home and in the community (Guttmann et al., 2009). Additionally, the NIV program at the Stollery serves a wide geographic region, so the large volume of starts in 2017 could be directly connected to its large catchment area. Therefore, it is also important to examine the geographic location of the infants and children who were enrolled in the Stollery's NIV program that year.

Geographic location. The Stollery Children's Hospital has a large catchment area, however the majority of infants and children started on NIV therapy in 2017 came from the Edmonton metro area. This is surprising, as complex diagnoses that may necessitate infants and children to require increased respiratory support are not focused in one geographic location.

However, it does highlight that Edmonton possesses the appropriate resources to treat the local population. There is the possibility that this finding could represent some hesitancy to treat infants and children who live in remote areas with a therapy that requires intensive, multidisciplinary follow-up. This may be because access to comprehensive healthcare services in those areas are limited, necessitating frequent travel and resulting in increased costs to families. Additionally, as a large number of the population that lives in these remote areas belong to Indigenous groups, there might be some underlying bias and prejudice that influence this. Recent research has examined the issue of access to healthcare services for Indigenous peoples in Canada and suggests that there are large gaps that exist in referral for treatment, access to treatment and follow-up for treatment, largely as a result of racist stereotypes and biases that continue to exist in the country (Billie, 2015; Vives, & Sinha, 2019). To determine if there is a hesitancy to initiate NIV therapy for infants and children from remote locations due to the lack of comprehensive healthcare services or due to pre-existing bias and prejudice, physician referral patterns for NIV initiation and reasons surrounding the decision to refer, or not refer, an infant or child for NIV therapy should be examined through further research. Examining the experiences of Indigenous families with chronically ill children who require respiratory support with NIV therapy is another area that might be examined through qualitative work; this would provide the opportunity to understand their experiences as well as the barriers that they face when accessing healthcare services. Another demographic characteristic that was examined in this cohort was the age at NIV initiation.

Age at initiation of non-invasive ventilation. The age at which NIV is initiated is an outcome of interest in this study, with the most common age group for NIV initiation being school-age children as well as a large number of initiations occurring in infants under one year

old. The most common age category being school-age children is similar to other studies of the pediatric population that identify this as the most common age for NIV initiation (Amin et al., 2014; Chatwin et al., 2015; Edwards et al., 2005; Machaalani et al., 2016; O'Donnell et al., 2006). This could be because at this age symptoms of obstructive sleep apnea or other types of upper airway obstruction may become more prominent, such as sleepiness at school or inability to concentrate (Kuhle et al., 2009; Marcus et al., 2012), leading to diagnosis and treatment with NIV. In this cohort, the most common age for NIV initiation due to upper airway obstruction was between 10 and 15 years (28%), which supports this finding. A large number of NIV initiations also occurred in infants under one year old. This is a significant finding, as it may represent the continued improvement of technology, leading to improved outcomes for these vulnerable infants (Castro-Codesal et al., 2018ab; Guttmann et al., 2009; Khirani et al., 2013). Upper airway obstruction was also a common reason for NIV initiation in infants, with 25% happening under six months of age and another 14% happening between six months and one year old. This may be due to diagnoses such as Pierre Robin sequence or tracheomalacia, both of which are diagnosed and treated as infants (Essouri et al., 2005; Leonardis et al., 2013). However, this may also represent increased pressures to do more to save critically ill infants, which can lead to ethical dilemmas (Geevasinga, & Ryan, 2007; Massie, & Gillam, 2015), as well as increased pressures on the healthcare system and the need for appropriate planning and additional health human resources and as these infants age (Guttmann et al., 2009). The age of an infant or child at initiation may also be closely related to the underlying indication for NIV therapy.

Indications for non-invasive ventilation. The reason for NIV initiation in the study population was most commonly upper airway obstruction. While upper airway obstruction is a common reason for NIV initiation, the population enrolled in the Stollery's NIV program is

unique due to the large number of initiations for this reason; at other sites CNS or musculoskeletal disorders were more common (Amin et al., 2014; Machaalani et al., 2016; Wallis et al., 2011). The Stollery has a well-developed otolaryngology department that works closely with the NIV program to refer children for treatment. While many children who have obstructive sleep apnea improve after surgical intervention such as tonsillectomy and adenoidectomy, those with complex medical conditions may continue to require increased respiratory support. These reasons may contribute to the large number of NIV initiations for airway concerns. The majority of those who no longer required NIV therapy throughout 2017 were due to improvement in condition. This may be partially related to the high number of initiations for upper airway concerns, as some of the disorders that cause upper airway obstruction may see improvement with age and growth (Edwards et al., 2005; Leboulanger et al., 2010). Another outcome of interest was the location of initiation of NIV therapy, either in an inpatient or outpatient setting.

Location of non-invasive ventilation initiation. The majority of new initiations occurred in an outpatient setting; the trend of outpatient NIV initiation at the Stollery has been previously documented (Castro-Codesal et al., 2018a) and is similar to other pediatric NIV programs (Amin et al., 2014; Chatwin et al., 2015; Machaalani et al., 2016; Marcus et al., 2005). This may represent a tendency to anticipate the need for NIV initiation in this complex population through elective sleep studies in symptomatic patients, rather than waiting for an acute illness to necessitate emergent initiation. Initiation as outpatients also allows infants and children to become comfortable with the mask interfaces in a familiar setting and limits some of the stress associated with a hospital admission (Castro-Codesal et al., 2019). Outpatient initiation would also be less expensive when compared to the cost of an inpatient hospital bed, representing a

fiscally responsible use of resources. When examining adherence related to inpatient versus outpatient NIV initiation, there was no statistically significant difference between groups for adherence to NIV therapy. This may suggest that adequate supports are in place to ease the transition for families from inpatient to outpatient to help ensure adherence to therapy. The majority of NIV initiations at the Stollery in 2017 occurred in an outpatient setting, a finding that is similar to other pediatric NIV programs. Another similarity between the Stollery's NIV program and other pediatric centres was the type of NIV therapy initiated as well as the mask interfaces used.

Type of non-invasive ventilation therapy and interfaces. The type of NIV therapy and mask interface most commonly used in this population is in alignment with previously published data on NIV use in infants and children. CPAP therapy was the most common type of therapy initiated at the Stollery in 2017, this was also a common finding throughout the literature (Chatwin et al., 2015; Marcus et al., 2005; O'Donnell et al., 2006). In this cohort, the use of CPAP is unsurprising as the majority of NIV initiations were due to upper airway obstruction, with CPAP being the therapy of choice for this respiratory condition (Kuhle et al., 2009; Marcus et al., 2005, 2012). The use of mainly nasal masks for NIV therapy is comparable to other pediatric NIV programs (Chatwin et al., 2015; Machaalani et al., 2016; O'Donnell et al., 2006; Ramirez et al., 2012), and is unsurprising. Full-face masks present additional challenges, including more complications such as an increased aspiration risk (Castro-Codesal et al., 2019; O'Donnell et al., 2006; Ramirez et al., 2012). The most common type of therapy initiated at the Stollery in 2017 was CPAP using a nasal interface; while the use of a nasal interface may help minimize some complications associated with NIV, this cohort experienced a wide range of other complications related to their therapy.

Complications from non-invasive ventilation. This study demonstrated that NIV-related complications are frequent, as previously described (Castro-Codesal et al., 2018b). Although literature about NIV complications varies, nasal symptoms, poor mask fit and skin injury are the most commonly reported complications, as demonstrated with this cohort. The presence of side effects from therapy has previously been found to impact long-term use of NIV therapy (Amaddeo et al., 2016; Ennis et al., 2015; Hull, 2014). In relation to discontinuation of NIV therapy, it is surprising that due to the large number of side effects reported none of the infants and children in this cohort who stopped using NIV therapy over the course of the year cited non-compliance or non-tolerance as the reason. This may represent the successful mitigation of side effects at the Stollery, perhaps through the unique practice of headgear adaptation that is offered through its NIV program.

Custom headgear adaptation. The Stollery offers a unique service, allowing for the custom adaptation of headgear, to the infants and children enrolled in its NIV program. While customization of headgear has been identified as potentially improving adherence (Slifer et al., 2007), the coordinated customization of NIV equipment has not been documented in the literature related to NIV therapy in infants and children, making this a practice unique to the Stollery's NIV program. The majority of infants and children enrolled in the Stollery's program during 2017 had at least one custom headgear adaptation performed. When examining adherence in relation to those who had custom headgear adaptation performed versus those who did not, there was no statistically significant difference found. This is surprising, because custom headgear adaptation attempts to improve NIV equipment fit for infants and children, as commercially made equipment may not fit well. In theory, this should help alleviate side effects and potentially improve adherence, as issues with equipment fit have been cited as one reason for

non-adherence to NIV therapy (Ennis et al., 2015; Hull, 2014; Marcus et al., 2005; O'Donnell et al., 2006; Ramirez et al., 2012). However, this study was not designed to predict relationships or outcomes which may have prevented the ability to find a statistically significant relationship between custom headgear adaptation and adherence to NIV therapy. This is an opportunity for further research, perhaps the development of a randomized-controlled trial or a cohort study, in order to determine if customization of headgear has any significant impact on adherence to NIV therapy. This represents a unique and exciting opportunity to contribute to the literature surrounding NIV therapy in infants and children as minimal information surrounding customization of NIV equipment currently exists. Custom headgear adaptation is provided through the Stollery's NIV program at regular follow-up visits, another outcome of interest for this study.

Follow-Up Practices

Despite identification of clear follow-up intervals, follow-up practices at the Stollery varied, and were difficult to compare to other pediatric NIV programs. Some programs make weekly follow-up phone calls (O'Donnell et al., 2006), others require monthly clinic visits (Marcus et al., 2005), others only have a clinic visit at six months (Edwards et al., 2005; Machaalani et al., 2016), and others do not describe a clear follow-up protocol (Amin et al., 2014; Uong et al., 2007). At the Stollery, the most common follow-up time was between zero and six months, with the average being three months, likely because families require extra support and reassurance during the initial stages of NIV therapy. However, it should be noted that there was no consistent approach to follow-up visits found, with a wide variation in the number of in-person follow-up visits per infant or child in the first year of NIV therapy.

A surprising finding was that most infants and children in 2017 only had one in-person follow-up appointment throughout their first year of enrollment in the NIV program at the Stollery. This could be for several reasons: first, if there were no issues with NIV therapy then families may not see the need for regular follow-up; second, the NIV program at the Stollery recently implemented regular phone calls to families enrolled in the program, these may take the place of in-person clinic visits if there are no immediate or pressing concerns; third, geographic location may make it difficult for regular follow-up, however this is unlikely as the majority of the study population resides in the Edmonton metro area. It would be interesting to further examine the phone call follow-up system that the Stollery has begun to utilize; this might present an opportunity to move away from in-person clinic visits unless a serious concern arises, limiting the amount of travel that medically complex families have to undertake as well as potentially reducing caseload for practitioners working with this population. This may also be an opportunity for more appropriate allocation of resources, allowing for a more streamlined and less time-consuming approach to NIV therapy follow-up. A study that examines adherence to NIV therapy in the context of increased phone call follow-up visits would be interesting, allowing for an understanding of whether this type of change in practice would have a positive or negative impact on the population.

The follow-up practices at the Stollery varied, with most of those enrolled in the program having one follow-up visit within the first year of NIV therapy. It would be interesting to separate the infants and children who had fewer follow-up visits from those who had more follow-up visits to better understand if follow-up practices have an impact on adherence to NIV therapy. This presents an opportunity for further research to examine potential relationships between follow-up practices and adherence to NIV therapy.

Adherence to Non-Invasive Ventilation Therapy

This study found that the total average hours of NIV use was five, the total average hours of NIV use on nights used was six, and the total average percentage of nights with NIV use greater than four hours was 54%. This represents acceptable adherence in this cohort, with some room for improvement. Other studies that have examined adherence in the pediatric population have found variable numbers, including an average of NIV use of four to seven hours (Machaalani et al., 2016; Marcus et al., 2005; O'Donnell et al., 2006; Uong et al., 2007), an average of NIV use on nights used of six hours (Machaalani et al., 2016; O'Donnell et al., 2006), and an average percentage of 80-85% of nights with NIV use greater than four hours, (Machaalani et al., 2016; Uong et al., 2007). Adherence to NIV therapy and related factors should be researched further, perhaps through qualitative work, as it is well documented that infants and children who require NIV therapy also require extensive support from their caregivers (DiFeo et al., 2012; Edwards et al., 2005; King et al., 2014; Machaalani et al., 2016). If caregiver burden becomes overwhelming, families with infants or children who require NIV therapy may require additional supports, training, and potentially the opportunity to access respite care in order to ensure adherence to NIV therapy remains manageable. The variation with which adherence is measured in infants and children requiring NIV therapy suggests that it may be beneficial to develop a standardized criterion for adherence that is specific to the pediatric population. Using the same parameters would allow for adherence data to be compared between NIV programs and for research data related to adherence to be easily interpreted and understood.

Limitations

This research had several imitations. First, it was limited because it used retrospective data. If data was missing or had not been collected, then it was not accessible or able to be used

for analysis. This resulted in some gaps in the data. I set out to collect follow-up and adherence data at standard intervals over the course of the first year of enrollment in the Stollery's NIV program; however, if some information was incomplete or a visit was not documented then it was not included in this research. The results of this study were descriptive in nature and as such are unable to predict relationships or outcomes but suggest possible associations. Additionally, because this data was collected from one site, the results of this study may not be generalizable to other pediatric populations requiring NIV therapy.

Conclusion

The use of NIV in infants and children is increasing and is being used to treat a wide variety of respiratory conditions. This was a retrospective chart review that included the development of a comprehensive patient database and focused on describing the population of infants and children enrolled in the NIV program at the Stollery Children's Hospital in 2017.

This study found that the number of NIV starts at the Stollery in 2017 was large for a specialty service, and that while the Stollery Children's Hospital has a large catchment area, the majority of infants and children started on NIV therapy in 2017 came from the Edmonton metro area. It also found that the most common age group for NIV initiation was school-age children, although a large number of initiations occurred infants under one year old as well, with the most common reason for NIV initiation in the study population being upper airway obstruction. In terms of therapy, the most commonly initiated type of NIV was CPAP with a nasal interface in an outpatient setting. It also found a large number of NIV-related complications; one way of addressing these complications was through the use of custom headgear adaptation, a service unique to the Stollery's NIV program. Follow-up practices varied, with most follow-up occurring between zero and six months after NIV initiation. Adherence in this population was average,

however the lack of standard criteria for adherence in infants and children made it difficult to compare to other pediatric cohorts. This study identified that the program at the Stollery is similar to other pediatric NIV programs in several ways, but also has characteristics that are unique. These unique characteristics provide an exciting opportunity for further research into the population that requires NIV support at the Stollery Children's Hospital.

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CHAPTER FOUR

Summary

NIV is currently used in the pediatric population to treat a wide variety of sleep and respiratory disorders (Castro-Codesal et al., 2018b). It has been established that NIV provides the ventilatory assistance needed for infants and children who have complex medical conditions with respiratory involvement, often avoiding the need for tracheostomy and long-term invasive ventilation, as well as avoiding suboptimal treatment and death (Amin et al., 2014; Castro-Codesal et al., 2018b; Chatwin et al., 2015; Edwards et al., 2005; Leboulanger, et al. 2010). For most infants and children, NIV can ultimately be managed at home as long as appropriate supports are present (Amaddeo et al., 2016; Amin et al., 2014; McDougall et al., 2013), allowing for fewer hospitalizations and improved quality of life, and for some an extended life span (Chatwin et al., 2015). While home NIV therapy is an excellent option for many infants and children, it requires extensive supports to be in place, as well as the coordination of specialized care by a multidisciplinary team that can actively follow their care and provide support (Ennis et al., 2015; Jambhekar et al., 2013; Machaalani et al., 2016; McDougall, et al., 2013; O'Donnell et al., 2006; Ramirez et al., 2013; Wallis et al., 2011). The NIV program at the Stollery Children's Hospital is an example of coordinated specialized care, as it is comprehensive in nature. It includes a multidisciplinary team that works with each infant and child who requires NIV therapy. This program, and the population that it serves, was the focus of my thesis work.

This thesis aimed to describe the clinical characteristics of the population of infants and children over a one-year period who were enrolled in the NIV program at the Stollery Children's Hospital in 2017. The goal of describing this population was that additional research into this area would be feasible, supported and encouraged. In order to facilitate this, a REDCap database

was created; it was designed to be user friendly with the hope that the NIV program at the Stollery would continue to use the database for future research into this area. Once the database was created, data was collected from the charts of all infants and children who were initiated onto NIV therapy in 2017 at the Stollery, and statistical analysis of the data was then conducted using SPSS. Descriptive statistics included the mean or median and standard deviations or ranges for continuous variables, and frequency distributions for all categorical data. Independent t-tests and ANOVAs were also used to compare differences between groups. Once the analysis was completed, the results allowed for a detailed description of the population of infants and children enrolled in the NIV program at the Stollery Children's Hospital in 2017.

Implications

The results of this thesis work highlight the diverse characteristics of the population of infants and children that require respiratory support with NIV therapy at the Stollery Children's Hospital. There are several implications for this thesis work that will be outlined in this section.

Increased Knowledge

To provide excellent care to a specialized population, a thorough understanding of that population is necessary. By describing the population of infants and children who require NIV therapy at the Stollery Children's Hospital, practitioners who work with this population will be able to better understand their needs and the resources they require. Understanding the various indications for NIV therapy, as well as the unique challenges that this population faces will help improve the care that is provided. Practitioners will be well-equipped to answer questions from anxious parents regarding NIV therapy in relation to duration of therapy, complications of therapy and reasons to initiate therapy if they thoroughly understand the population characteristics and trends. Identification of common indications for NIV therapy and

dissemination of this knowledge may help increase referrals to the NIV program. By providing clear descriptions of the characteristics of infants and children that require NIV therapy, not only will practitioners who work with this population be educated, but families who have medically complex infants or children will also benefit.

Increased knowledge is not limited to the practitioners who work with infants and children who require NIV therapy, but also should also include families. By conducting research that describes this population, educational resources can be created for families who are new to the NIV program. This will help them become more comfortable with caring for their infant or child who requires NIV and also provides the unique opportunity to create a community of support. It is clear that infants and children who require NIV therapy also require family members who are competent in their care, however the need for additional caregivers and respite programs cannot be understated. This research also provides the opportunity to foresee special needs for this population, allowing for program planning and resource allocation.

Program Planning and Resource Allocation

Families who are responsible for caring for infants and children who require NIV therapy require extensive training and support. This requires funding for training and education, the availability of additional caregivers and access to respite programs so that families are able to care for their infant or child as well as fulfill their other responsibilities. By understanding the size of the population that requires NIV therapy, it will be easier to plan ahead to ensure that these resources and supports are in place for families.

An understanding of the resources required to provide specialized care to this population will also help drive long-term program planning and resource allocation for the NIV program at the Stollery. This study demonstrated that the Stollery's NIV program sees a large volume of

initiations on a yearly basis. This supports the necessity of the NIV program at the Stollery and the funding it requires. Continued research into this area that also demonstrates the large population the NIV program serves may help procure additional funding and supports in the future.

Research Support

One of the biggest implications of this thesis is the creation of a user-friendly database that will allow continued research in this area to be feasible, supported and encouraged. The REDCap database that was created as part of this thesis is fully functional and is intended for continued use. Future research should utilize the REDCap database that was created for this thesis research, and focus on ongoing collection of data to analyze longitudinal trends of the population that continues to be enrolled in the NIV program at the Stollery Children's Hospital. Utilization of the REDCap database will allow for easy collection and aggregation of data, facilitating timely conduction of research.

It is my hope that by undertaking this project as a practicing registered nurse, not only will physician-led research continue to be supported, but advanced-practice nurses will also see the benefit of research into this area and will be encouraged to undertake it. It also presents an opportunity to involve undergraduate students in nursing and other health-related disciplines in the research process early on in their education, this can help to create an early interest in research and scholarly work for undergraduate students. The ultimate goal is that future research into this area, by all practitioners and disciplines, will be supported and encouraged by the work undertaken in this thesis.

Final Thoughts

This thesis has provided me with an opportunity to open my eyes to the value of nursing research. I have worked with wonderful mentors throughout the duration of my thesis work who have challenged me, pushed me and encouraged me. Examining the population of infants and children who require NIV therapy in detail has been deeply interesting and has re-affirmed what I already knew; that this population is unique and resilient. I hope that I have been able to inspire further research, like suggested in the previous section, into this area with the goal of improving the care that these special infants and children receive.

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APPENDIX A

Confidential

Who Is Using Non-Invasive Ventilation? A Descriptive Study Examining the Population Enrolled in a Pediatric Non-Invasive Ventilaton Program

Demographics

Record ID	_____
Last Name	_____
First Name	_____
Date of Birth	_____
Alberta Healthcare Number	_____
Sex	◯ Male ◯ Female
Geographic Location	◯ Edmonton metro area ◯ Outside Edmonton ◯ Saskatchewan ◯ BC ◯ NWT ◯ Yukon ◯ Nunavut
Reason for NIV Initiation	◯ Central nervous system disorder ◯ Musculoskeletal disorder ◯ Upper airway obstruction ◯ Pulmonary disease ◯ Cardiac Disease ◯ Cancer ◯ Other (unclear)

APPENDIX B

Confidential

Who Is Using Non-Invasive Ventilation? A Descriptive Study Examining the Population Enrolled in a Pediatric Non-Invasive Ventilaton Program

NIV Therapy

Record ID	_____
Initiation Date	_____
Initiation Location	◯ Inpatient ◯ Outpatient ◯ Not Documented
Age at NIV Initiation	◯ < 6 months ◯ 6 months to < 1 year ◯ 1 year to < 5 years ◯ 5 years to < 10 years ◯ 10 years to < 15 years ◯ 15 years and up
Therapy Type	◯ CPAP ◯ BPAP
Mask Type	◯ nasal ◯ full face
NIV Discontinuation	◯ Yes ◯ No
Reason For Discontinuation	◯ Deceased ◯ Transfer (to adult program or other site) ◯ Escalation of therapy (tracheostomy, intubation) ◯ Improvement ◯ Non-compliance/non-tolerance ◯ Unclear/Not documented
Headgear Adaptation Done?	◯ Yes ◯ No
Complications from NIV	◯ Yes ◯ No
Complication Type	☐ Eye Irritation ☐ Nasal Congestion ☐ Skin Breakdown
Concerns with Equipment Fit	◯ Yes ◯ No
Fit Issue	☐ Mask Fit ☐ Mask Leak ☐ Headgear Fit

APPENDIX C

Visits and Adherence

Record ID	_____
0-3 month visit	◯ Yes ◯ No
Date of Visit	_____
0-3 month adherence download available?	◯ Yes ◯ No
Download Date	_____
Days Included	_____
Total Average Hours of Use	_____
Average Hours of Use Days Used	_____
Percentage of Nights with Use >4 Hours	_____
3-6 month visit	◯ Yes ◯ No
Date of Visit	_____
3-6 months adherence download available?	◯ Yes ◯ No
Download Date	_____
Days Included	_____
Total Average Hours of Use	_____
Average Hours of Use Days Used	_____
Percentage of Nights with Use >4 Hours	_____

6-9 month visit	◯ Yes ◯ No
Date of Visit	_____
6-9 months adherence download available?	◯ Yes ◯ No
Download Date	_____
Days Included	_____
Total Average Hours of Use	_____
Average Hours of Use Days Used	_____
Percentage of Nights with Use >4 Hours	_____
9-12 month visit	◯ Yes ◯ No
Date of Visit	_____
9-12 months adherence download available?	◯ Yes ◯ No
Download Date	_____
Days Included	_____
Total Average Hours of Use	_____
Average Hours of Use Days Used	_____
Percentage of Nights with Use >4 Hours	_____
Total Number of Follow-Up Visits	◯ 1 ◯ 2 ◯ 3 ◯ 4
Time from NIV Start to First Follow-up Visit	_____

Total Average Hours of Use	
Total Average Use on Days Used	
Total Average Percentage of Nights with Use > 4 Hours	